Review Article

Primary extradural tumors of the spinal column: A comprehensive treatment guide for the spine surgeon based on the 5th Edition of the World Health Organization bone and soft-tissue tumor classification

ABSTRACT

Background: In 2020, the World Health Organization (WHO) published the 5th version of the soft tissue and bone tumor classification. Based on this novel classification system, we reviewed the current knowledge on all tumor entities with spinal manifestations, their biologic behavior, and most importantly the appropriate treatment options as well as surgical approaches.

Methods: All tumor entities were extracted from the WHO Soft-Tissue and Bone Tumor Classification (5th Edition). PubMed and Google Scholar were searched for the published cases of spinal tumor manifestations for each entity, and the following characteristics were extracted: Growth pattern, ability to metastasize, peak age, incidence, treatment, type of surgical resection indicated, recurrence rate, risk factors, 5-year survival rate, key molecular or genetic alterations, and possible associated tumor syndromes. Surgical treatment strategies as well as nonsurgical treatment recommendations are presented based on the biologic behavior of each lesion.

Results: Out of 163 primary tumor entities of bone and soft tissue, 92 lesions have been reported along the spinal axis. Of these 92 entities, 54 have the potential to metastasize. The peak age ranges from conatal lesions to 72 years. For each tumor entity, we present recommended surgical treatment strategies based on the ability to locally destruct tissue, to grow, recur after resection, undergo malignant transformation as well as survival rates. In addition, potential systemic treatment recommendations for each tumor entity are outlined.

Conclusion: Based on the 5th Edition of the WHO bone and soft tumor classification, we identified 92 out of 163 tumor entities, which potentially can have spinal manifestations. Exact preoperative tissue diagnosis and interdisciplinary case discussions are crucial. Surgical resection is indicated in a significant subset of patients and has to be tailored to the specific biologic behavior of the targeted tumor entity based on the considerations outlined in detail in this article.

Keywords: Chordoma, primary spinal tumors, sarcoma

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INTRODUCTION

The core principles guiding surgical treatment for primary bone and soft-tissue tumors have been introduced by Enneking *et al*. more than 40 years ago and comprise three different types of surgical tumor resection: Intralesional, marginal *en bloc*, and wide *en bloc* resection.[1] It has been suggested that tumor location (intracompartmental versus extracompartmental) and histologic grade should be used

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to determine the mode of resection. Since the introduction of Enneking's system additional research regarding primary bone and soft-tissue tumors, new nonsurgical treatment modalities such as stereotactic radiosurgery or targeted molecular therapies and novel radiographic techniques together have significantly improved demarcating tumor extent and curbing tumor invasion.

This article is based on the 5th Edition of the World Health Organization (WHO) tumor classification of bone and soft-tissue tumors, published in 2020. We compiled the most recent knowledge of all tumor entities, which have been described to occur along the spinal axis and surrounding soft tissues.[2] This comprehensive overview summarizes clinical knowledge as well as imaging findings of all primary, extradural spinal tumors described in the literature.

We describe our treatment algorithms, which is individualized for each tumor entity and loosely based on Enneking's classification system, and modified by contemporary imaging protocols.

METHODS

The 5th Edition of the WHO soft tissue and bone tumors classification, published in 2020 was reviewed and individual tumor entities extracted into a spreadsheet. Medical databases (PubMed and Google Scholar) were searched for publications reporting occurrences of each entity listed in the WHO classification along the spinal axis (spinal bones or paraspinal soft tissues). If an entity has been reported to occur along the spinal axis, a case report with exemplary imaging findings was obtained. For each tumor entity, the following data were extracted from the WHO classification or other key references: Relevant differential diagnoses, growth pattern (infiltrative/destructive), potential for malignant transformation, potential to metastasize, peak age, incidence, recommended type of surgical resection (A, B, C), recurrence rate, treatment, risk factors, 5-year overall survival rate, key molecular or genetic alterations, and possible associated tumor syndromes. All primary bone and soft tissue tumor entities listed in the 5th Edition of the WHO tumor classification were listed in a spreadsheet and a note was made on entities reported to occur along the spinal axis. In a second spreadsheet, exemplary imaging findings of each entity have been listed or say: "Exemplary imaging findings of each entity are listed in a second spreadsheet." Moreover, finally, in a third spreadsheet, the above-mentioned key characteristics for each entity have been listed.

RESULTS

A comprehensive list of all primary bone and soft-tissue tumors, as listed in the most recent WHO classification is given in Appendix 1 and comprises a total of 163 entities. Of note, the following tumors can arise in either bone or soft tissue: Hemangioma, epitheloid hemangioma, epitheloid hemangioendothelioma, angiosarcoma, desomplastic fibroma, fibrosarcoma, chondroma, and osteosarcoma.

Tumor entities are classified by the cell of tumor origin [Appendix 1]. For soft-tissue neoplasms, the following cells of origin are as follows: Adipocytic, fibroblastic and myofibroblastic, fibrohistiocytic, vascular, pericytic (perivascular), smooth muscle, skeletal muscle, gastrointestinal stromal, chondro-osseous, and peripheral nerve sheath. Two further categories exist for all soft-tissue tumors that do not fall into the above mentioned: Tumors of uncertain differentiation and undifferentiated small round cell sarcomas. In the case of bone tumors, the following subclassification based on the cell population of origin exists: Chondrogenic, osteogenic, fibrogenic, vascular, osteoclastic giant cell-rich, or notochordal. Two further subcategories are listed in the WHO classification: Other mesenchymal bone tumors and hematopoietic neoplasms of the bone.

The results of our literature search are outlined in Appendixes 2 and 3 and show that 92 out of 163 entities were reported to occur either in spinal bones or paraspinal soft tissue. We categorized 92 entities with imaging [Appendix 2] and clinical/molecular findings [Appendix 3], as well as recommended surgical and nonsurgical treatment options.

Appendix 3 shows a comprehensive characterization of each tumor by: Growth pattern (infiltrative/locally destructive or not), ability to metastasize, ability to undergo malignant transformation, mean age at diagnosis, incidence, suggested mode of resection (intralesional resection A, marginal *en bloc* resection B, wide, or compartmental *en bloc* excision C), recurrence rate, treatment strategy, tumor risk factors, 5-year overall survival (OS) rate, genetic/molecular tumor characteristics, possible associated tumor syndromes, and corresponding cross -sectional imaging findings are presented in Appendix 2.

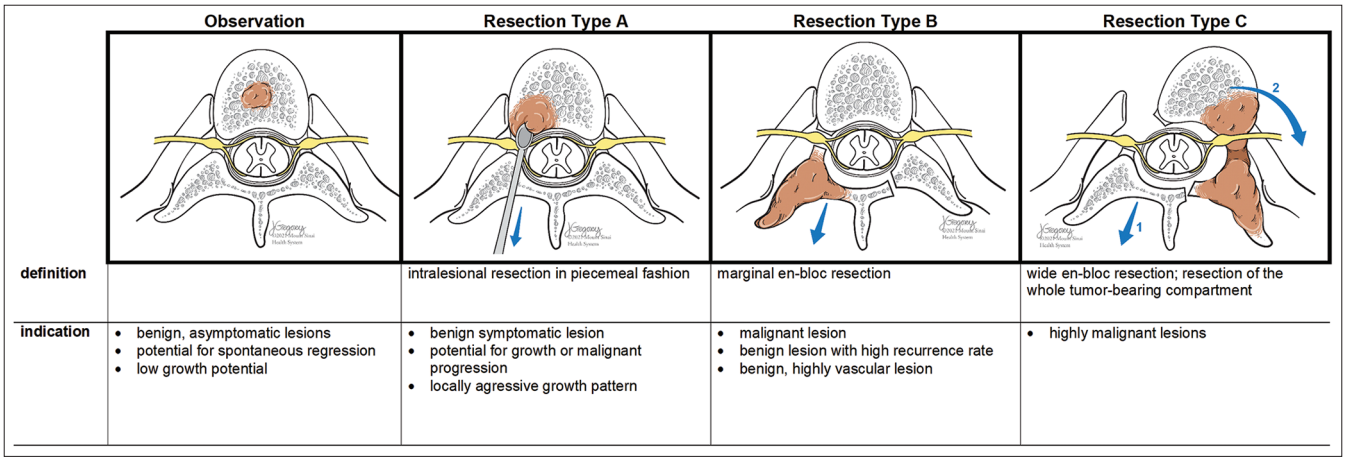
As shown in Appendix 3, the incidence rates for primary extradural spinal bone or soft-tissue tumors range from 2% (hemangioma) to a low of only two published cases for spinal nodular fasciitis. The survival rates of malignant lesions range from 94% for 5 year OS for ossifying fibromyxoid tumor to 7% for dedifferentiated osteosarcoma. A total of 54 entities

are capable of forming metastases, 1 additional entity can form so called benign pulmonary metastases (chondroblastoma). The peak age ranges from conatal lesions (lymphangioma) to 72 years (pleomorphic rhabdomyosarcoma).

DISCUSSION

The most recent edition of the WHO classification of bone and soft-tissue tumors lists a total of 163 tumor entities, out of which 92 have been previously reported in the literature to potentially occur in the spine. Surgical resection is the integral part of treatment for most of these lesions and follows the overriding principles outlined by Enneking *et al*. in 1980,[1] as shown in Figure 1. Type B and C resections are more complex than type A resections with higher rates of complications; however, type B/C resections are associated with superior oncologic outcome as compared to type A resections for malignant lesions.[3] It must be noted that given to the unique anatomy of the spine, when compared to long bones, in many cases, a type B resection might be indicated. While type B resections may not be technically feasible, spine surgeons may opt for type C resections with a wider excision. Figure 2 provides an overview of important growth characteristics of malignant bone and soft-tissue tumors. As indicated, the growth pattern of sarcomas is infiltrative. Even with a rim of reactive tissue, the pseudocapsule may act only as a weak barrier to prevent tumor spread. While the pseudocapsule has been shown to restrict tumor permeation after radio- or chemotherapy it is not a true barrier for tumor spread.[4] Cortical bone as well as major fascial planes, such as pleura or peritoneum are considered bone fide barriers. It is known from radiologic studies that infiltrating tumor nests, known as skip lesions, outside the primary tumor can be depicted on magnetic resonance imaging (MRI) in up to 16.5% of patients.[5] As shown in Figure 2, once the cortical bone of the vertebra is breached, the tumor cells can freely spread until they reach the next level of solid barrier [routes A-D in Figure 2]. As has been shown in previous correlating studies between preoperative imaging and intraoperative histologic analysis, the mean discrepancy between tumor margin on preoperative MRI and intraoperative histology for osteosarcomas is 5 mm.[6,7] Since short-tau inversion recovery and postcontrast T1 imaging overestimates the tumor extend by 1.68 cm, tumor outline is best depicted on noncontrast-enhanced T1 images.[8] Therefore, in our own experience if a malignant tumor is confined to one compartment, we perform either a type B resection with a margin of 5 mm on top of the tumor outline in the preoperative noncontrast T1 images, or we perform a type C resection, which will remove the whole tumor bearing compartment. If a malignant tumor extends into more than one compartment (e.g., cortical bone erosion in the case of vertebral osteosarcomas), we prefect to discuss either neo-adjuvant treatment to "downsize" the tumor (the more compartments the tumor extends into, the less likely a true wide *en bloc* resection can be achieved) or surgery to encompass an *en bloc* resection of the primary tumor bearing compartment plus the extension into a neighboring compartment with a safety margin of at least 5 mm.

How to incorporate these principles into surgical practice depends on the index level. In the case of C1 and C2, oncologic resections type B and C in most cases require a transmandibular approach [Figure 3]. When compared to the rest of the cervical spine negative margins are less likely to be obtained due to the anatomical complexity of the region.[9] For the rest of the mobile spine the WBB system has been proposed to choose the appropriate approach or combination of approach to perform a type B or C resection [Figure 4].[10] The choice of approach for oncologic resections of the sacrum is mainly determined by



	Observation	Resection Type A	Resection Type B	Resection Type C
definition		intralesional resection in piecemeal fashion	marginal en-bloc resection	wide en-bloc resection; resection of the whole tumor-bearing compartment
indication	• benign, asymptomatic lesions • potential for spontaneous regression • low growth potential	• benign symptomatic lesion • potential for growth or malignant progression • locally agressive growth pattern	• malignant lesion • benign lesion with high recurrence rate • benign, highly vascular lesion	• highly malignant lesions

Figure 1: Overview of the three different surgical types of resection in the treatment of spinal tumors

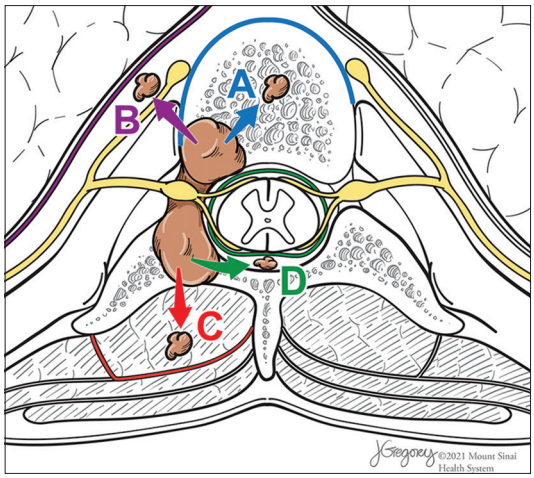


Figure 2: Illustration of potential routes of and barriers to spread of spinal sarcomas. Lesions, detached from the primary tumor are termed skip lesions. Barriers to skip lesions are: (A) Cortical bone, (B) pleura in cases when the lateral vertebral cortex has been breached, (C) muscle fascia in case of posterior cortical tumor breach, (D) dura in case of cortical breach of the spinal canal

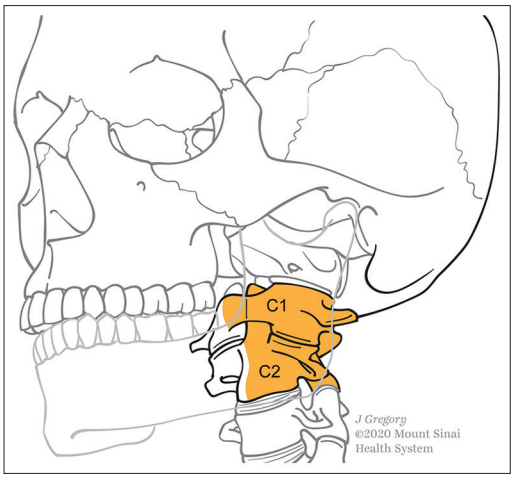


Figure 3: Oncologic resection (type B and C) of primary tumors of C1 and C2 are carried out in most cases utilizing a transmandibular approach

the anatomic level of the lesion as well as the presence of visceral tumor infiltration. Figure 5 outlines our institutional algorithm to such lesions. Only lesions located below the inferior margin of the sacroiliac joint (SIJ) without visceral invasion are resected using a posterior-only approach. All other lesions are resected using an anterior/posterior approach. Reconstruction of the pelvic ring is necessary if more than 50% of the SIJs are resected. In instances where the tumor extends by more than 3 cm beyond the SIJ, we consider them as primarily inoperable (due to the large tumor volume and complexity of reconstruction).

Reconstruction of large resection cavities in many cases requires the involvement of plastic surgery and is beyond the scope of this article.

En bloc resections are technically demanding and have been shown to have higher complication rates when compared to type A resections, particularly when more than 1 level is being resected (Spiessberger A, PubMed ID pending), even though lesion etiology seems to have less impact on complication rates.

Given the profile of potential complications in the case of type B and C resections, rigorous preoperative planning is of paramount importance. Neurologic deficits are particularly devastating to patients and should be avoided at all costs. Other than direct mechanical injury, ischemic spinal cord injury has been reported to occur on rare occasions.[11,12] Even though spinal cord blood supply is highly collateralized, postoperative infarcts can be a complication due to segmental vessel ligation.[11,13] Spinal cord blood supply is established through the anterior spinal artery, a branching vessel of the vertebral arteries, as well as from as posterior spinal arteries through branching vessels of either vertebral or posterior inferior cerebellar arteries. Collateral flow is provided through variable radiculomedullary vessels, typically 2-3 cervical (bilaterally equal), 2-3 thoracic (left more than right), and 0-1 lumbar (left more than right).[12] Three major radiculomedullary vessels are described: The artery of cervical enlargement (usually a branching vessel from the ascending cervical artery at C6), the artery "von Haller" (usually the T5 segmental vessel) as well as the artery of Adamkiewicz (usually the T10 segmental vessel).[14] Watershed areas, susceptible to ischemic infarction in cases of hypotension or hypoxia have been suggested in the mid thoracic spine as well as the posterior aspect of the conus medullaris.[15] Type B and C resections require segmental artery ligation; however, recent studies have suggested that up to three adjacent segmental vessel can be sacrificed safely.[16,17] We believe, that caution should be taken when ligating one of the three major radiculomedullary vessels, as described above. Preoperative high-resolution CT angiography can help localize the level of these three vessels. Intraoperative temporary nerve root/segmental vessel clamping with cautious observation of motor evoked potential/somatosensory evoked potential is important as well. In addition, intraoperative and postoperative hypotension should be avoided at all costs when a major radiculomedullary vessel has been sacrificed. It is also worth noting that the choice of vasopressor might make a difference as well. Animal studies comparing norepinephrine and phenylephrine in their properties to increase spinal cord perfusion in the setting of hypotension have shown, that norephinephrine provides better restoration of blood flow and oxygenation.[18] One should also recognize that radiculomedullary vessel ligation may not only render the patient more susceptible to ischemic cord injury, but also

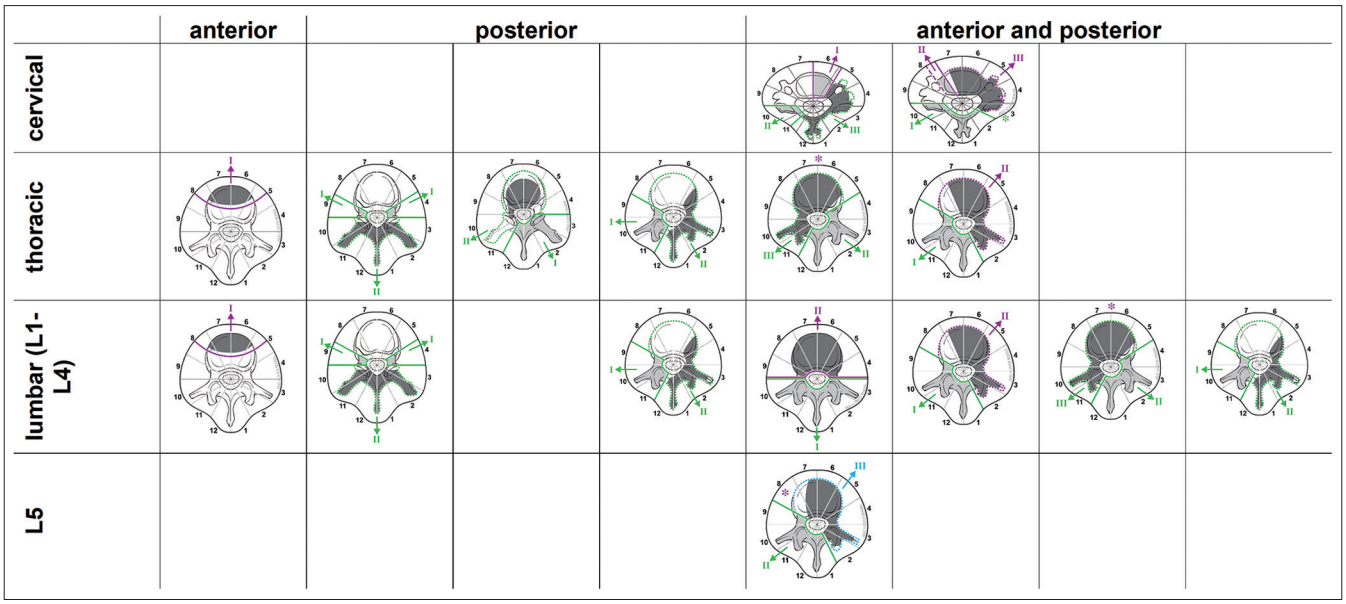


Figure 4: Choice of approach for oncologic tumor resections of the subaxial spine (excluding sacrum), based on the Weinstein-Boriani-Biagini system. The vertebra is divided in 12 zones and based on the tumor location either an anterior approach (purple), posterior approach (green) are chosen. For each scenario the osteotomy sites are indicated and in cases necessitating combined approach the suggested order is indicated (I, II)

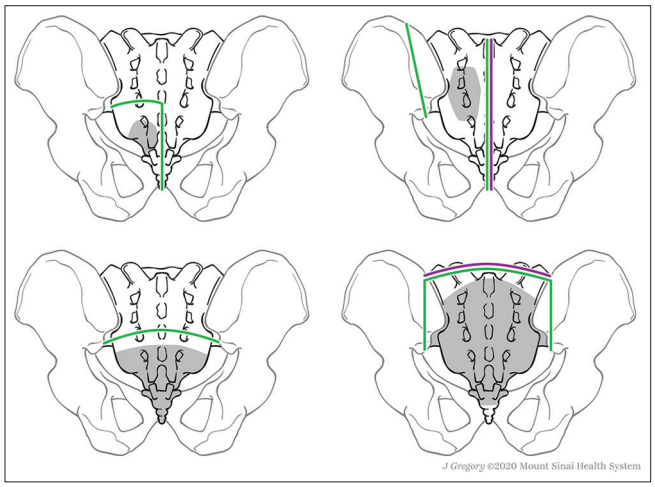


Figure 5: Sacral resections can be performed in a posterior only approach (green) or combined anterior/posterior approach (purple and green). It is our practice to liberally perform anterior approaches to separate the tumor from the visceral structures, to ligate the bilateral internal iliac arteries, to harvest abdominal wall flaps for reconstruction

surgical trauma to segmental vessels or vertebral arteries can lead to embolic cord infarcts caused by vessel dissections.[19] In the case of cervical type B and C resections, preoperative endovascular sacrifice of one vertebral artery in case high degree tumor encasement (>180°) can be safely performed following careful study of a CT angiogram of both cervical vessels and posterior circulation. Side dominance, potential stenoses, size or absence of the posterior communicating arteries (in the case of fetal posterior cerebral artery variants) must be determined. Moreover, temporary endovascular balloon occlusion can be considered to determine the safety of vessel occlusion.

CONCLUSION

Based on the 5th Edition of the WHO bone and soft tumor classification, we identified 92 out of 163 tumor entities, which potentially can have spinal manifestations. Exact preoperative tissue diagnosis and interdisciplinary case discussions are crucial. Surgical planning has to be tailored to the specific biologic behavior of the targeted tumor entity based on the considerations outlined in detail in this article.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Enneking WF, Spanier SS, Goodman MA. A system for the surgical staging of musculoskeletal sarcoma. Clin Orthop Relat Res 1980;153:106-20.
2. WHO Classification of Tumours Editorial Board. Soft Tissue and Bone Tumours: WHO Classification of Tumours (Medicine). 5th ed. Lyon: World Health Organization; 2020.
3. Yamazaki T, McLoughlin GS, Patel S, Rhines LD, Fourney DR. Feasibility and safety of *en bloc* resection for primary spine tumors: A systematic review by the Spine Oncology Study Group. Spine (Phila Pa 1976) 2009;34:S31-8.
4. Gitelis S, Thomas R, Templeton A, Schajowicz F. Characterization of the pseudocapsule of soft-tissue sarcomas. An experimental study in rats. Clin Orthop Relat Res 1989;246:285-92.
5. Saifuddin A, Sharif B, Oliveira I, Kalus S, Barnett J, Pressney I. The incidence of skip metastases on whole bone MRI in high-grade bone sarcomas. Skeletal Radiol 2020;49:945-54.

6. Jin T, Deng ZP, Liu WF, Xu HR, Li Y, Niu XH. Magnetic resonance imaging for the assessment of long bone tumors. Chin Med J (Engl) 2017;130:2547-50.
7. O'Flanagan SJ, Stack JP, McGee HM, Dervan P, Hurson B. Imaging of intramedullary tumour spread in osteosarcoma. A comparison of techniques. J Bone Joint Surg Br 1991;73:998-1001.
8. Putta T, Gibikote S, Madhuri V, Walter N. Accuracy of various MRI sequences in determining the tumour margin in musculoskeletal tumours. Pol J Radiol 2016;81:540-8.
9. Molina CA, Ames CP, Chou D, Rhines LD, Hsieh PC, Zadnik PL, *et al.* Outcomes following attempted *en bloc* resection of cervical chordomas in the C-1 and C-2 region versus the subaxial region: A multiinstitutional experience. J Neurosurg Spine 2014;21:348-56.
10. Boriani S, Weinstein JN, Biagini R. Primary bone tumors of the spine. Terminology and surgical staging. Spine (Phila Pa 1976) 1997;22:1036-44.
11. Ng YH, Kato S, Demura S, Shinmura K, Yokogawa N, Nakade Y, *et al.* Delayed ischemic spinal cord injury after total *en bloc* spondylectomy in the thoracic spine. J Orthop Sci 2021 Jan 8;S0949-2658(20)30373-0.
12. Melissano G, Civilini E, Bertoglio L, Calliari F, Campos Moraes Amato A, Chiesa R. Angio-CT imaging of the spinal cord vascularisation: A pictorial essay. Eur J Vasc Endovasc Surg 2010;39:436-40.
13. Du C, Liu S, Jia F, Liu X, Wei F. Delayed incomplete paraplegia after *en bloc* spondylectomy of thoracic metastasis, a case report. 2020.
14. Gailloud P. The artery of von Haller: A constant anterior radiculomedullary artery at the upper thoracic level. Neurosurgery 2013;73:1034-43.
15. Gailloud P, Gregg L, Galan P, Becker D, Pardo C. Periconal arterial anastomotic circle and posterior lumbosacral watershed zone of the spinal cord. J Neurointerv Surg 2015;7:848-53.
16. Kato S, Kawahara N, Tomita K, Murakami H, Demura S, Fujimaki Y. Effects on spinal cord blood flow and neurologic function secondary to interruption of bilateral segmental arteries which supply the artery of Adamkiewicz: An experimental study using a dog model. Spine (Phila Pa 1976) 2008;33:1533-41.
17. Murakami H, Kawahara N, Tomita K, Demura S, Kato S, Yoshioka K. Does interruption of the artery of Adamkiewicz during total *en bloc* spondylectomy affect neurologic function? Spine (Phila Pa 1976) 2010;35:E1187-92.
18. Streijger F, So K, Manouchehri N, Gheorghe A, Okon EB, Chan RM, *et al.* A direct comparison between norepinephrine and phenylephrine for augmenting spinal cord perfusion in a porcine model of spinal cord injury. J Neurotrauma 2018;35:1345-57.
19. Montalvo M, Bayer A, Azher I, Knopf L, Yaghi S. Spinal cord infarction because of spontaneous vertebral artery dissection. Stroke 2018;49:e314-7.

APPENDIXES

Appendix 1: List of bone and soft tissue tumors

- Soft tissue tumours
  - Adipocytic tumours
    - Angiolipoma
    - Atypical lipomatous tumour/well-differentiated liposarcoma
    - Atypical spindle cell/pleomorphic lipomatous tumour
    - Chondroid lipoma
    - Hibernoma
    - Lipoblastoma and lipoblastomatosis
    - Lipoma
    - Lipomatosis
    - Lipomatosis of nerve
    - Liposarcoma, dedifferentiated
    - Liposarcoma, myxoid
    - Liposarcoma, myxoid pleomorphic
    - Liposarcoma, pleomorphic
    - Myolipoma of soft tissue
    - Spindle cell lipoma and pleomorphic lipoma
  - Fibroblastic and myofibroblastic tumours
    - Acral fibromyxoma
    - Angiofibroma of soft tissue
    - Angiomyofibroblastoma
    - Calcifying aponeurotic fibroma
    - Cellular angiofibroma
    - Dermatofibrosarcoma protuberans
    - Desmoid fibromatosis
    - Desmoplastic fibroblastoma
    - Elastofibroma
    - EWSR1-SMAD3-positive fibroblastic tumour (emerging)
    - Fibroma of tendon sheath
    - Fibromatosis colli
    - Fibrosarcoma, adult
    - Fibrosarcoma, infantile
    - Fibrous hamartoma of infancy
    - Gardner fibroma
    - Giant cell fibroblastoma
    - Inclusion body fibromatosis
    - Inflammatory myofibroblastic tumour
    - Ischaemic fasciitis
    - Juvenile hyaline fibromatosis
    - Lipofibromatosis
    - Low-grade fibromyxoid sarcoma
    - Low-grade myofibroblastic sarcoma
    - Myofibroblastoma
    - Myositis ossificans and fibro-osseous pseudotumour of digits
    - Myxofibrosarcoma
    - Myxoinflammatory fibroblastic sarcoma
    - Nodular fasciitis
    - Nuchal-type fibroma
    - Palmar fibromatosis and plantar fibromatosis
    - Proliferative fasciitis and proliferative myositis
    - Sclerosing epithelioid fibrosarcoma
    - Solitary fibrous tumour

Contd...

Appendix 1: Contd...

- Soft tissue tumours
  - Fibroblastic and myofibroblastic tumours
    - Superficial CD34-positive fibroblastic tumour
  - So-called fibrohistiocytic tumours
    - Deep fibrous histiocytoma
    - Giant cell tumour of soft tissue
    - Plexiform fibrohistiocytic tumour
    - Tenosynovial giant cell tumour
  - Vascular tumours
    - Angiosarcoma
    - Haemangioendothelioma, composite
    - Haemangioendothelioma, epitheloid
    - Haemangioendothelioma, pseudomyogenic
    - Haemangioendothelioma, retiform
    - Haemangioma
    - Haemangioma, anastomosing
    - Haemangioma, epitheloid
    - Intramuscular angioma
    - Kaposi sarcoma
    - Lymphangioma and lymphangiomatosis
    - Papillary intralymphatic angioendothelioma
    - Synovial haemangioma
    - Tufted angioma and kaposiform haemangioendothelioma
    - Venous haemangioma, venous
  - Pericytic (perivascular) tumours
    - Angioleiomyoma
    - Glomus tumour
    - Myopericytoma, including myofibroma
  - Smooth muscle tumours
    - EBV-associated smooth muscle tumour
    - Inflammatory leiomyosarcoma
    - Leiomyoma
    - Leiomyosarcoma
  - Skeletal muscle tumours
    - Ectomesenchymoma
    - Rhabdomyoma
    - Rhabdomyosarcoma, alveolar
    - Rhabdomyosarcoma, embryonal
    - Rhabdomyosarcoma, pleomorphic
    - Rhabdomyosarcoma, spindle cell
  - Gastrointestinal stromal tumour
    - Gastrointestinal stromal tumour
  - Chondro-osseous tumours
    - Soft tissue chondroma
    - Extraskeletal osteosarcoma
  - Peripheral nerve sheath tumours
    - Benign triton tumour/neuromuscular choristoma
    - Dermal nerve sheath myxoma
    - Ectopic meningioma and meningothelial hamartoma
    - Granular cell tumour
    - Hybrid nerve sheath tumour
    - Malignant melanotic nerve sheath tumour
    - Malignant peripheral nerve sheath tumour
    - Neurofibroma

Contd...

Appendix 1: Contd...

- Perineurioma
- Schwannoma
- Solitary circumscribed neuroma
- Tumours of uncertain differentiation
  - Alveolar soft part sarcoma
  - Angiomatoid fibrous histiocytoma
  - Atypical fibroxanthoma
  - Clear cell sarcoma of soft tissue
  - Deep (aggressive) angiomyxoma
  - Desmoplastic small round cell tumour
  - Epithelioid sarcoma
  - Extrarenal rhabdoid tumour
  - Extraskeletal myxoid chondrosarcoma
  - Haemosiderotic fibrolipomatous tumour
  - Intimal sarcoma
  - Intramuscular myxoma
  - Juxta-articular myxoma
  - Myoepithelioma, myoepithelial carcinoma, and mixed tumour
  - NTRK-rearranged spindle cell neoplasm (emerging)
  - Ossifying fibromyxoid tumour
  - PEComa
  - Phosphaturic mesenchymal tumour
  - Pleomorphic hyalinizing angiectatic tumour of soft parts
  - Synovial sarcoma
  - Undifferentiated sarcoma
  - Undifferentiated small round cell sarcomas of bone and soft tissue
- CIC-rearranged sarcoma
  - Ewing sarcoma
  - Round cell sarcoma with EWSR1-non-ETS fusions
  - Sarcoma with BCOR genetic alterations
- Bone tumours
- Chondrogenic tumours
  - Bizarre parosteal osteochondromatous proliferation
  - Central atypical cartilaginous tumour/chondrosarcoma, Grade 1
  - Chondroblastoma
  - Chondromyxoid fibroma
  - Chondrosarcoma, central Grades 2 and 3
  - Chondrosarcoma, clear cell
  - Chondrosarcoma, dedifferentiated
  - Chondrosarcoma, mesenchymal
  - Chondrosarcoma, periosteal
  - Chondrosarcoma, secondary peripheral Grades 2 and 3
  - Enchondroma
  - Osteochondroma
  - Osteochondromyxoma
  - Periosteal chondroma
  - Secondary peripheral atypical cartilaginous tumour/chondrosarcoma, Grade 1
  - Subungual exostosis
  - Synovial chondromatosis
- Osteogenic tumours
  - Osteoblastoma
  - Osteoid osteoma
  - Osteoma
  - Osteosarcoma

Contd...

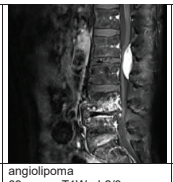
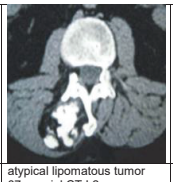
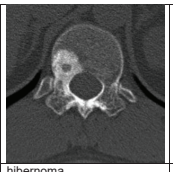
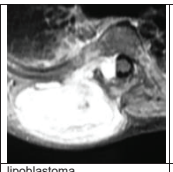
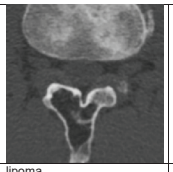
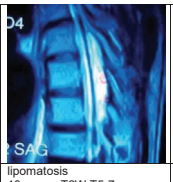
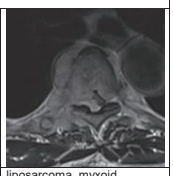
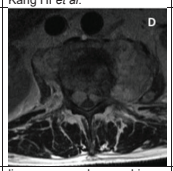
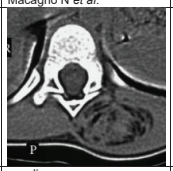
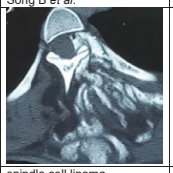
Appendix 1: Contd...

- Osteosarcoma, high-grade surface
- Osteosarcoma, low-grade central
- Osteosarcoma, parosteal
- Osteosarcoma, periosteal
- Osteosarcoma, secondary
- Fibrogenic tumours (see soft tissue tumors)
- Vascular tumours of bone (seesoft tissue tumors)
- Osteoclastic giant cell-rich tumours
  - Aneurysmal bone cyst
  - Giant cell tumour of bone
  - Nonossifying fibroma
- Notochordal tumours
  - Benign notochordal cell tumour
  - Conventional chordoma
  - Dedifferentiated chordoma
  - Poorly differentiated chordoma
- Other mesenchymal tumors of bone (see soft-tissue tumors)
- Haematopoietic neoplasms of bone
  - Erdheim-chester disease
  - Langerhans cell histiocytosis
  - Plasmacytoma of bone
  - Primary non-Hodgkin lymphoma of bone
  - Rosai-Dorfman disease

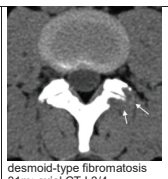
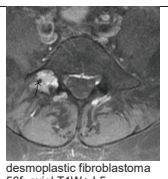
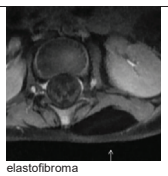
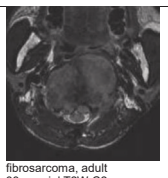
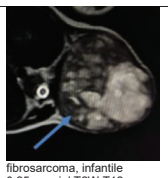
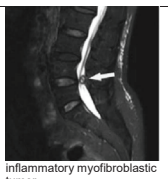
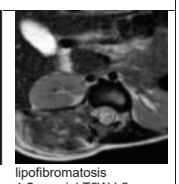
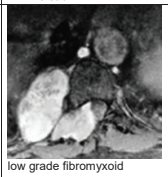
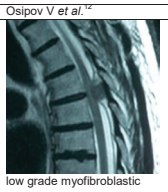
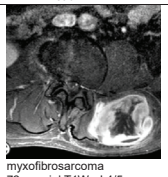
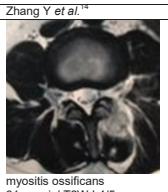
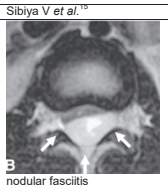
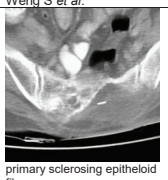
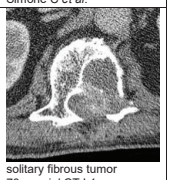
EBV - Ebstein Barr virus

Appendix 2: Radiographic overview of primary spinal neoplasms

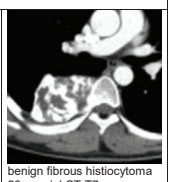
adipocytic tumors

							
tumor entity patient age/sex; imaging	angiolipoma 69m; sag T1W+ L2/3	atypical lipomatous tumor 67m; axial CT L2	hibernoma 71f; axial CT L3	lipoblastoma 0.8m; axial T1W+ T7	lipoma 54m; axial CT L3	lipomatosis 18m; sag T2W T5-7	liposarcoma, myxoid 79m; axial T2W T5
source	Kang HI *et al.*[1]	Macagno N *et al.*[2]	Song B *et al.*[3]	Gupta G *et al.*[4]	Teekhasaenee C *et al.*[5]	Rajput D *et al.*[6]	Rovlias A *et al.*[7]
							
tumor entity patient age/sex; imaging	liposarcoma, pleomorphic 60m; axial T2W L2	myolipoma 4m; axial CT lower thoracic	spindle cell lipoma 14f; axial CT T7				
source	Morales-Codina A *et al.*[8]	Parratt M *et al.*[9]	Sah H *et al.*[10]				

fibroblastic / myofibroblastic tumors

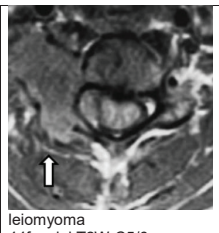
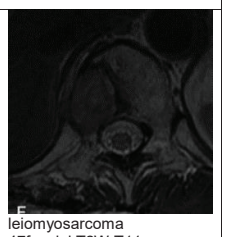
							
tumor entity patient age/sex; imaging	desmoid-type fibromatosis 31m; axial CT L3/4	desmoplastic fibroblastoma 56f; axial T1W+ L5	elastofibroma 1m; axial T1W+ L2	fibrosarcoma, adult 33m; axial T2W C2	fibrosarcoma, infantile 0.25m; axial T2W T12	inflammatory myofibroblastic tumor 56m; sag T2W L4/5	lipofibromatosis 1.5m; axial T2W L2
source	Kim SJ *et al.*.[11]	Osipov V *et al.*[12]	Bulam H *et al.*[13]	Zhang Y *et al.*[14]	Sibiya V *et al.*[15]	Weng S *et al.*[16]	Simone C *et al.*[17]
							
tumor entity patient age/sex; imaging	low grade fibromyxoid sarcoma 46f; axial T1W+ T9	low grade myofibroblastic sarcoma 55m; sag T2W T5	myxofibrosarcoma 72m; axial T1W+ L4/5	myositis ossificans 31m; axial T2W L4/5	nodular fasciitis 7m; axial T2W L2	primary sclerosing epitheloid fibrosarcoma 48f; axial CT S2	solitary fibrous tumor 73m; axial CT L1
source	Singhania BK *et al.*[18]	Hadjigorgiu GF *et al.*[19]	Nam DH *et al.*[20]	Abdallah A *et al.*[21]	Kubota K *et al.*[22]	Chow LTC *et al.*[23]	Farooq Z *et al.*[24]

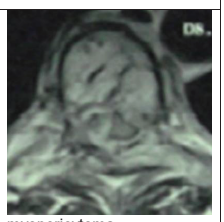
fibrohistiocytic tumors

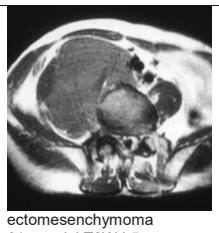
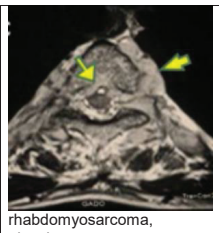
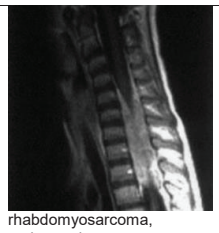
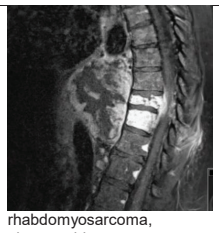
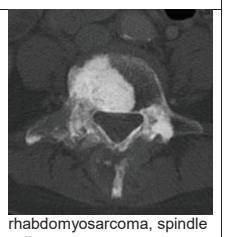
	
tumor entity patient age/sex; imaging	benign fibrous histiocytoma 23m; axial CT T7
source	Liu S *et al.*[25]

Contd...

Appendix 2: Contd...

smooth muscle tumors			
			
tumor entity patient age/sex; imaging	EBV associated smooth muscle tumor 24m: axial T1W T2	leiomyoma 44f; axial T2W C5/6	leiomyosarcoma 47f; axial T2W T11
source	Ehresman JS *et al.* [26]	Iwakura R *et al.* [27]	Lo TH *et al.* [28]

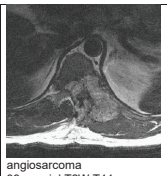
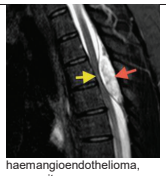
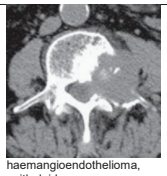
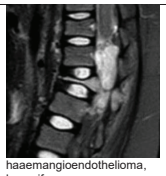
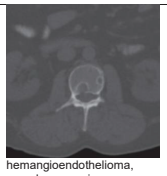
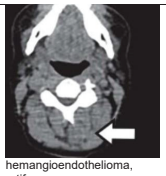
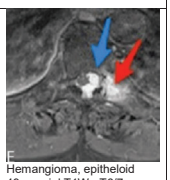
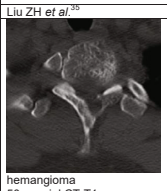
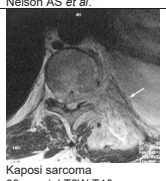
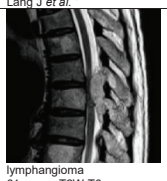
pericytic tumors	
	
tumor entity patient age/sex; imaging	myopericytoma 50f: axial T2W T8
source	Agrawal N *et al.* [29]

skeletal muscle tumors					
					
tumor entity patient age/sex; imaging	ectomesenchymoma 61m; axial T2W L5	rhabdomyosarcoma, alveolar 20f; axial T2w T3/4	rhabdomyosarcoma, embryonal 5m; sag T1W+ C5-T3	rhabdomyosarcoma, pleomorphic 59m; sag T2W T9/10	rhabdomyosarcoma, spindle cell 70f; axial CT L5
source	Kimura S *et al.* [30]	Sofiene B *et al.* [31]	Rumboldt Z *et al.* [32]	Spaleholz M *et al.* [33]	Tagami M *et al.* [34]

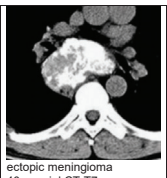
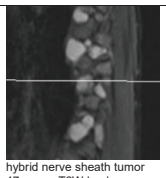
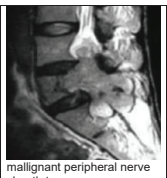
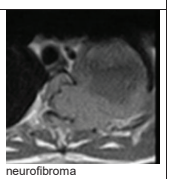
Contd...

Appendix 2: Contd...

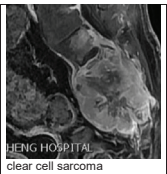
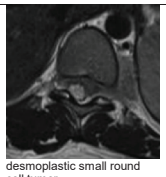
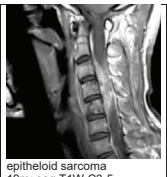
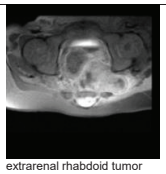
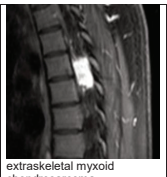
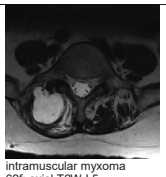
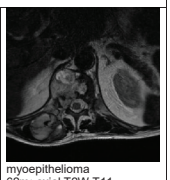
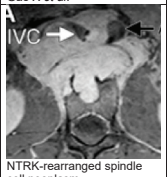
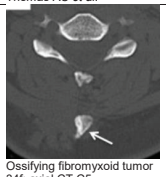
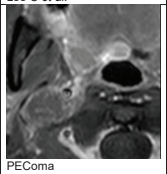
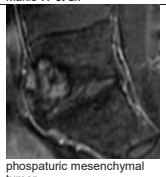
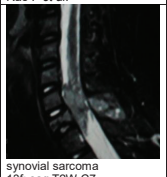
vascular tumors

							
tumor entity patient age/sex; imaging	angiosarcoma 39m; axial T2W T11	haemangioendothelioma, composite 38f; sag T2W T4-6	haemangioendothelioma, epitheloid 76m; axial CT L4	haaemangioendothelioma, kaposiforme 5f; sag T1W+ L1	hemangioendothelioma, pseudomyogenic 59m; axial CT L2	hemangioendothelioma, retiform 52m; axial CT C3	Hemangioma, epitheloid 49m; axial T1W+ T6/7
source	Liu ZH *et al.*[35]	Nelson AS *et al.*[36]	Lang J *et al.*[37]	Eseonu K *et al.*[38]	Bryanton M *et al.*[39]	Vadrucci M *et al.*[40]	O'Shea BM *et al.*[41]
							
tumor entity patient age/sex; imaging	hemangioma 50m; axial CT T1	Kaposi sarcoma 33m; axial T2W T10	lymphangioma 61m: sag T2W T6				
source	Moattari KA *et al.*[42]	Di Bella S *et al.*[43]	Fattahi A *et al.*[44]				

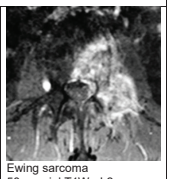
peripheral nerve sheath tumours

				
tumor entity patient age/sex; imaging	ectopic meningioma 40m; axial CT T7	hybrid nerve sheath tumor 47m; sag T2W lumbar	mallignant peripheral nerve sheath tumor 35f; sag T2W L5	neurofibroma 34f; axial T1W T3
source	Lu C *et al.*[45]	Hussain NS *et al.*[46]	Senapati SB *et al.*[47]	Kumar SA *et al.*[48]

uncertain differentiation

							
tumor entity patient age/sex; imaging	clear cell sarcoma 46m; sag T1W+ sacral	desmoplastic small round cell tumor 18f; axial T2W T11	epitheloid sarcoma 19m; sag T1W C3-5	extrarenal rhabdoid tumor 0.7m; axial T1W+ sacral	extraskeletal myxoid chondrosarcoma 29m; sag T1W T5	intramuscular myxoma 62f; axial T2W L5	myoepithelioma 62m; axial T2W T11
source	Gao X *et al.*[49]	Thomas AC *et al.*[50]	Lee C *et al.*[51]	Makis W *et al.*[52]	Rao P *et al.*[53]	Choi DY *et al.*[54]	Ghermandi R *et al.*[55]
							
tumor entity patient age/sex; imaging	NTRK-rearranged spindle cell neoplasm 21m; axial T1W+ L1	Ossifying fibromyxoid tumor 34f; axial CT C5	PEComa 29f; axial T1W+ C2	phospaturic mesenchymal tumor 54m; sag T1W L5	synovial sarcoma 13f; sag T2W C7		
source	Dupuis M *et al.*[56]	De Wandeler T *et al.*[57]	Komune N *et al.*[58]	Maehara J *et al.*[59]	Yang M *et al.*[60]		

undifferentiated small round cell tumors

	
tumor entity patient age/sex; imaging	Ewing sarcoma 58m; axial T1W+ L2
source	Iacoangeli M *et al.*[61]

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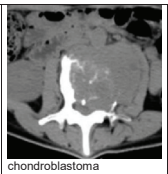
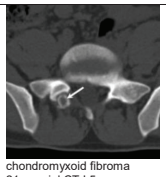
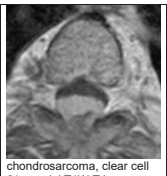
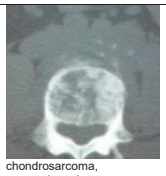
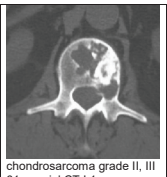
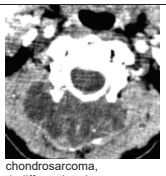
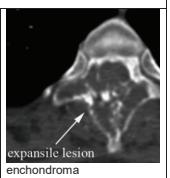
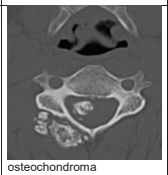
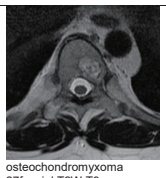
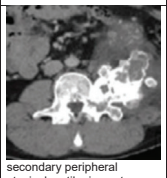
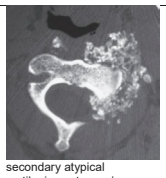
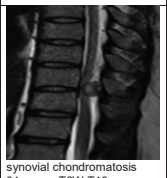
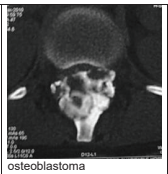
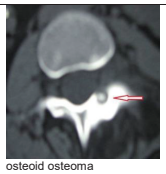
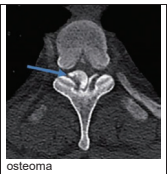
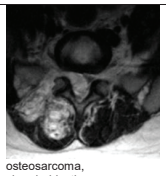
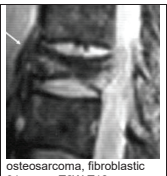
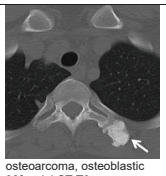
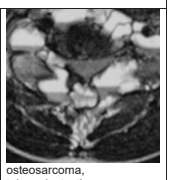
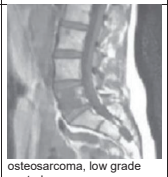
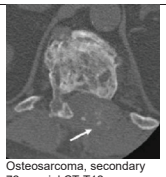
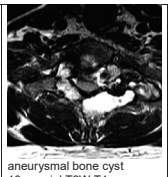
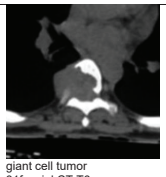
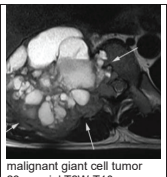
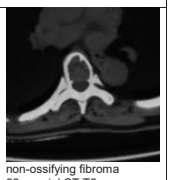
Appendix 2: Contd...

chondrogenic tumors							
							
tumor entity patient age/sex; imaging	chondroblastoma 22f; axial CT L3	chondromyxoid fibroma 21m; axial CT L5	chondrosarcoma, clear cell 61m; axial T1W T4	chondrosarcoma, mesenchymal 30m; axial CT L4	chondrosarcoma grade II, III 61m; axial CT L1	chondrosarcoma, dedifferentiated 81m; axial CT C2	enchondroma 49f; axial CT T3
source	Shakir TM *et al.* [62]	Gutierrez-Gonzalez R *et al.* [63]	Paidakakos NA *et al.* [64]	Fukuda A *et al.* [65]	Strike SA *et al.* [66]	Matsumoto Y *et al.* [67]	Guo J *et al.* [68]
							
tumor entity patient age/sex; imaging	osteochondroma 36f; axial CT C3	osteochondromyxoma 27f: axial T2W T9	secondary peripheral atypical cartilaginous tumor 29m; axial CT L2	secondary atypical cartilaginous tumor / chondrosarcoma grade I 46m;axial CT C2	synovial chondromatosis 31m; sag T2W T10		
source	Yakkanti R *et al.* [69]	Yu W *et al.* [70]	Adimonye A *et al.* [71]	Strike SA *et al.* [66]	Ghorpade RS *et al.* [72]		

osteogenic tumors							
							
tumor entity patient age/sex; imaging	osteoblastoma 20f; axial CT T12	osteoid osteoma 8m; axial CT T12	osteoma 47m; axial CT T3	osteosarcoma, chondroblastic 25f; axial T2W L5	osteosarcoma, fibroblastic 81m; sag T2W T12	osteoarcoma, osteoblastic 32f; axial CT T3	osteosarcoma, teleangiectactic 18f; axial T2W T11
source	Bhargava P *et al.* [73]	Sapkas G *et al.* [74]	Forlizzi J *et al.* [75]	Scudday TS *et al.* [76]	Kokubo Y *et al.* [77]	Katonis P *et al.* [78]	Katonis P *et al.* [78]
							
tumor entity patient age/sex; imaging	osteosarcoma, low grade central 42f: sag T1W L5	Osteosarcoma, secondary 72m; axial CT T12					
source	Asdi ARB *et al.* [79]	Sofka CM *et al.* [80]					

osteoclastic giant cell-rich tumors				
				
tumor entity patient age/sex; imaging	aneurysmal bone cyst 18m; axial T2W T1	giant cell tumor 31f; axial CT T6	malignant giant cell tumor 23m; axial T2W T10	non-ossifying fibroma 52m; axial CT T5
source	Eun J *et al.* [81]	Zheng K *et al.* [82]	Yu H *et al.* [83]	Yang J *et al.* [84]

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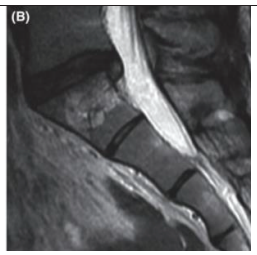
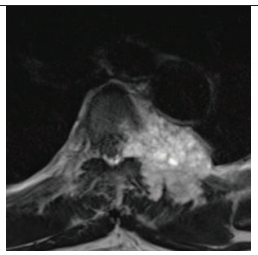
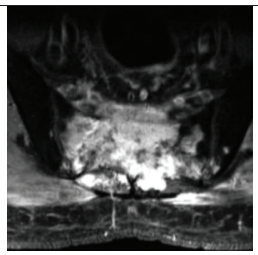
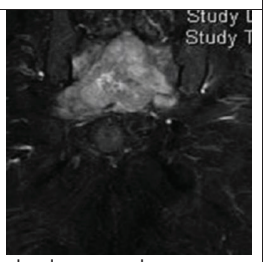
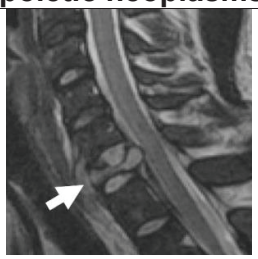
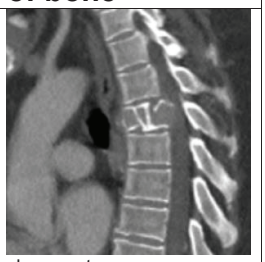
Appendix 2: Contd...

notochordal tumors				
				
tumor entity patient age/sex; imaging	benign notochordal tumor 22m; sag T2W S1	chordoma, conventional 64m; axial T2W T6	chordoma, dedifferentiated 41f; axial T2W S2	chordoma, poorly differentiated 58m; cor T2W S1
source	Tateda S *et al*. [85]	Liu S *et al*. [86]	Kim SC *et al*. [87]	Rekhi B *et al*. [88]

haematopoietic neoplasms of bone		
		
tumor entity patient age/sex; imaging	Non-Hodgkin lymphoma of the bone 23m; sag T2W C7	plasmacytoma 64f; sag CT T5
source	Smith ZA *et al*. [89]	Röpke EF *et al*. [90]

REFERENCES

1. Kang HI, Kim TW, Kim EJ, Park KH. Angiolipoma on the Lumbar Spine. Korean J Spine 2017;14:112-4.
2. Macagno N, Fuentes S, de Pinieux G, *et al*. Paravertebral well-differentiated liposarcoma with low-grade osteosarcomatous component: Case report with 11-year follow-up, radiological, pathological, and genetic data, and literature review. Case Rep Pathol 2017;2017:2346316.
3. Song B, Ryu HJ, Lee C, Moon KC. Intraosseous hibernoma: A rare and unique intraosseous lesion. J Pathol Transl Med 2017;51: 499-504.
4. Gupta G, Garg R, Wadhwa C, Bansal H, Kaushal RK. A rare primary dumbbell lipoblastoma. Asian J Neurosurg 2018;13:83-5.
5. Teekhasaenee C, Kita K, Takegami K, Kawakita E, Sakakibara T, Kasai Y. Intraosseous lipoma of the third lumbar spine: A case report. J Med Case Rep 2015;9:52.
6. Rajput D, Srivastava AK, Kumar R. Spinal epidural lipomatosis: An unusual cause of relapsing and remitting paraparesis. J Pediatr Neurosci 2010;5:150-2.
7. Rovlias A, Balanika A, Nomikos A, Melissaris S. Primary myxoid liposarcoma of the upper thoracic spine in an elderly patient. J Neurosci Rural Pract. 2017;8(Suppl 1): S120-S122.
8. Morales-Codina AM, Martin-Benlloch JA, Corbellas Aparicio M. Primary pleomorphic liposarcoma of the spine. Case report and review of the literature. Int J Surg Case Rep 2016;25:114-9.
9. Parratt MT, Gokaraju K, Spiegelberg BG, Miles J, Cannon SR, Briggs TW. Myolipoma affecting the erector spinae: A case report in a child. Case Rep Med 2009;2009:520126.
10. Sah H, Shrestha DK, Rajbhandari B, Pradhnang AB, Shilpakar SK. Recurrent extensively calcified spindle cell lipoma in cervicothoracic paraspinous region mimicking liposarcoma: A case report. Interdisciplinary Neurosurgery. 2020;20: 100665.
11. Kim SJ, Ha DH, Lee SM, Kang H. Desmoid type fibromatosis in the facet joint of lumbar spine: case report and review of literature. Korean J Radiol 2013;14:818-22.
12. Osipov V, Carrera GF. Collagenous fibroma (desmoplastic fibroblastoma) with vertebral body erosion. Sarcoma 2009;2009:682687.
13. Bulam H, Sezgin B, Findikcioglu K, Cesur N. A 1-Year-Old Boy With Paraspinal Elastofibroma: The Youngest Diagnosed Elastofibroma. Ann Thorac Surg 2015;100:302-4.
14. Zhang Y, Ai F, Fu S, *et al*. An application of posterior cervical and transoral approaches to treating primary malignant osseous tumors in craniovertebral junction. Zhongguo xiu fu Chong Jian wai ke za zhi= Zhongguo Xiufu Chongjian Waike Zazhi= Chinese J Reparative and Reconstructive Surgery 2020;34:1149-57.
15. Sibiya V, Manimekalai V, Manikandan P. Infantile fibrosarcoma-An unusual presentation. J Adv Med Med Res 2017:1-4.
16. Wang S, Chen L, Cao Z, Mao X, Zhang L, Wang B. Inflammatory myofibroblastic tumor of the lumbar spinal canal: A Case Report With Literature Review. Med (Baltimore) 2017;96:e6488.
17. Simone C, Marco C, Alessandro G, Alberto R, Stefano B. Lipofibromatosis of the paravertebral soft tissue in a child of two years old. Clin Oncol 2017;2:1262.
18. Singhania BK, Jini J, Pramanick G. Primary paravertebral low-grade fibromyxoid sarcoma. Apollo Med 2019;16:36.
19. Hadjigeorgiou GF, Samaras V, Varsos V. Low-grade myofibroblastic sarcoma of the thoracic spine: Report of an extreme rare case. British journal of neurosurgery 2017;31:731-3.
20. Nam DH, Kim TW, Park KH, Kim JO. High grade myxofibrosarcoma in the paravertebral muscle: A case report. Korean J Spine 2010;7:268-71.
21. Abdallah A, Gokcedag A, Ofluoglu AE, Emel E. Non-traumatic myositis ossificans in the lumbar spine. American J Case Rep 2014;15: 421.
22. Kubota K, Okada S, Maeda T, *et al*. Extradural nodular fasciitis arising in the spinal canal. Spine (Phila Pa 1976) 2012;37:E133-137.
23. Chow L, Lui Y, Kumta S, Allen P. Primary sclerosing epithelioid fibrosarcoma of the sacrum: A case report and review of the literature. Journal of clinical pathology 2004;57:90-4.
24. Farooq Z, Badar Z, Zaccarini D, Tavernier FB, Mohamed A, Mangla R. Recurrent solitary fibrous tumor of lumbar spine with vertebral body involvement: imaging features and differential diagnosis with report of a case. Rad Case Rep 2016;11:450-5.
25. Liu S, Zhou X, Song A, *et al*. Management of giant benign fibrous histiocytoma in the spinal region with pleural involvement: A case report. Med 2019;98.
26. Ehresman JS, Ahmed AK, Palsgrove DN, Pennington Z, Goodwin CR, Sciubba DM. Epstein-Barr virus-associated smooth muscle tumor involving the spine of an HIV-infected patient: Case report and review of the literature. J Clin Neurosci. 2018;52:145-50.
27. Iwakura R, Tachibana T, Maruo K, *et al*. Benign metastasizing leiomyoma mimicking dumbbell tumor of the spine: A report of two cases. Interdisciplinary Neurosurg 2017;7:71-2.
28. Lo T, Van Rooij W, Teepen J, Verhagen I. Primary leiomyosarcoma of the spine. Neuroradiology 1995;37:465-7.
29. Agrawal N, Nag K. Myopericytoma of the thoracic spine: A case report and review of literature. The Spine J 2013;13:e23-e27.
30. Kimura S, Kawaguchi S, Wada T, Nagoya S, Yamashita T, Kikuchi K. Rhabdomyosarcoma arising from a dormant dumbbell ganglioneuroma of the lumbar spine: a case report. Spine 2002;27:E513-E517.
31. Sofiene B, Asma B, Adenane B, *et al*. Primary spinal epidural rhabdomyosarcoma of upper thoracic spine. J Spine 2014;3:2.
32. Rumboldt Z, Jednačak H, Talan-Hranilović J, Kalousek V. Spinal epidural rhabdomyosarcoma. Acta Neurochirurgica 2004;146:195-7.
33. Spalteholz M, Gulow J. Pleomorphic rhabdomyosarcoma infiltrating thoracic spine in a 59-year-old female patient: Case report. GMS Interdisciplinary plastic and reconstructive surgery DGPW. 2017;6.
34. Tagami Y, Sugita S, Kubo T, *et al*. Spindle cell rhabdomyosarcoma in a lumbar vertebra with FUS-TFCP2 fusion. Pathology-Research and Practice 2019;215:152399.
35. Liu Z-H, Lee S-T, Jung S-M, Tu P-H. Primary spinal angiosarcoma. Journal of Clinical Neuroscience. 2010;17:387-389.
36. Nelson A, Biswas S, Baborie A, Pigott T, Bhojak M, Das K. Composite hemangioendothelioma in the spine: A case report. Neurographics 2014;4:97-100.
37. Lang J, Chen L, Chen B, *et al*. Epithelioid angiosarcoma of the spine: a case report of a rare bone tumor. Oncology Letters 2014;7:2170-4.
38. Eseonu K, Anwar H. Kaposiform Hemangioendothelioma in the thoracic spine: A case report and review of the literature. Int J Spine Surg 2020;14:426-32.
39. Bryanton M, Makis W. Pseudomyogenic hemangioendothelioma mimicking multiple myeloma on 18F-FDG PET/CT, followed by spontaneous regression. Clin Nuclear Med 2015;40:579-81.
40. Vadrucci M, Vandoni A, Gilardi L. Incidental Detection of Retiform Hemangioendothelioma by 68Ga DOTATOC PET/CT. Clin Nuclear Med 2017;42:149-51.
41. O'Shea BM, Kim J. Epithelioid hemangioma of the spine: Two cases. Radiol Case Rep 2014;9:984.
42. Moattari KA, Melikian R, Khurana SK. Rapidly Progressive Myelopathy Caused by Aggressive Vertebral Hemangioma. Case Reports Orthopedics 2019;2019.
43. Di Bella S, Capone A, Olearo F, *et al*. Vertebral lesions from AIDS-related Kaposi's sarcoma. Current HIV Research 2011;9:270-5.
44. Fattahi A, Taheri M, Majdi M. Lymphangioma of the thoracic spine with epidural compression: A case report. Iranian J Med Sci 2019;44:172.
45. Lu C, Hu X, Xu M, *et al*. Posterior mediastinal ectopic meningioma: A case report. World J Surg Oncol 2015;13:1-4.
46. Hussain NS, Specht CS, Frauenhoffer E, Glantz M, Harbaugh K. Hybrid Neurofibroma-Schwannoma. Cureus 2016;8.
47. Senapati SB, Mishra SS, Dhir MK, Das S. Malignant peripheral

nerve sheath tumor in spine: two case reports. South Asian J Cancer 2013;2:141.
48. Kumar SA, Kumar M, Malgonde M. Dumbbell-shaped neurofibroma of the upper thoracic spine: A case report. South Asian J Cancer 2013;2:26.
49. Gao X, Zhao C, Wang J, *et al*. Surgical management and outcomes of spinal clear cell sarcoma: A retrospective study of five cases and literature review. J Bone Oncol 2017;6:27-31.
50. Thomas AC, Rajashekharan R. A rare case of dumbbell-shaped primary intraspinal peripheral primitive neuroectodermal tumor involving thoracic spinal epidural space. Asian journal of neurosurgery. 2018;13:1216.
51. Lee C, Choe WJ, Kim N. Epithelioid sarcoma in the cervical spine: A case report. Korean J Spine 2015;12:165.
52. Makis W, Ciarallo A, Hickeson M. Malignant extrarenal rhabdoid tumor of the spine: staging and evaluation of response to therapy with F-18 FDG PET/CT. Clinical Nuclear Med 2011;36:599-602.
53. Rao P, Colen RR, Bruner JM, Meis JM. Extraskeletal myxoid chondrosarcoma presenting as an intradural spinal mass: Report of a rare clinical presentation with an emphasis on differential diagnostic considerations. Rare Tumors 2014;6:150-3.
54. Choi DY, Kim JT, Kim J, Lee HJ. Atypical intramuscular myxoma of the lumbosacral paraspinal muscle: The first case report in Asian. Journal of Korean Neurosurgical Society 2015;58:566.
55. Ghermandi R, Pala E, Gambarotti M, Colangeli S, Boriani S. Myoepithelioma of the spine: first case in the literature. Eur Rev Med Pharmacol Sci 2014;18(suppl 1):66-71.
56. Dupuis M, Shen Y, Curcio C, *et al*. Successful treatment of lipofibromatosis-like neural tumor of the lumbar spine with an NTRK-fusion inhibitor. Clin Sarcoma Res 2020;10:1-7.
57. De Wandeler T, Van Gestel D, Tchofo PJ. Ossifying Fibromyxoid Tumor with Spinal Cord Compression and Epiduritis. J Belgian Society Radiol 2019;103(1).
58. Komune N, Masuda S, Yasumatsu R, *et al*. Malignant perivascular epithelioid cell tumor mimicking jugular foramen schwannoma: A case report and literature review. Heliyon. 2020;6:e03200.
59. Maehara J, Yamashita K, Hiwatashi A, *et al*. Primary phosphaturic mesenchymal tumour of the lumbar spine: utility of 68Ga-DOTATOC PET/CT findings. BJR| Case Rep 2016:20150497.
60. Yang M, Zhong N, Zhao C, *et al*. Surgical management and outcome of synovial sarcoma in the spine. World J Surg Oncol 2018;16:1-11.
61. Iacoangeli M, Dobran M, Di Rienzo A, *et al*. Nonmetastatic Ewing's sarcoma of the lumbar spine in an adult patient. Case Rep Oncol Med 2012;2012.
62. Shakir TM, Li W, Niu C, Zhang M. Chondroblastoma of the lumbar vertebra. Journal of Ayub Medical College Abbottabad. 2018;30(4): 604-606.
63. Gutiérrez-González R, De Reina L, Saab A, Jiménez-Heffernan J, García-Uría J. Chondromyxoid fibroma of the lumbar spine: Case report and literature review. European Spine J 2012;21:458-462.
64. Paidakakos NA, Rovlias A, Rokas E, Theodoropoulos S, Katafygiotis P. Primary clear cell chondrosarcoma of the spine: A case report of a rare entity and a review of the literature. Case Rep Oncological Med 2012;2012.
65. Fukuda A, de Castro Oliveira DL, Fernandes Joaquim A, Amstalden EMI, de Souza Queiroz L, Reis F. Vertebral body chondrosarcoma with metastasis to the scalp. BJR| Case Reports 2019;5:20180037.
66. Strike SA, McCarthy EF. Chondrosarcoma of the spine: A series of 16 cases and a review of the literature. The Iowa Orthopaedic J 2011;31: 154.
67. Matsumoto Y, Takahashi Y, Harimaya K, *et al*. Dedifferentiated chondrosarcoma of the cervical spine: A case report. World J Surg Oncol 2013;11:1-6.
68. Guo J, Gao J-z, Guo L-j, Yin Z-x, He E-x. Large enchondroma of the thoracic spine: A rare case report and review of the literature. BMC Musculoskeletal Disorders 2017;18:1-5.
69. Yakkanti R, Onyekwelu I, Carreon LY, Dimar JR. Solitary osteochondroma of the spine—a case series: Review of solitary osteochondroma with myelopathic symptoms. Global Spine J 2018;8:323-39.
70. Yu W, Zhang Z-Z, Wang O, Huang M-Q, Xia W-B, Guermazi A. Ring sign: An imaging sign for osteochondromyxoma in Carney complex. Quantitative Imaging Med Surg 2019;9:1958.
71. Adimonye A. A rare and interesting case of a massive secondary spinal chondrosarcoma and review of the literature. Case Rep Orthopedics 2015;2015.
72. Ghorpade RS, Lokanath YK. Synovial chondromatosis of dorsal spine: Case report of rare pathological entity and review. Korean J Spine 2016;13:196.
73. Bhargava P, Singh R, Garg BB. Dorsal spine osteoblastoma. Asian J Neurosurg 2016;11:180.
74. Sapkas G, Efstathopoulos NE, Papadakis M. Undiagnosed osteoid osteoma of the spine presenting as painful scoliosis from adolescence to adulthood: A case report. Scoliosis 2009;4:1-4.
75. Forlizzi J, Mennona S, McDonnell M, Chiappetta G, Rogalski S. Discovery of thoracic osteoma after elective lumbar spinal surgery: a case report. AME Case Reports 2019;3.
76. Scudday TS, Danisa OA, Zuckerman LM. Management of pelvic chondroblastic osteosarcoma after urgent spinal decompression-A report of 2 cases. J Orthopaedic Case Reports 2016;6:72.
77. Kokubo Y, Uchida K, Kobayashi S, *et al*. Primary osteosarcoma of the thoracic spine: report of an unusual elderly patient with autopsy findings. Spinal cord. 2005;43(8): 508-511.
78. Katonis P, Datsis G, Karantanas A, *et al*. Spinal osteosarcoma. Clinical Medicine Insights: Oncology. 2013;7:CMO. S10099.
79. Asdi ARB, Kamal AF. Low-grade intramedullary osteosarcoma presenting with multiple sclerotic bone lesions. Radiol Case Rep 2018;13:1042-7.
80. Sofka CM, Ciavarra G, Saboeiro G, Ghelman B. Paget's disease of the spine and secondary osteosarcoma. HSS Journal®. 2006;2:188-90.
81. Eun J, Oh Y. A case report of aneurysmal bone cyst of the thoracic spine treated by serial anterior and posterior fusion. Med 2019;98.
82. Zheng K, Xu M, Wang B, Yu Xc, Hu Yc. Giant cell tumor of the mobile spine occurring in pregnancy: A case report and literature review. Orthopaedic Surg 2017;9:252-6.
83. Yu H, Shi R, Peng Z-G, Yu B-H, Cui J-L. Primary malignancy in giant cell tumor of thoracic vertebrae: A case report. Med 2018;97.
84. Yang J, Zhong N, Hu J, Yang X, Xiao J. Total vertebral involvement by benign fibrous histiocytoma: A case report and literature review. J Bone Oncology 2020;20:100274.
85. Tateda S, Hashimoto K, Aizawa T, *et al*. Diagnosis of benign notochordal cell tumor of the spine: Is a biopsy necessary? Clin Case Rep 2018;6:63.
86. Liu S, Zhou X, Song A, *et al*. Surgical treatment of giant chordoma in the thoracic spine combining thoracoscopic and posterior spinal surgery: A case report. Med 2019;98.
87. Kim SC, Cho W, Chang U-K, Youn SM. Two cases of dedifferentiated chordoma in the sacrum. Korean J Spine 2015;12:230.
88. Rekhi B, Banerjee D, Ramadwar M, Bajpai J, Jambhekar NA. Clinicopathologic features of four rare types of chordomas, confirmed by brachyury immunostaining. Indian J Pathol and Microbiology 2017;60:350.
89. Smith ZA, Sedrak MF, Khoo LT. Primary bony non-Hodgkin lymphoma of the cervical spine: A case report. J Medical Case Rep 2010;4:1-4.
90. Röpke E, Theissig F, Ulrich G, *et al*. Solitary plasmacytoma of thoracic vertebra in a woman with Lynch syndrome: A case report. Int J Surg Case Rep 2019;65:44-47.

Appendix 3: Characteristics of primary spinal neoplasms

Adipocytic Tumors

	Important differential diagnosis	Infiltrating/malignant transformation/ local destruction/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	treatment	Risk factors	5y OS	Protein/gene	Possible associated Tumor syndroms
Angiolipoma[1]	-	possible/no/no	2nd-3rd decade	~1% of spinal tumors	A	<5%	resection	-	NA	PRKD2	-
Atypical lipomatous tumour/ well-differentiated liposarcoma[1, 2]	-	yes/yes (2-20%)/no	4th-5th decade	50% of liposarcomas	B	11%	resection; RT or Sx + RT**	-	92%	MDM2 and/or CDK4 amplification	Li Fraumeni
Hibernoma[1, 3]	atypical lipomas, well-differentiated liposarcoma	no/no/no	38	1% of adipocytic tumors	A	<5%	resection if symptomatic	-	NA	Chromosome 11q13 deletion	MEN 1
Lipoblastoma[1, 4]	lipoma, hibernoma, liposarcoma	no/no/no	4	?	B	13-46%*	resection	-	NA	PLAG1	-
Lipoma[1, 5]	liposarcoma	no/no/no	36	14 cases	A	<5%	resection if symptomatic	obesity	NA	HMGA2 protein	PTEN hamartoma tumor syndrome
Lipomatosis[1, 6-9]	-	no/no/no	68	6% of patients with spinal stenosis)	A	5%*	resection if symptomatic	steroid, alcohol	NA	-	-
Liposarcoma, myxoid[1, 10]	-	yes/-/yes	childhood, 4th-5th decade	20% of liposarcomas	C	12-25%	resection; RT*, CH*	-	89%	FUS-DDIT3 or rarely EWSR1-DDIT3	-
Liposarcoma, pleomorphic[1, 11, 12]	-	Yes/-/yes	7th decade	<5% of liposarcomas	C	45%	resection, CH	-	57%	-	-
Myolipoma[1]	-	no/no/no	adulthood	?	A		resection if symptomatic	-	NA	HMGA2	-
Spindle cell lipoma[1]	Liposarcoma	possible/no/no	45-60	?	A	<5%	resection	-	NA	Chromosome 13 and/or 16 deletion	-

Fibroblastic and Myofibroblastic Tumours

	Important differential diagnosis	Infiltrating/ malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/gene	Possible associated Tumor syndroms
Desmoid-type fibromatosis[1,13]	-	yes/no/no	37-39	0.4/100000	B or C**	33%	resection vs close observation; CH alone in FAP associated cases	trauma, pregnancy	52%***	*CTNNB1* or APC mutations	FAP
Desmoplastic fibroblastoma[1]	-	yes/no/no	6th decade	?	A	<5%	resection	-	NA	t (2;11)(q31;q12)	-
Elastofibroma[1]	-	no/no/no	7th-8th decade	2%	A	<5%	resection if symptomatic	-	NA	gains of 6p25-q25 and Xq12-q22	-
Fibrosarcoma, adult[1,14,15]	-	yes/yes/yes	50	<1% of STS	C	20%	resection + CH vs neoadjuvant CH + resection*	foreign body, previous irradiation	55%	*STRN3-NTRK3* fusion	-

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Appendix 3: Contd...

Fibrosarcoma, infantile[1]	-	yes/no/ rarely (8-15%)	1	0.5/100000	B	25%-40%	resection + CH/TG	-	89%	*ETV6-NTRK3* fusion	-
Inflammatory myofibroblastic tumor[1,16-18]	-	occasionally/no/ yes	10	0.04%	B or C	25%-86%****	Resection vs RT, CH/TT*	-	15m*****	*ALK*	-
Lipofibromatosis[1,19]	-	yes/no/no	1	?	B	70%	resection	-	NA	fusions (*EGF, HBEGF, TGF-α)* to *EGFR (HER1)* or *EGFR*	-
Low grade fibromyxoid sarcoma[1,20]		yes/no/rarely	41	5% of STS	B or C	64%	Resection + CH*	-	83%	*FUS-CREB3L2 or FUS-CREB3L1 gene* fusions	-
Low grade myofibroblastic sarcoma[1]	-	yes/yes/rarely	42	12 cases	C	25%	resection vs RT, CH*	-	83%	-	-
Myofibrosarcoma[1]	-	yes/yes/yes	66	?	C	40%	resection + RT	-	-65%	gains of chromosome 5p	-
Myositis ossificans[1,21]	-	no/no/no/no	young adults	0.4%	A	<5%	resection if symptomatic	-	NA	*COL1A1-USP6* fusion	-
Nodular fasciitis[1,22]	-	rarely/rarely/rarely	young adults	2 cases	A	<5%	resection	trauma	NA	*USP6* rearrangement	-
Primary sclerosing epitheloid fibrosarcoma[1,23,24]	-	yes/no/yes (85%)	40	89 cases	C	50%	Resection + CH*	-	66%******	*EWSR1-CREB3L1* fusion	-
Solitary fibrous tumor[1,25-27]	-	no/no/yes	55	0.14/100000	B or C	10-30%	resection + CH/TT*	-	49-83%	*NAB2-STAT6* rearrangement	-

Fibrohistiocytic Tumors

	Important differential diagnosis	Infiltrating/malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/gene	Possible associated Tumor syndroms
Deep benign fibrous histiocytoma[1]	-	no/no/rarely (5%)	37 years	< 1% of fibrohistiocytic tumours	A or B	20%	resection	-	NA	*PRKCB* or *PRKCD* rearrangements	-

Smooth Muscle Tumors

	Important differential diagnosis	Infiltrating/ malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/gene	Possible associated Tumor syndroms
EBV associated smooth muscle tumor[1,28]	-	no/no/no	32	11 spinal cases	A	<5%	resection if symptomatic; immunreconstitution	immunodeficiency	-	-	-
Leiomyoma[1,29]	-	no/rare/no	37	<10% of leiomyoma	B	<10%	resection if symptomatic	uterine leimyomas	-	*KAT6B-KANSL1* and *EWSR1-PBX3* fusion genes	-
Leiomyosarcoma[1,30]		yes/NA/yes	7th decade	11% of STS	C	40%	Resection, RT and CH	radiation	57%	complex	Li-Fraumeni syndrome, hereditary retinoblastoma

Pericytic Tumors

	Important differential diagnosis	Infiltrating/malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/ gene	Possible associated Tumor syndroms
Myopericytoma[1,31]	-	no/no/very rare*******	52	?	A	<5%	resection if symptomatic	AIDS	-	*PDGFRB* gene	Infantile myofibromatosis

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Skeletal Muscle Tumors											
	Important differential diagnosis	**Infiltrating/malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Ectomesenchymoma[1,32]	-	yes/NA/yes	0.6	50 cases	C	50%	resection, CH/RT	-	83%	*HRAS* mutations	-
Rhabdomyosarcoma, pleomorphic[1,33,34]	-	yes/NA/yes	72	3.5% of STS (all rhabdos)	C	54%	resection, CH/RT	-	26%	complex	-
Rhabdomyosarcoma, alveolar[1,35]	-	yes/NA/yes	10-24	25% of rhabdos	C	63%	resection, CH/RT	-	27%	*PAX3-FOXO1* or a *PAX7-FOXO1* fusion gene	-
Rhabdomyosarcoma, embryonal[1,35]	-	yes/NA/yes	2-20	0.45/100000	C	28%	resection, CH/RT	-	58%	complex	Costello syndrome, NF 1, Noonan syndrome, Li–Fraumeni syndrome
Rhabdomyosarcoma, spindle cell[1,36] WHO, 26823695	-	yes/NA/yes	34	3-10% of rhabdos	C	33%	resection, CH/RT	-	18%	*VGLL2/NCOA2/ CITED2* or *MYOD1* or *TFCP2/NCOA2* rearrangements	-

Vascular Tumors											
	Important differential diagnosis	**Infiltrating/ malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Angiosarcoma[1,37]	-	yes/no/yes	7th decade	2% of STS	C	20%	resection + RT/ CH/TT	radiation, lymph-edema, forieg bodies, AV fistulas, hemangiomas	30-40%	*MYC* gene amplifications	NF, Maffucci syndrome
Hemangioendothelioma, composite[1,38,39]	-	yes/no/rarely	43	26 cases	B	50%	resection	radiation, lymph-edema,	62-83%	*PTBP1-MAML2 and EPC1-PCH2 gene*	-
Hemangioendothelioma, epitheloid[1,40]	-	yes/no/yes	adulthood	0.1/100000	C	?	resection + CH/RT	-	59%	*WWTR1-CAMTA1 gene* fusion	-
Hemangioendothelioma, kaposiforme[1,41]	-	yes/no/rarely (lymph nodes)	1	0.9/100000 children	B	<5%	vincristine, steroid, sirolimus vs resection	-	-	*GNA14* mutations	-
Hemangioendothelioma, pseudomyogenic[1]	-	yes/no/rarely	30	?	A	60%	resection	-	-	*SERPINE1 to FOSB or ACTB-FOSB* fusion	-
Hemangioendothelioma, retiform[1]	-	no/no/rarely (lymph nodes)	childhood	40 cases	B or C	60%	resection	radiation, lymph-edema, lymph-angioma	-	-	-
Hemangioma, epitheloid[1,42]	-	rarely/no/rarely (lymph nodes)	4th decade	?	A or B	33%	resection	trauma	-	*FOS or FOSB gene*	-
Hemangioma[43,44]	-	no/no/no	51	2%	A or B	3-50%	if symptomatic: Embo + resection (+/- kypho, +/- adjuvant RT) vs Rt alone, vs	-	-	-	-
Kaposi sarcoma[1,45,46]	-	yes/no/yes	?	400-600/ 100000	-	?	immun-reconstitution, CH	immuno-suppression	74%	-	-
Lymphangioma[1]	-	no/no/no	congenital	?	A or B	20%	resection	-	-	*PIK3CA* mutations	Turner syndrome

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Appendix 3: Contd...

Peripheral Nerve Sheath Tumors

	Important differential diagnosis	Infiltrating/malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/ gene	Possible associated Tumor syndroms
Ectopic meningioma[1,47,48]	-	occasionally/no/ occasionally/6%	2nd + 5th decade	1% of menigiomas	A	26%	resection if high grade, symptomatic or pogressive	-	92% (3y)	-	Cowden, Li-Fraumeni, Von Hippel-Lindau syndrome
Hybrid nerve sheath tumors[1,49]	-	no/no/no	38	?	A	< 5%	resection if high grade, symptomatic or pogressive	-	-	-	NF1, NF2, schannomatosis
Malignant periperhal Nerve sheath tumor[1,50-52]	-	yes/NA/yes	20-50 years	2-5% of STS	C	56%	Resection + CH/TT	benign nerve sheath tumor, radiation	53%	complex	*NF1*
Neurofibroma[1,53,54]		rarely/in NF1/rarely/no	45	0.3/100000	A or B	17%	resection if symptomatic	-	-	inactivation *NF1* gene	*NF1*

Uncertain Differentiation

	Important differential diagnosis	Infiltrating/ malignant transformation/ metastasis	Peak age	Incidence	Type of surgical resection	Recurrence rate	Treatment	Risk factors	5y OS	Protein/gene	Possible associated Tumor syndroms
Clear cell sarcoma[1,55,56]	-	yes/no/yes	3-4th decade	?	C	40%	resection + RT/CH	-	60%	*reciprocal translocation t (12;22)(q13;q12)*	-
Desmoplastic small round cell tumor[1,57_61]	-	yes/no/yes	19	0.1/100000	C	89%	neo CH + resection + CH/RT vs TT	-	15%	*EWSR1-WT1 gene fusion*	-
Epitheloid sarcoma[1,62,63]	-	yes/no/yes	39	<1% STS	C	25%	resection + RT/CH	trauma	54%	*loss of SMARCB1 expression*	-
Extrarenal rhabdoid tumor[1,64,65]	-	yes/no/yes	13	<1% of childhood STS	C	22%	resection + CH/TT	-	15%	*SMARCB1 gene alterations*	-
Extraskeletal myxoid chondrosarcoma[1,66]	-	yes/no/yes	50	<1% STS	C	37%	resection + RT/TT	-	82-90%	*NR4A3 gene rearrangement*	-
Intramuscular myxoma[1]	-	yes/no/no	40-70 years	?	A	<5%	resection	fibrous dysplasia	-	*GNAS* mutation	-
Myoepithelioma[1,67]	-	possible/no/ possible	40 years	?	B	20-50%	resection	-	90%	*EWSR1 gene rearrangements*	-
NTRK-rearranged spindle cell neoplasm[1,68,69]	-	yes/no/no	1-2nd decade	1% of STS	B	11-44%	resection + CH/TT	-	?	*NTRK-rearrangements*	-
Ossifying fibromyxoid tumor[1,70]	-	yes/no/possible	58 years	?	B	0-60%	resection	-	94%	*PHF1 gene fusion*	-
Pecoma[1,71,72]	-	yes/no/yes	45	234 cases	C	0-70%	neo CH + resection + CH/RT vs TT	-	45%	*LOH TSC2* locus	-
Phosphaturic mesenchymal tumour[1,73]	-	no/yes/possible	53 years	< 0.01% of all STS	B	0-13%	resection	-	100%	*α-Klotho* upregulation	-
Synovial sarcoma[1,74,75]	-	yes/no/yes	3-4th decade	0.08/100000	C	42%	Resection + RT	radiotherapy	75-83%	*SS18-SSX1/2/4* fusion gene	-

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Undifferentiated Small Round Cell Tumors											
	Important differential diagnosis	**Infiltrating/malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Ewing Sarcoma[1,76-78]		yes/no/yes	16	0.3/100000	-	50%	chemotherapy	-	39-69%	FET-ETS fusion genes	-

Chondrogenic Tumors											
	Important differential diagnosis	**Infiltrating/ malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Chondroblastoma[1]	chondroblastoma-like osteosarcoma	no/no/benign lung mets	2-3 decade	<1% of bone tumors	A	10-18%	resection vs RFA	-	NA	*H3.3 alterations*	-
Chondromyxoid fibroma[1,79]	-	no/very rare/no	2-3rd decade	?	A or B	15%	resection	-	NA	*GRM1 gene recombination*	-
Chondrosarcoma, clear cell[1]	renal cell carcinoma, chondroblastoma, osteosarcoma	yes/rare/rare	3-4th decade	2% of chondrosarcomas	C	86%	resection	-	85%	*Chromosome 9, 20 aberrations*	-
Chondrosarcoma, mesenchymal[1,80]	-	yes/no/yes	26	2-9% of chondrosarcomas	C	55%	resection + CH	-	60%	*HEY1-NCOA2 rearrangement*	-
Chondrosarcoma, central grade II, III[1]	chondroblastic osteosarcoma	possible/yes/no	3-6th decade	0.18/100000	C	19-26%	resection	-	31-74%	*WNT/β-catenin signalling loss*	-
Chondrosarcoma, dedifferentiated[1,81]	-	yes/no/yes	59	11% of chondrosarcomas	C	50%	Resection + CH	-	7-24%	*IDH1 or IDH2 mutation*	-
Enchondroma[1]	secondary peripheral atypical cartilaginous tumour/chondrosarcoma	no/very rare/no/	36	2%	A	<5%	resection if symptomatic	-	NA	IDH1 or IDH2 mutations	Enchondromatosis
Osteochondroma[1]	secondary peripheral atypical cartilaginous tumour/chondrosarcoma	no/possible/no	18	0.9/100000	A	<5%	resection	radiation	NA	inactivation *EXT1* or *EXT2* gene	multiple osteochondromas syndrome
Osteochondromyxoma[1,82]	-	possible/no/ possible/no	1	?	A or B	?	resection	-	NA	*PRKAR1A* gene mutation	Carney complex
Secondary peripheral atypical cartilaginous tumor/ chondrosarcoma grade I[1,83]	-	yes/yes/yes/	49	0.66/100000	A or B	11%	resection vs RFA	-	87-99%	*IDH1* or *IDH2* mutation	Enchondromatosis
Secondary peripheral atypical cartilaginous tumor/ chondrosarcoma grade II, III[1]	periosteal osteosarcoma	yes/no/rarely	3-4th decade	5% of osteochondromas	B or C	16%	resection	-	98%	-	-
Synovial chondromatosis[1]r		yes/possible/ possible	3-5th decade	0.18/100000	B	20%	resection	-	NA	*FN1-ACVR2A* and *ACVR2A-FN1* fusions	-

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Osteogenic Tumors											
	Important differential diagnosis	**Infiltrating/ malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Osteoblastoma[1]	-	yes/rare/no	2-3rd decade	1% of bone tumors	B	23%	resection	-	NA	FOS rearrangements	-
Osteoid osteoma[1]	-	no/no/no	24	10% of all bone tumors	A	<5%	resection if symptomatic vs RFA (lesion might disappear)	-	NA	FOS rearrangements	-
Osteoma[1]	-	no/no/no	37	6.4%	A	<5%	resection if symptomatic	-	NA	LEMD3 gene	Gardner Syndrome, Osteopoikilosis
Osteosarcoma, (chondroblastic, fibroblastic, osteoblastic, telenagiectactic)[1,84]	-	yes/no/yes	10-14 years and 65 years	0.46/100000	C	30-50%	neoadjvuant CH + resection + RT/CH	-	68%	Gains 6p, 8q	LiFraumeni, Werner, Rothmund-Thomson, Bloom syndrome
Osteosarcoma, low grade central[1]	fibrosarcoma	yes/rare/rare	3rd decade	1-2% of osteosarcomas	B	7%	resection	-	90%	Amplification of 12q13-q15	-
Osteosarcoma, secondary	-	yes/no/yes	6-7th decade	1-7% in Paget disease	C	?	neoadjuvant CH + resection + RT/CH	Paget diaseas, radiation, Caisson disease, Sickle cell disease, implants, chronic osteomyleitis	10-32%	?	Rothmund-Thomson syndrome

Osteoclastic giant cell-rich Tumors											
	Important differential diagnosis	**Infiltrating/malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Aneurysmal bone cyst[1]	teleangiectactic osteosarcoma	no/no/no	1-2nd decade	0.015/100000	A or B	20-70%	resection vs denosumab vs embo vs RT	-	NA	*USP6* rearrangements	-
Giant cell tumor[1,85,86]	-	yes/rarely/rarely	31	0.15/100000	B	15-50%	resection vs denosumab vs embo vs RT	Paget disease, radiation	87%****	H3.3 mutation	Gorlin-Goltz syndrome, Jaffe-Campanacci syndrome
Non-ossifying fibroma[1]		no/no/no	2nd decade	?	A	<5%	resection if symptomatic	-	NA	*KRAS* and *FGFR1* mutations	Jaffe-Campanacci syndrome, *NF1, KRAS*

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Notochordal Tumors											
	Important differential diagnosis	**Infiltrating/malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/ gene**	**Possible associated Tumor syndroms**
Benign notochordal tumor[1,87]	-	no/rarely/no	58	1.7%	A	<5%	resection if symptomatic	-	NA	expression of brachyury	-
Chordoma, conventional, dedifferentiated, poorly differentiated[1]	-	yes/yes/yes	6-8th decade	0.08/100000	C	35%	resection + RT/TT	-	68%	expression of brachyury	-
Haematopoietic Neoplasms of Bone											
	Important differential diagnosis	**Infiltrating/malignant transformation/ metastasis**	**Peak age**	**Incidence**	**Type of surgical resection**	**Recurrence rate**	**Treatment**	**Risk factors**	**5y OS**	**Protein/gene**	**Possible associated Tumor syndroms**
Plasmacytoma[1] WHO	-	yes/yes/yes	55-60	6.8/100000	-	22%	RT	-	57%	-	-
Non-Hodgkin lymphoma of the bone[1,88]	-	yes/yes/yes	50-60	7% of bone tumors	-	10%	CH, RT	HIV	75%	Immunglobulin rearrangements	-

*In high risk/systemic/recurrence patients, **Depending on mutation status: CTNNB1 p.Ser45Phe, ***20 year survival rate in patients with FAP associated lesions, ****Patients with malignant variant, *****Mean survival time in aggressive variant (EIMS), ******At 46 month, *******Very rare malignant variant, „ congenital variants less aggressive, , among immunospurressed, {{{ can arise secondarily in previous enchondroma, {{{{ can arise secondarily on the surface of osteochondromas. CH: Chemotherapy; TT: Targeted therapy

REFERENCES

1. WHO Classification of Tumours Editorial Board. Soft Tissue and Bone Tumours: WHO Classification of Tumours (Medicine). 5th ed. Lyon: World Health Organization; 2020.
2. Mavrogenis AF, Lesensky J, Romagnoli C, Alberghini M, Letson GD, Ruggieri P. Atypical lipomatous tumors/well-differentiated liposarcomas: Clinical outcome of 67 patients. Orthopedics 2011;34:e893-8.
3. Furlong MA, Fanburg-Smith JC, Miettinen M. The morphologic spectrum of hibernoma: A clinicopathologic study of 170 cases. Am J Surg Pathol 2001;25:809-14.
4. Abdul-Ghafar J, Ahmad Z, Tariq MU, Kayani N, Uddin N. Lipoblastoma: A clinicopathologic review of 23 cases from a major tertiary care center plus detailed review of literature. BMC Res Notes 2018;11:42.
5. Teekhasaenee C, Kita K, Takegami K, Kawakita E, Sakakibara T, Kasai Y. Intraosseous lipoma of the third lumbar spine: A case report. J Med Case Rep 2015;9:52.
6. Rajput D, Srivastava AK, Kumar R. Spinal epidural lipomatosis: An unusual cause of relapsing and remitting paraparesis. J Pediatr Neurosci 2010;5:150-2.
7. Kim K, Mendelis J, Cho W. Spinal epidural lipomatosis: A review of pathogenesis, characteristics, clinical presentation, and management. Global Spine J 2019;9:658-65.
8. Ferlic PW, Mannion AF, Jeszenszky D, Porchet F, Fülöp Fekete T, Kleinstück F, *et al*. Patient-reported outcome of surgical treatment for lumbar spinal epidural lipomatosis. Spine J 2016;16:1333-41.
9. Malone JB, Bevan PJ, Lewis TJ, Nelson AD, Blaty DE, Kahan ME. Incidence of spinal epidural lipomatosis in patients with spinal stenosis. J Orthop 2018;15:36-9.
10. Chowdhry V, Goldberg S, DeLaney TF, Cote GM, Chebib I, Kim J, *et al*. Myxoid liposarcoma: Treatment outcomes from chemotherapy and radiation therapy. Sarcoma 2018;2018:8029157.
11. Gebhard S, Coindre JM, Michels JJ, Terrier P, Bertrand G, Trassard M, *et al*. Pleomorphic liposarcoma: Clinicopathologic, immunohistochemical, and follow-up analysis of 63 cases: A study from the French Federation of Cancer Centers Sarcoma Group. Am J Surg Pathol 2002;26:601-16.
12. Gupta G, Garg R, Wadhwa C, Bansal H, Kaushal RK. A rare primary dumbbell lipoblastoma. Asian J Neurosurg 2018;13:83-5.
13. Desurmont T, Lefevre JH, Shields C, Colas C, Tiret E, Parc Y. Desmoid tumour in familial adenomatous polyposis patients: responses to treatments. Fam Cancer 2015;14:31-9.
14. Bahrami A, Folpe AL. Adult-type fibrosarcoma: A reevaluation of 163 putative cases diagnosed at a single institution over a 48-year period. Am J Surg Pathol 2010;34:1504-13.
15. Augsburger D, Nelson PJ, Kalinski T, Udelnow A, Knösel T, Hofstetter M, *et al*. Current diagnostics and treatment of fibrosarcoma -perspectives for future therapeutic targets and strategies. Oncotarget 2017;8:104638-53.
16. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol 1995;19:859-72.
17. Yu L, Liu J, Lao IW, Luo Z, Wang J. Epithelioid inflammatory myofibroblastic sarcoma: A clinicopathological, immunohistochemical and molecular cytogenetic analysis of five additional cases and review of the literature. Diagn Pathol 2016;11:67.
18. Panagiotopoulos N, Patrini D, Gvinianidze L, Woo WL, Borg E, Lawrence D. Inflammatory myofibroblastic tumour of the lung: A reactive lesion or a true neoplasm? J Thorac Dis 2015;7:908-11.
19. Fetsch JF, Miettinen M, Laskin WB, Michal M, Enzinger FM. A clinicopathologic study of 45 pediatric soft tissue tumors with an admixture of adipose tissue and fibroblastic elements, and a proposal for classification as lipofibromatosis. Am J Surg Pathol 2000;24:1491-500.
20. Chamberlain F, Engelmann B, Al-Muderis O, Messiou C, Thway K, Miah A, *et al*. Low-grade fibromyxoid sarcoma: Treatment outcomes and efficacy of chemotherapy. *In Vivo* 2020;34:239-45.
21. Yalcin ED, Ararat E. Prevalence of soft tissue calcifications in the head and neck region: A cone-beam computed tomography study. Niger J Clin Pract 2020;23:759-63.
22. Kubota K, Okada S, Maeda T, Matsumoto Y, Sakamoto A, Harimaya K, *et al*. Extradural nodular fasciitis arising in the spinal canal. Spine (Phila Pa 1976) 2012;37:E133-7.
23. Antonescu CR, Rosenblum MK, Pereira P, Nascimento AG, Woodruff JM. Sclerosing epithelioid fibrosarcoma: A study of 16 cases and confirmation of a clinicopathologically distinct tumor. Am J Surg Pathol 2001;25:699-709.
24. Ossendorf C, Studer GM, Bode B, Fuchs B. Sclerosing epithelioid fibrosarcoma: Case presentation and a systematic review. Clin Orthop Relat Res 2008;466:1485-91.
25. Demicco EG, Park MS, Araujo DM, Fox PS, Bassett RL, Pollock RE, *et al*. Solitary fibrous tumor: A clinicopathological study of 110 cases and proposed risk assessment model. Mod Pathol 2012;25:1298-306.
26. Outani H, Kobayashi E, Wasa J, Saito M, Takenaka S, Hayakawa K, *et al*. Clinical outcomes of patients with metastatic solitary fibrous tumors: A Japanese Musculoskeletal Oncology Group (JMOG) Multiinstitutional Study. Ann Surg Oncol 2021;28:3893-901.
27. Ardissone F. Thoracic malignant solitary fibrous tumors: Prognostic factors and long-term survival. J Thorac Dis 2011;3:84-5.
28. Ehresman JS, Ahmed AK, Palsgrove DN, Pennington Z, Goodwin CR, Sciubba DM. Epstein-Barr virus-associated smooth muscle tumor involving the spine of an HIV-infected patient: Case report and review of the literature. J Clin Neurosci 2018;52:145-50.
29. Kilpatrick SE, Mentzel T, Fletcher CD. Leiomyoma of deep soft tissue. Clinicopathologic analysis of a series. Am J Surg Pathol 1994;18:576-82.
30. Gladdy RA, Qin LX, Moraco N, Agaram NP, Brennan MF, Singer S. Predictors of survival and recurrence in primary leiomyosarcoma. Ann Surg Oncol 2013;20:1851-7.
31. Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: Clinicopathologic and immunohistochemical study of 54 cases. Am J Surg Pathol 2006;30:104-13.
32. Dantonello TM, Leuschner I, Vokuhl C, Gfroerer S, Schuck A, Kube S, *et al*. Malignant ectomesenchymoma in children and adolescents: Report from the Cooperative Weichteilsarkom Studiengruppe (CWS). Pediatr Blood Cancer 2013;60:224-9.
33. Esnaola NF, Rubin BP, Baldini EH, Vasudevan N, Demetri GD, Fletcher CD, *et al*. Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. Ann Surg 2001;234:215-23.
34. Noujaim J, Thway K, Jones RL, Miah A, Khabra K, Langer R, *et al*. Adult pleomorphic rhabdomyosarcoma: A multicentre retrospective study. Anticancer Res 2015;35:6213-7.
35. Drabbe C, Benson C, Younger E, Zaidi S, Jones RL, Judson I, *et al*. Embryonal and alveolar rhabdomyosarcoma in adults: Real-life data from a tertiary sarcoma centre. Clin Oncol (R Coll Radiol) 2020;32:e27-35.
36. Zhao Z, Yin Y, Zhang J, Qi J, Zhang D, Ma Y, *et al*. Spindle cell/sclerosing rhabdomyosarcoma: Case series from a single institution emphasizing morphology, immunohistochemistry and follow-up. Int J Clin Exp Pathol 2015;8:13814-20.
37. Cao J, Wang J, He C, Fang M. Angiosarcoma: A review of diagnosis and current treatment. Am J Cancer Res 2019;9:2303-13.
38. Nayler SJ, Rubin BP, Calonje E, Chan JK, Fletcher CD. Composite hemangioendothelioma: A complex, low-grade vascular lesion mimicking angiosarcoma. Am J Surg Pathol 2000;24:352-61.
39. McNab PM, Quigley BC, Glass LF, Jukic DM. Composite hemangioendothelioma and its classification as a low-grade malignancy. Am J Dermatopathol 2013;35:517-22.
40. McCulloch M, Russin M, Nachat A. Recurrence of epithelioid

hemangioendothelioma during pregnancy: Case report and systematic review. Perm J 2016;20:15-152.
41. Ji Y, Chen S, Yang K, Xia C, Li L. Kaposiform hemangioendothelioma: Current knowledge and future perspectives. Orphanet J Rare Dis 2020;15:39.
42. Fetsch JF, Weiss SW. Observations concerning the pathogenesis of epithelioid hemangioma (angiolymphoid hyperplasia). Mod Pathol 1991;4:449-55.
43. Santos RH, Franca CN, Guimaraes VD, Prigenzi CK, Fontoura EA, Botelho RV. Aggressive vertebral hemangiomas in asymptomatic patients: A review. Surg Neurol Int 2020;11:453.
44. Goldstein CL, Varga PP, Gokaslan ZL, Boriani S, Luzzati A, Rhines L, *et al*. Spinal hemangiomas: Results of surgical management for local recurrence and mortality in a multicenter study. Spine (Phila Pa 1976) 2015;40:656-64.
45. Liu Z, Fang Q, Zuo J, Minhas V, Wood C, Zhang T. The world-wide incidence of Kaposi's sarcoma in the HIV/AIDS era. HIV Med 2018;19:355-64.
46. Schneider JW, Dittmer DP. Diagnosis and treatment of Kaposi sarcoma. Am J Clin Dermatol 2017;18:529-39.
47. Lang FF, Macdonald OK, Fuller GN, DeMonte F. Primary extradural meningiomas: A report on nine cases and review of the literature from the era of computerized tomography scanning. J Neurosurg 2000;93:940-50.
48. Liu Y, Wang H, Shao H, Wang C. Primary extradural meningiomas in head: A report of 19 cases and review of literature. Int J Clin Exp Pathol 2015;8:5624-32.
49. Hornick JL, Bundock EA, Fletcher CD. Hybrid schwannoma/perineurioma: Clinicopathologic analysis of 42 distinctive benign nerve sheath tumors. Am J Surg Pathol 2009;33:1554-61.
50. Le Guellec S, Decouvelaere AV, Filleron T, Valo I, Charon-Barra C, Robin YM, *et al*. Malignant peripheral nerve sheath tumor is a challenging diagnosis: A systematic pathology review, immunohistochemistry, and molecular analysis in 160 patients from the French Sarcoma Group Database. Am J Surg Pathol 2016;40:896-908.
51. Watson KL, Al Sannaa GA, Kivlin CM, Ingram DR, Landers SM, Roland CL, *et al*. Patterns of recurrence and survival in sporadic, neurofibromatosis type 1-associated, and radiation-associated malignant peripheral nerve sheath tumors. J Neurosurg 2017;126:319-29.
52. Lekovic GP, Mehta GU, Maxwell AK, Peng KA, Brackmann DE. Radiation-induced malignant peripheral nerve sheath tumor of the vagus nerve following radiation treatment of cervical paraganglioma. J Neurol Surg Rep 2020;81:e66-70.
53. Xin Z, Orazmyradov B, Li J, Zhou Q, Peng L, Li H, *et al*. A novel classification and its clinical significance in spinal schwannoma based on the membranous hierarchy. Neurosurgery 2020;87:1206-22.
54. Safaee M, Parsa AT, Barbaro NM, Chou D, Mummaneni PV, Weinstein PR, *et al*. Association of tumor location, extent of resection, and neurofibromatosis status with clinical outcomes for 221 spinal nerve sheath tumors. Neurosurg Focus 2015;39:E5.
55. Eckardt JJ, Pritchard DJ, Soule EH. Clear cell sarcoma. A clinicopathologic study of 27 cases. Cancer 1983;52:1482-8.
56. Friesenbichler W, Luftinger R, Kropshofer G, Henkel M, Amann G, Furtwängler R, *et al*. Clear cell sarcoma of the kidney in Austrian children: Long-term survival after relapse. Pediatr Blood Cancer 2021;68:e28860.
57. Gerald WL, Miller HK, Battifora H, Miettinen M, Silva EG, Rosai J. Intra-abdominal desmoplastic small round-cell tumor. Report of 19 cases of a distinctive type of high-grade polyphenotypic malignancy affecting young individuals. Am J Surg Pathol 1991;15:499-513.
58. Kallianpur AA, Shukla NK, Deo SV, Yadav P, Mudaly D, Yadav R, *et al*. Updates on the multimodality management of desmoplastic small round cell tumor. J Surg Oncol 2012;105:617-21.
59. Honore C, Delhorme JB, Nassif E, Faron M, Ferron G, Bompas E, *et al*. Can we cure patients with abdominal desmoplastic small round cell tumor? Results of a retrospective multicentric study on 100 patients. Surg Oncol 2019;29:107-12.
60. Mello CA, Campos FA, Santos TG, Gobo Silva ML, Tardin Torrezan G, D'Almeida Costa F, *et al*. Desmoplastic small round cell tumor: A review of main molecular abnormalities and emerging therapy. Cancers (Basel) 2021;13:498.
61. Stiles ZE, Murphy AJ, Anghelescu DL, Brown CL, Davidoff AM, Dickson PV, *et al*. Desmoplastic small round cell tumor: Long-term complications after cytoreduction and hyperthermic intraperitoneal chemotherapy. Ann Surg Oncol 2020;27:171-8.
62. Kim SJ, Ha DH, Lee SM, Kang H. Desmoid type fibromatosis in the facet joint of lumbar spine: Case report and review of literature. Korean J Radiol 2013;14:818-22.
63. Pradhan A, Grimer RJ, Abudu A, Tillman RM, Carter SR, Jeys L, *et al*. Epithelioid sarcomas: How important is loco-regional control? Eur J Surg Oncol 2017;43:1746-52.
64. Fanburg-Smith JC, Hengge M, Hengge UR, Smith JS Jr., Miettinen M. Extrarenal rhabdoid tumors of soft tissue: A clinicopathologic and immunohistochemical study of 18 cases. Ann Diagn Pathol 1998;2:351-62.
65. Geller JI, Roth JJ, Biegel JA. Biology and treatment of rhabdoid tumor. Crit Rev Oncog 2015;20:199-216.
66. Drilon AD, Popat S, Bhuchar G, D'Adamo DR, Louise Keohan M, Fisher C, *et al*. Extraskeletal myxoid chondrosarcoma: A retrospective review from 2 referral centers emphasizing long-term outcomes with surgery and chemotherapy. Cancer 2008;113:3364-71.
67. Hornick JL, Fletcher CD. Myoepithelial tumors of soft tissue: A clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. Am J Surg Pathol 2003;27:1183-96.
68. Brcic I, Godschachner TM, Bergovec M, Igrec J, Till H, Lackner H, *et al*. Broadening the spectrum of NTRK rearranged mesenchymal tumors and usefulness of pan-TRK immunohistochemistry for identification of NTRK fusions. Mod Pathol 2021;34:396-407.
69. Davis JL, Lockwood CM, Stohr B, Boecking C, Al-Ibraheemi A, DuBois SG, *et al*. Expanding the spectrum of pediatric NTRK-rearranged mesenchymal tumors. Am J Surg Pathol 2019;43:435-45.
70. Folpe AL, Weiss SW. Ossifying fibromyxoid tumor of soft parts: A clinicopathologic study of 70 cases with emphasis on atypical and malignant variants. Am J Surg Pathol 2003;27:421-31.
71. Bleeker JS, Quevedo JF, Folpe AL. "Malignant" perivascular epithelioid cell neoplasm: Risk stratification and treatment strategies. Sarcoma 2012;2012:541626.
72. Sanfilippo R, Jones RL, Blay JY, Le Cesne A, Provenzano S, Antoniou G, *et al*. Role of chemotherapy, VEGFR inhibitors, and mTOR inhibitors in advanced perivascular epithelioid cell tumors (PEComas). Clin Cancer Res 2019;25:5295-300.
73. Folpe AL, Fanburg-Smith JC, Billings SD, Bisceglia M, Bertoni F, Cho JY, *et al*. Most osteomalacia-associated mesenchymal tumors are a single histopathologic entity: An analysis of 32 cases and a comprehensive review of the literature. Am J Surg Pathol 2004;28:1-30.
74. Scheer M, Dantonello T, Hallmen E, Blank B, Sparber-Sauer M, Vokuhl C, *et al*. Synovial sarcoma recurrence in children and young adults. Ann Surg Oncol 2016;23 Suppl 5:618-26.
75. Andrassy RJ, Okcu MF, Despa S, Raney RB. Synovial sarcoma in children: Surgical lessons from a single institution and review of the literature. J Am Coll Surg 2001;192:305-13.
76. Esiashvili N, Goodman M, Marcus RB Jr. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance epidemiology and end results data. J Pediatr Hematol Oncol 2008;30:425-30.
77. Albergo JI, Gaston CL, Laitinen M, Darbyshire A, Jeys LM, Sumathi V, *et al*. Ewing's sarcoma: Only patients with 100% of necrosis after chemotherapy should be classified as having a good response. Bone Joint J 2016;98-B: 1138-44.

78. Van Mater D, Wagner L. Management of recurrent Ewing sarcoma: Challenges and approaches. Onco Targets Ther 2019;12:2279-88.
79. Bhamra JS, Al-Khateeb H, Dhinsa BS, Gikas PD, Tirabosco R, Pollock R, *et al*. Chondromyxoid fibroma management: A single institution experience of 22 cases. World J Surg Oncol 2014;12:283.
80. Xu J, Li D, Xie L, Tang S, Guo W. Mesenchymal chondrosarcoma of bone and soft tissue: A systematic review of 107 patients in the past 20 years. PLoS One 2015;10:e0122216.
81. Gomez CD, Anderson MS, Epperly SC, Zuckerman LM. Successful treatment of a dedifferentiated chondrosarcoma of the proximal humerus with a hemicortical articular surface sparing allograft: A case report. Int J Surg Case Rep 2020;72:590-5.
82. Carney JA, Boccon-Gibod L, Jarka DE, Tanaka Y, Swee RG, Unni KK, *et al*. Osteochondromyxoma of bone: A congenital tumor associated with lentigines and other unusual disorders. Am J Surg Pathol 2001;25:164-76.
83. Deckers C, Schreuder BH, Hannink G, de Rooy JW, van der Geest IC. Radiologic follow-up of untreated enchondroma and atypical cartilaginous tumors in the long bones. J Surg Oncol 2016;114:987-91.
84. Sun HH, Chen XY, Cui JQ, Zhou ZM, Guo KJ. Prognostic factors to survival of patients with chondroblastic osteosarcoma. Medicine (Baltimore) 2018;97:e12636.
85. Martin C, McCarthy EF. Giant cell tumor of the sacrum and spine: Series of 23 cases and a review of the literature. Iowa Orthop J 2010;30:69-75.
86. Palmerini E, Picci P, Reichardt P, Downey G. Malignancy in giant cell tumor of bone: A review of the literature. Technol Cancer Res Treat 2019;18:1533033819840000.
87. Deshpande V, Nielsen GP, Rosenthal DI, Rosenberg AE. Intraosseous benign notochord cell tumors (BNCT): Further evidence supporting a relationship to chordoma. Am J Surg Pathol 2007;31:1573-7.
88. Heyning FH, Hogendoorn PC, Kramer MH, Holland CT, Dreef E, Jansen PM. Primary lymphoma of bone: Extranodal lymphoma with favourable survival independent of germinal centre, post-germinal centre or indeterminate phenotype. J Clin Pathol 2009;62:820-4.