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Original Article

Primary bone tumors of the spine revisited: A 10-year single-center experience of the management and outcome in a neurosurgical department

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Abstract

Objective: To report a large clinical series of primary bone tumors of the spine (PBTS) and review the current concepts of management. **Materials and Methods:** We retrospectively analyzed a clinical series of PBTS treated over the last decade (2004-2014) in the spine unit of a large European tertiary care center. Every PBTS was identified from an electronic medical-record system. Analysis comprised medical records and clinical imaging. Overall survival and outcome was measured using the Glasgow Outcome Scale (GOS) at six weeks, six months and one year postoperatively. Surgical management and adjuvant/neoadjuvant strategies were analyzed. A thorough review of the current literature was performed. **Results:** A total of 79 patients were included. Of these, 44 (55.7%) were male. The age ranged from 9 to 90 years (mean 55), and most patients were adults (93.6%). Local pain was the most common symptom and was present in 91.1% of the patients. The majority of the tumors occurred in the thoracic spine (52 patients, 65.8%). Overall 86% (68 patients) of PBTS were classified as malignant and at the time of diagnosis, 7 patients (8.9%) presented with non-spinal metastasis. The most common histologic types were hematopoietic tumors (72.2%), followed by chondrogenic ones (12.7%). Within hematopoietic tumors, plasmacytoma was the most frequent type (49 patients, 62%). In 12 patients (15.2%) recurrences were seen during the follow-up period. Overall mean survival of benign PBTS was 100%, malignant non-hematopoietic PBTS 50% and, malignant hematopoietic PBTS 84% at one year, respectively. At six weeks and one year after the initial surgery, 79% and 54% of the patients presented a GOS >3, respectively. **Conclusion:** PBTS were almost exclusively seen in adults. Malignant tumors were markedly more frequent than benign tumors, with hematopoietic tumors being the most common type. For PBTS, early surgery is important in order to restore spinal stability and decompress the spinal cord. This allows pain reduction and prevention of neurological deficits.

Key words: Management, plasmacytoma, primary bone tumor, spine, WHO

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INTRODUCTION

Primary bone tumors (PBT) represent less than 0.2% of all newly diagnosed tumors and half of these results in death.[1] Only 5% of all PBT arise in the spine.[2-4] In most series published, hematopoietic tumors are the most common malignant histological type.[5] In adults, almost 80% of the PBT are malignant, as opposed to children in which 40% are classified as malignant.[4]

Epidemiological data shows, that in many bone tumors with benign radiological features, which occur with greater frequency than tumors with malignant features, no tissue diagnosis is pursued, and consequently the precise incidence of a specific bone tumor is not known.[2,4,6-8] Although primary bone tumors of the spine (PBTS) are rare, they harbor significant cancer morbidity and mortality, especially among young people. This impact is primarily caused by the characteristic local invasiveness, destruction of adjacent structures and neurological impairment, as well as metastases.[7,8]

The rarity of this pathology explains the lack of high-level evidence, mainly based on case series.[4,8-10]

The World Health Organization (WHO) classifies hematopoietic type tumors (e.g., plasma cell myeloma, solitary or osseous plasmacytoma, and lymphoma of the bone) as PBT.[11] However, they show different features compared to other PBTS, e.g., early systemic spreading and favorable responsiveness to chemo- and/or radiotherapy.[5]

The purpose of this study was to evaluate the management and outcome of PBTS in a single neurosurgical department over the last decade (2004-2014), providing homogeneity in diagnosis and treatment.

A secondary objective was to separately analyze the hematopoietic tumors, considering them as a distinct group, and to critically question the current WHO Classification for PBT.

MATERIALS AND METHODS

We retrospectively analyzed consecutive series of patients with histologically confirmed PBTS treated from 2004 to 2014 in the neurosurgical department of a large European tertiary care center. All patients were identified using the International Classification of Diseases (ICD) discharge codes. Patients with a newly diagnosed PBTS at our institution were included. Regarding the hematopoietic type, we only included osseous plasmacytoma and lymphoma of the bone, in which the first manifestation of the tumor was located in the spine. Spinal metastases and recurrences were excluded.

The clinical characteristics, tumor type, localization, treatment and outcome were collected and analyzed. The incidence of each tumor type was determined. To determine if hematopoietic tumors represent a distinct group, we stratified the population into benign, malignant hematopoietic and, malignant non-hematopoietic. For these subgroups clinical outcome was evaluated using the Glasgow Outcome Scale (GOS) at the first follow-up visit at six weeks, six months and one year postoperatively. Overall survival was established by Kaplan-Meier estimates at the same intervals. The local research ethics board approved the study.

RESULTS

Population and clinical presentation

Between 2004 and 2014, a total of 79 patients were newly diagnosed with PBTS and treated at our department. There were 44 (55.7%) male and 35 (44.3%) female patients. The mean age at diagnosis was 55 years (range 9-90 years) [Table 1].

Local pain was the most common symptom (91.1%), followed by gait disturbances (51.9%). Table 2 summarizes the clinical symptoms.

The duration of symptoms was less than three months in 67 patients (84.8%), and only 3 patients (3.9%) had symptoms for more than a year.

Diagnosis

The most utilized diagnostic tool was CT (89.9%), followed by MRI (84.8%). In 69.6% of the cases, both CT and MRI were performed. Although X-ray is no longer a standard of care diagnostic in spine tumors, it was used in 4 patients (5.1%) as a complimentary diagnostic tool. A preoperative biopsy was only performed in 3 patients (3.8%). A bone scintigraphy was performed preoperatively in 2 patients (2.5%) and as complimentary diagnostic tool after surgery in 32 patients (40.5%).

Table 3 summarizes the tumor distribution along the spine.

At the time of diagnosis, most of the patients (84.7%) had 1-3 lesions. One patient (1.3%) presented 8 lesions in different spinal locations and 7 patients (8.9%) already had distant, non-spinal metastases [Table 4].

The mean axial diameter of the lesions measured on CT and/or MRI was 40 mm (SD 16 mm-range 9 to 81 mm).

Table 1: Distribution according to age and sex

Decade	No.	Sex (M/F)	Age %	Sex (M/F) %
0-10	3	3/0	3.8	3.8/0
11-20	4	1/3	5	1.3/3.7
21-30	5	3/2	6.3	3.8/2.5
31-40	3	1/2	3.8	1.3/2.5
41-50	13	8/5	16.5	10.1/6.4
51-60	12	8/4	15.2	10.1/5.1
61-70	27	18/9	34.2	22.8/11.4
71-80	6	0/6	7.6	0/7.6
81-90	6	2/4	7.6	2.5/5.1
	79	44/35	100	55.7/44.3

Table 2: Symptoms

Symptom	Preoperative		Postoperative		Improvement	
	No.	%	No.	%	No.	%
Local Pain	72	91.1	31	39.2	41	56.9
Gait Disturbances	41	51.9	26	32.9	15	36.6
Paresthesia	24	44.6	14	17.7	10	41.2
Paresia	22	27.8	13	16.4	9	41
Plegia	8	10.1	1	1.2	7	87.5
Swelling	2	2.5	0	0	2	100
Dysphagia	2	2.5	0	0	2	100
Other	2	2.5	3	3.8	0	0

Table 3: Localization of lesions

Localization	No.	%
Cervical	23	29.1
Thoracic	52	65.8
Lumbar	18	22.8
Sacral	5	6.3

Table 4: Number of lesions and/or simultaneous lesions of the spine per patient

No. Lesions	No.	%
1	28	35.4
2	22	27.8
3	17	21.5
4	8	10.1
6	2	2.5
7	1	1.3
8	1	1.3

Treatment

A total of 77 (97%) patients underwent an invasive procedure. The remaining 2 patients did not wish to proceed with surgical therapy following the tissue diagnosis after biopsy. Complete tumor resection was radiologically achieved in 43 cases (55.8%). In 18 cases (23.4%), a partial resection of the tumor or thecal sac decompression was performed. In 16 patients (20.8%), a biopsy and vertebroplasty were performed, in order to reduce pain and restore stability. A dorsal approach was used in 64 patients (83.1%), a ventral approach in 10 cases (12.9%), and a combined dorso-ventral (360°) approach was required in 3 cases (4%) [Figure 1].

Complications were seen in 25.9% (20 cases) requiring a second intervention. Complications in our study included misplacement of osteosynthesis material in dorsal instrumentations without neurological deficits and one postoperative non-tumor associated hemorrhage. All complications were corrected by second look surgery and did not result in additional morbidity.

Clinical outcome

Postoperatively, significant symptom reduction was achieved. Pain improved in 57% and gait disturbances in 37%. Sensory deficits improved in 42% and motor deficits (paresis/plegia) in 53%. Remarkably, 7 of 8 patients with pre-operative paraplegia recovered (87.5%), whilst only 1 patient (1.3%) remained paraplegic [Table 2].

Functional outcome, recurrence and survival

The majority of patients (72 (93.5%)) were evaluated at 6-weeks follow-up, where 23 patients (31.9%) showed low disability according to the GOS, 38 (52.7%) showed moderate disability and 11 (15.2%) had severe disability. Six patients were lost to follow-up and one patient died from cardiac disease one month following the surgery. Six months after the surgery, 58 patients (75%) were evaluated. At this time, 10 patients had been lost to follow-up and 11 had died. At one year after surgery, 51 patients (66.2%) remained in the control group, the residual patients were either lost to follow-up or had died [Table 5 and Figure 2]. At two years following the surgery, only 31 patients (40%) were available for follow-up. Kaplan-Meier analysis was performed to assess the general overall survival rate at six weeks, six months, one year and two years [Figure 3].

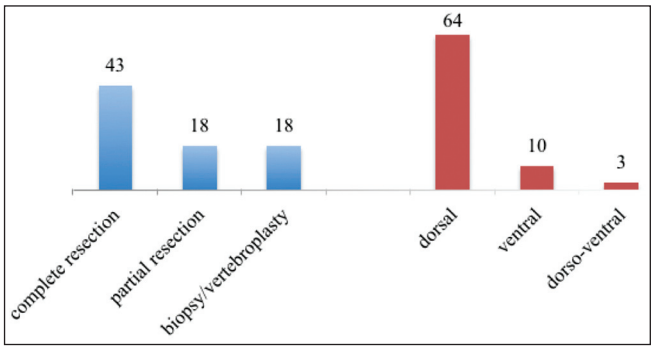

Figure 1: Treatment type and sub-type utilized

In 12 patients (15.6%), tumor recurrence occurred during the follow-up period. A total of 8 (10.1%) patients died within 3 months following surgery or had no follow-up controls. The cause of death was in all cases unrelated to the surgery performed.

Histology

Malignant PBTS was seen in 68 patients (86%), the most common histological presentations being hematopoietic (72.2%) and chondrogenic (12.7%), followed by neuroectodermal (5.1%), osteogenic (3.8%), notochordal (1.3%) and other types (6.3%) [Table 6]. The most common PBTS diagnosed was plasmacytoma in 49 patients (62%), followed by chondrosarcoma in 5 (6.3%), B-cell lymphoma in 5 (6.3%), and Ewing sarcoma in 4 patients (5%). Hemangioma, T-cell lymphoma, osteochondroma and osteoblastoma were found in 2 patients (2.5%) each; chondroblastoma, chordoma, enchondroma, Langerhans histiocytosis, leiomyosarcoma, mixed B-cell and T-cell lymphoma, osteosarcoma and Giant-Cell tumor were diagnosed in 1 patient (1.3%) each.

DISCUSSION

Etiology

The majority of PBT arises within the first three decades of life, with spine involvement in only 5% of cases.[2,3,6] Although there are no known predisposing or risk factors, some subtypes such as bone infarctions, chronic osteomyelitis, Paget's disease, radiation and metal prostheses have been associated with bone tumors.[6,12] Nevertheless, the etiology of PBT remains unknown. Recent molecular studies suggest mutations in the tumor suppressor gene p53, the nuclear factor kB ligand (RANKL) and osteoprotegerin (OPG).[13] These studies may reveal new targets for alternative or coadjuvant treatment options.[14-17]

Additionally, certain syndromes have been associated to some PBT [Tables 7 and 8]. In our study, none of the patients had an associated syndrome.

Epidemiology and presentation

The incidence of benign bone tumors is higher than malignant ones, and their incidence is still underestimated, as most remain asymptomatic and therefore undetected. Moreover, benign tumors are seldom subject to biopsy or surgery, and as a consequence, the real incidence is impossible to establish.[8,12] Furthermore, referral bias may play a role, especially in surgical studies.

Our series comprises a high percentage of malignant tumors (86%) where a referral bias is the most likely explanation. Interestingly, all patients in our study were first referred to the neurosurgical department. Most of them were diagnosed and treated within 3 months and, if necessary, transferred then to the hemato-oncology or radiation oncology unit for adjuvant treatment.

Our extensive literature review yielded only a few large series analyzing PBTS [Table 9]. In our study, the distribution of patients according to gender and age did not differ from the literature, in fact the male to female ratio is about 1:1. However, the mean age in our population was higher than in the Leeds Registry and the PTS Database (55 vs. 42, and 43 years, respectively). The high rate of malignant PBTS in our study (86%) was comparable to the Leeds Registry (77%), but significantly higher as compared to the Instituto Rizzoli series (43%) and the PTS Database (49%).[4,9,10] Also similar to the Leeds Registry, plasmacytoma (62%) was by far the most common tumor type. The second most common type was also of hematopoietic origin (lymphoma, 6.5%) equal to chondrosarcoma (6.5%). Other authors reported chordoma and osteosarcoma as the second and third most frequent types.[4] Local and ethnic characteristics, as well as a referral practice, may play an important role in these rare pathologies.[4]

In our series, the patients with PBT almost always presented an acute, semi-acute or chronic localized pain (91%), typically exacerbated by movement or at night hours. In contrast, a palpable mass was exceptional (2.5%). Clinical signs such as gait disturbances as well as paresis and/or paresthesias were the most common neurological findings [Table 2]. In most cases of acute and severe onset, diagnosis and treatment was conducted in less than three days.

Diagnosis

When the suspicion of PBTS arises, spinal CT or MRI are the standard diagnostic tools. They provide a precise assessment of the lesion, anatomic origin, extension, and allow differential diagnosis.[8,18-20] A multidisciplinary approach, including surgeons, pathologists, oncologists and radiologists allows the best local and systemic management.[8,12,18-22]

Table 5: Glasgow outcome scale at 6 weeks, 6 months and 1 year

GOS	6 Weeks	6 Months	1 Year
1	1	10	2
2	0	0	0
3	11	8	9
4	38	30	23
5	23	20	19
%	94.8	88.3	68.8
Lost to F/U	6	10	15
Deceased	1	11	13

Table 6: Distribution according to end-diagnosis

Diagnostic	No.	%	Sex (m/f)	Histological
Plasmacytoma	49	62	28/21	Hematopoietic
Chondrosarcoma	5	6.3	3/2	Chondrogenic
B-cell Lymphoma	5	6.3	3/2	Hematopoietic
Ewing Sarcoma	4	5	4/0	Neuroectodermal
Hemangioma	2	2.5	0/2	Vascular
T-cell Lymphoma	2	2.5	1/1	Hematopoietic
Osteochondroma	2	2.5	2/0	Chondrogenic
Osteoblastoma	2	2.5	1/1	Osteogenic
Chondroblastoma	1	1.3	0/1	Chondrogenic
Chordoma	1	1.3	0/1	Notochordal
Enchondroma	1	1.3	0/1	Chondrogenic
Langerhans-cell Histiocytosis	1	1.3	1/0	Miscellaneous
Leiomyosarcoma	1	1.3	0/1	Smooth Muscle
Mixed B-T-cell Lymphoma	1	1.3	1/0	Hematopoietic
Osteosarcoma	1	1.3	0/1	Osteogenic
Giant Cell Tumor	1	1.3	0/1	Giant Cell Tumor

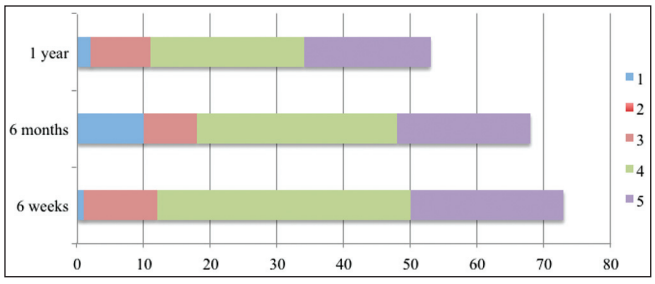

Figure 2: Glasgow Outcome Scale (GOS) at six weeks, six months and one year

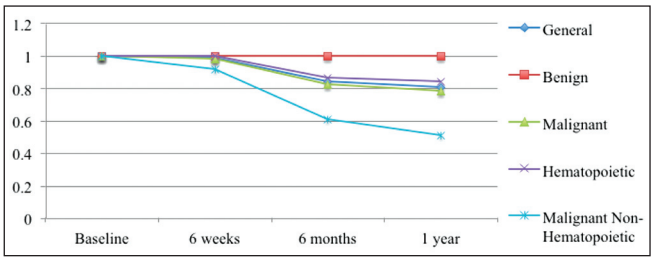

Figure 3: Kaplan-Meier curve showing the general overall survival at one year

Table 7: Associated pathology according to classification of bone tumors

Tumor Type	Pathology
Osteoma	Gardner Syndrome [60]
Osteosarcoma	Paget Disease, Retinoblastoma, Li-Fraumeni, Rothman-Thompson, Werner, Bloom, Langer-Giedion, Mazabraud [61]
Osteochondroma	Multiple Hereditary Exostosis Syndrome [60]
Chondroma	Ollier Disease, Mafucci Syndrome [60, 62]
Fibrous Dysplasia	McCune-Albright Syndrome [62]
Hemangiomatosis	Gorham's Disease [60]

In our study, both CT and MRI were performed in most patients (69.6%). However, radiographs were considered obsolete for the diagnosis of PBTS. Bone scintigraphy should not be performed initially, but it may still be considered as a complimentary diagnostic tool after histological diagnosis has been made.

Our study also confirmed the thoracic region as the most common location for the PBTS (65.8%) followed by the cervical region with 29.1% [Tables 3 and 9]. It is important to mention, that at the time of diagnosis, many patients (64.6%) presented with more than one lesion. In most cases, only the symptomatic lesion was surgically treated [Table 4].

Table 8: Epidemiological, genetic and histological factors

Tumor	Histology	Cytogenetic	Age (decade)	Vertebral Predisposition	Location
Osteochondroma	Chondrogenic	Mutations in TSG: *Exostosin-1 at 8q24; Exostosin-2 at 11p11-p12[63]*	2 [60]	No Difference	No Difference
Chondrosarcoma	Chondrogenic	Aberrations: *20q, 20p, 14q23-qter[63]*	5 [61, 64]	No Difference	Thoracic [64]
Osteoid Osteoma	Osteogenic	Involvement: *22q13, 17q [63]*	2 [60, 64, 65]	Posterior [60, 64, 65]	Lumbar [64, 65]
Osteoblastoma	Osteogenic	No Data	2-3 [60, 64]	Posterior [60, 64, 65]	No Difference [60]
Osteosarcoma	Osteogenic	Amplifications: *1q21-23 & 17p, 12q13-15, PRIM1, CDK4, 12p, MYC [63]*	4 [64]	No Difference	Lumbosacral [64]
Ewing Sarcoma	Neuroectodermal	Translocation: *t(11;22)(q24:q12), fusion protein EWS/FLI1 [63]*	1-3 [64]	No Difference	Lumbosacral [64]
Malignant Fibrous Histiocytoma	Fibrohistiocytic	LOH 9p21-22 [63]	5	No Difference	No Difference
Plasma Cell Myeloma	Hematopoietic	No Data	4-5 [64, 65]	Body [65]	Thoracolumbar [64]
Giant Cell Tumor	Unknown	Telomere Alteration: *11p, 13p, 14p, 15p, 19q, 20q, 21p [63]*	3 [64, 65]	Body [60, 65]	Sacral [60, 64, 65]
Chordoma	Notochordal	Lost Chromosomes: *3,4,10,13;TSP gene thought to exist on distal 1p [63]*	5 [64]	Body [65]	Cervical, Sacral [64, 65]
Hemangioma	Vascular	No Data	Any	Body [60, 64, 65]	Thoracic [65]
Aneurysmal Bone Cyst	Unknown	Rearranged Bands in Chromosomes: *16q22, 17p13 (USP6 gene); recurrent t(16;17)(q22;p13) [63]*	1-2 [60, 65]	Posterior [60, 64, 65]	Lumbosacral [65]
Langerhans Cell Histiocytosis	Unknown	X-Chromosome inactivation: *clonal [63]*	2[64]	Body [64]	Thoracic [64]

Table 9: Comparison between PBTS Series.

Study	Age (A/C)% Mean	Sex (m/f) %	Localization (Ce/T/L/S) % Total	Symptoms+ Yes/No	Hematopoietic/Non-Hematopoietic %	Benign/Malignant %	Survival
Leeds Registry[4]	84/16 42/13	52/48	14/38/16/31 127++	Yes	35/65	23/77	81% malignant at 6 years
Instituto Rizzoli[9]	N/A	N/A	17/33/49/NI 366	No	23/77	57/43	N/A
PTS Database[10]	*100 43	55/45	N/A	No	NI	51/49**	***72% at 5 years
Dusseldorf	74/5 55/12	56/44	^29/66/23/6 98	Yes	72/28	14/86	***80% at 1 year

A = adults; C = children; m = male; f = female; Ce = cervical; T = thoracic; L = lumbar; S = sacral; +If included in the study; ++Leeds Registry = one Patient reported had a bifocal chordoma (cervical & sacral); *No differentiation between adults and children; **Classification included nerve sheath tumors & unclassified benign/malignant tumors; *** No differentiation between benign and malignant; ^Number of lesions detected radiologically with same characteristics as the operated; NI = Not included; N/A = Not applicable

Patients presenting with a non-spinal metastasis had an osteosarcoma (1), chondrosarcoma (2), Ewing's sarcoma (2), lymphoma (1) and plasmacytoma i.e., multiple myeloma (1).

Surgical treatment

The two most common oncologic staging systems were described by Enneking and Weinstein-Boriani-Biagini. They allow the planning for the surgical procedure according to tumor extension.[2,8,18,23,24]

Over the years, there has been a trend to operate PBTS more aggressively, in order to achieve the best local control rates.[8,12,25,26] In the past, treatments such as curettage, embolization, bone grafting, marginal resection or decompression were utilized, but the recurrence rates were high, ranging from 40-60%.[21,22,24,27-31] In contrast, radical resection has a markedly reduced recurrence rate of 4-7%.[32-41] As a matter of fact, recurrence is highly correlated with mortality.[42,43] In our study, the recurrence rate was 15.2% and a strong negative predictive factor. All patients with malignant tumor recurrence died within 6 months following surgery. Analysis showed that the nature of the tumor was probably the cause of early recurrence rather than partial resection [Table 10].

In the present population, each surgery was individually planned based on tumor characteristics and clinical signs, as well as other criteria such as age, general condition, comorbidities and prognosis. In most cases, a near-total resection was performed in consideration of more than two-thirds of the population, who had hematopoietic tumor type, and an adjuvant therapy had to be initiated. In these cases, the aim was to minimize the surgical trauma and potential complications.

Biopsy

The value of tissue diagnosis by biopsy is still controversial. Despite modern imaging modalities, there are still numerous cases, in which it is impossible to differentiate benign from malignant tumors. In ambiguous cases, a biopsy may help to determine further management strategy.[14,25,44] Figures 4 and 5 illustrate such ambiguous cases. In our series, a preoperative biopsy was only performed in 3 patients (3.8%) and a percutaneous biopsy together with a vertebroplasty was performed in 16 patients (20.8%). Histopathological diagnosis could be established in 100% of specimen obtained either percutaneously or intraoperatively.

Adjuvant treatment

In some cases, adjuvant or neoadjuvant therapy is indicated. Unfortunately, only plasmacytoma, lymphoma and Ewing sarcoma are considered radiosensitive, with the latter being the only one, in which the patient also benefits from neoadjuvant radiotherapy.[26]

The use of radiotherapy in recurrent or unresectable osteoblastomas, osteosarcomas, and giant-cell tumors is controversial and is only considered if the patient has a recurrence or an unresectable lesion in order to achieve palliative local control.[45-47] The long-term risk of sarcomatous transformation in giant-cell tumors limits its use.[8,46,48-50] In hemangiomas, radiotherapy should only be used in partially resected tumors, but also showed good results as a primary treatment.[51,52] In chordomas or chondrosarcomas, there are no clinical trials, that studied the exclusive use of radiotherapy, but there are some studies suggesting better local control and increased survival rates as an adjuvant therapy.[53,54]

The use of chemotherapeutics is in many cases palliative, although neoadjuvant and adjuvant chemotherapy in osteosarcoma has shown to result in improved survival rates.[8,14,55,56] Ewing's sarcoma also favorably responds to combined neoadjuvant and/or adjuvant chemotherapy and radiotherapy, and is in conjunction with surgery, the mainstay of treatment for this type of tumor.[57] A combined approach is also employed for plasmacytoma, where radiotherapy is the established neoadjuvant treatment option.[57]

Despite the unknown mechanism of action in this pathology, the use of bisphosphonates is reported in the literature as an adjunct treatment for symptomatic benefit and local disease control.[8,57]

Hematopoietic tumors and the current WHO classification

The WHO Classification for PBT also includes plasma cell myeloma, solitary or osseous plasmacytoma and lymphoma of

Table 10: Association between resection, recurrence and histology

No	Age	Resection/Treatment	Time to Recurrence	Diagnosis	Histology	Classification
1	22	Complete	4 years	Chondroblastoma	Chondrogenic	Benign
2	76	Complete	9 years	Chondrosarcoma	Chondrogenic	Malignant
3	54	Partial	8 years	Chondrosarcoma	Chondrogenic	Malignant
4	47	Vertebroplasty	3 years	Hemangioma	Vascular	Benign
5	51	Complete	9 years	Osteochondroma	Chondrogenic	Benign
6	61	Complete	1 year	Osteosarcoma	Osteogenic	Malignant
7	59	Complete	2 years	Plasmacytoma	Hematopoietic	Malignant
8	56	Complete	2 years	Plasmacytoma	Hematopoietic	Malignant
9	35	Partial	2 years	Plasmacytoma	Hematopoietic	Malignant
10	79	Complete	1 year	Plasmacytoma	Hematopoietic	Malignant
11	62	Complete	6 months	Sarcoma	Smooth Muscle	Malignant
12	32	Complete	3 months	Ewing's Sarcoma	Neuroectodermal	Malignant

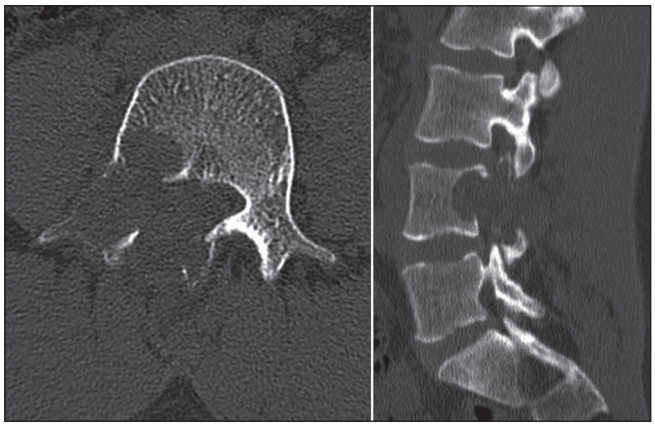

Figure 4: CT scan of the lumbar spine showing destruction of the right pedicle, a missing calcified tumor matrix and a non-sclerotic zone of transition to adjacent bone, which is not typical in benign bony lesions, but can occur in aneurysmal bone cysts

the bone. For the clinician, however, they may be considered a distinct group. Actually, they necessitate a more complex and different diagnostic work-up. Additionally, most of them do not require surgical treatment.

In most series of PBTS, including ours, hematopoietic tumors represent the most frequent type. If they were considered apart, the true incidence of PBTS would be even more rare. Therefore, international registries for PBTS are highly warranted and should be encouraged to further improve the epidemiology, classification, diagnostics and treatment of PBTS.

Outcome

In almost all patients, clinical improvement was seen at follow-up visits, especially in patients with paraparesis. The only patient with a persistent paraplegia had an unresectable tumor with symptoms, which already present for over 3 months prior to surgery. Although, a significant number of patients achieved symptom reduction at follow-up visits, 8 (10.1%) patients presented clinical worsening due to progressive disease or local recurrence [Figure 2].

Strong points and limitations

Our study represents the experience of a single tertiary care center over a 10-year period with a sizable number of patients and a reasonable length of follow-ups. This results in homogenous diagnostic work-up and management decisions. In comparison with other reported studies, the number of patients diagnosed and treated in a 10-year period was seemingly higher.[4,25,43,58,59]

Summarizing all PBTS in one study leads to reduced detail regarding individual tumor subtypes. On the contrary, it allows good overview of the complete spectrum of PBTS.

Referral bias likely explains the high proportion of malignant PBTS in our series, as a result of acute and/or severe onset of symptoms urging an immediate transfer to a surgical spine unit for therapy.

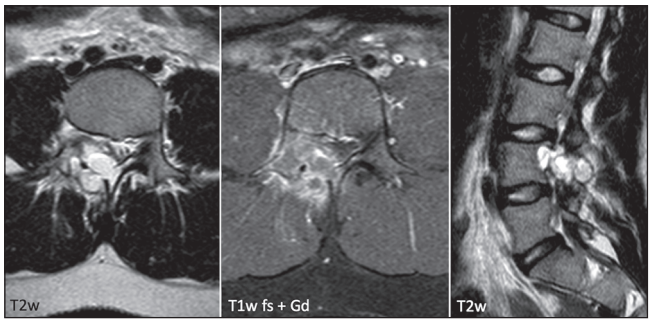

Figure 5: MRI of the lumbar spine (same patient) showing a cystic and expansile mass within the neural arch of the fourth lumbar vertebrae. The mass is well delineated typical for benign bony lesion, but it shows a diffuse, irregular enhancement and a peritumoral enhancing edema. Typical fluid-fluid levels within the cyst as, you can find in aneurysmal bone cyst were absent. Therefore, the differentiation between benign and malignant mass was difficult

Despite these limitations, we believe our study adds significant new insight regarding the rare PBTS. The large number of patients treated and studied at a single center over a decade gives this analysis relevant impact.

CONCLUSION

Most PBTS in our population occurred in adults and were malignant. Surgical treatment resulted in pain reduction and neurological improvement, allowing a rapid adjuvant therapy in good clinical condition. Malignant hematopoietic tumors had an intermediate one-year prognosis ranging between benign and other malignant PBTS. Although hematopoietic tumors are characterized by early systemic spread, spinal surgeons may well manage the ones presenting with acute primary spinal manifestation.

An experienced multidisciplinary team of tertiary care center should perform diagnosis and treatment of these extremely rare tumors.

In order to optimize management and elaborate guidelines, we encourage national or international registries, so that correct epidemiology, better classification and understanding, improved diagnostics, superior treatment and an overall better outcome may be achieved.

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