Radiology Case Reports 16 (2021) 145–151

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Malignant peripheral nerve sheath tumor in a child ☆,☆☆,★

Brian Daniel Noreña-Rengifo, MD[a,*], *Lina Marcela Cadavid-Alvarez, RR*[b,c], *Patricia Eugenia Gil-Serrano, RR*[d], *Gabriel Jaime Varela-Aguirre, PR*[b]

[a] *Department of Radiology, Universidad de Antioquia, Medellín, Colombia*
[b] *Department of Radiology, Hospital Pablo Tobón Uribe, Medellín, Colombia*
[c] *Department of Radiology, IMEDI, Rionegro, Colombia*
[d] *Department of Radiology, Sura Ayudas Diagnósticas, Cali, Colombia*

ARTICLE INFO

Article history:
Received 26 July 2020
Revised 12 October 2020
Accepted 17 October 2020

Keywords:
Malignant peripheral nerve sheath tumor
Nerve sheath neoplasms
Peripheral nerve
Child
Neurofibroma
Schwannoma

ABSTRACT

Among the diverse causes of posterior mediastinal masses, malignant peripheral nerve sheath tumors is a very rare neurogenic tumor. Imaging features tend to be variable. A 20-month-old toddler presented with a 3-month history of persistent diffuse thoracic and abdominal pain. A chest magnetic resonance imaging was taken and shown a posterior mediastinal lesion. Histopathology and immunohistochemical analysis confirmed the diagnosis of a malignant peripheral nerve sheath tumor with myxoid areas.

Malignant peripheral nerve sheath tumors are an uncommon entity in the children with a poor prognosis. Magnetic resonance imaging is the preferred technique in children to limit the use of ionizing radiation and because has a higher contrast resolution; however, all suspicious tumors should be biopsied to make an appropriate diagnosis. Treatment is radical surgery with excision of the entire mass; however, there is a high incidence of local recurrence.

© 2020 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Case report

Soft tissue sarcomas are rare childhood tumors [1]. Malignant peripheral nerve sheath tumors (MPNSTs) arise de novo or from preexisting benign neurofibromas or schwannomas [1].

We report the case of a 20-month-old toddler presented with a 3-month history of persistent diffuse thoracic and abdominal pain, weakness of extremities, fever and irritability. A chest X-ray reported a lung lesion that suggested a left basal pneumonia. After an initial antibiotic treatment, the patient persisted with the same symptoms, a thoracoabdominal computed tomography (CT) scan was then performed. CT showed a solid expansive lesion in the lower

☆ Competing Interests: The authors have declared that no competing interests exist..
☆☆ Consent: Did the author obtain written informed consent from the patient for submission of this manuscript for publication? Yes
★ Acknowledgments: None.

* *Corresponding author*.
E-mail address: brian.norena@udea.edu.co (B.D. Noreña-Rengifo).

https://doi.org/10.1016/j.radcr.2020.10.028

1930-0433/© 2020 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

lobe of the left lung. The lesion had a neoplastic appearance, with heterogeneous enhancement after administration of intravenous contrast material, mass effect and displacement of the mediastinum towards the right.

A chest magnetic resonance imaging (MRI) was then taken and a posterior mediastinal lesion with mediastinal mass effect, mostly on the left atrium, the left main bronchus and the pulmonary vessels, without stenosis of the supra aortic vessels or the descending aorta was observed (Fig. 1). No evidence of metastasis was found. Given these findings, a diagnosis of ganglioneuroma or ganglioneuroblastoma was made.

A CT-guided biopsy was then taken. Malignant peripheral nerve sheath tumor was reported in the pathology. Further studies checking for metastasis were negative.

Surgical resection was then performed through thoracotomy by the pediatric surgery group. An extra pleural mass of 10 × 10 × 6 cm was found in the posterior mediastinum, in close contact with the thoracic wall, compressing the entire left hemithorax and with a moderate amount of clear pleural fluid associated. There was no evidence of involvement of the spinal roots or the spinal canal. The pathology report showed a grade 2 MPNST with myxoid areas in French Federation of Cancer Centers Sarcoma Group grading soft tissue sarcomas, low mitotic rate, tumor edge involvement and tumoral necrosis. Immunohistochemical analysis showed no evidence of S100, calretinin, synaptophysin, smooth muscle actin or myogenin; and positive CD56, PGFA, and CD10 confirming the diagnosis of MPNST with myxoid areas (Fig. 2).

A postsurgical contrasted chest MRI showed an oval shaped residual lesion with well-defined borders, homogeneous enhancement and restrictive diffusion, involving the intervertebral foramen of spinal nerve T7, widening it to 14 × 13 × 10 mm. A postsurgical residual hematoma of 35 × 15 × 20 mm on the seventh intercostal space and a right laminar pleural effusion were also seen, with no consolidations or nodular images. No mediastinal or hilar lymphadenopathies were observed.

When the toddler was 22-month-old, he presented again with thoracic and abdominal pain, fever and irritability. A contrast-enhanced chest MRI was taken and showed an enlargement of the previously reported oval shaped lesion adjacent to the intervertebral foramen of spinal nerve T7, invading the spinal canal and occupying 50% of the epidural space, with displacement of the spinal cord to the right and no evidence of spinal injury (Fig. 3). The lesion had necrotic areas and fibrous septa (myxoid variety), and was surrounding 50% of the thoracic descending aorta, extending to the intercostal and intervertebral foramina T4 through T10 without invading the canal. Several lobulated masses were observed on the lateral and anterior pleural spaces, following the surgical incision's path (Fig. 4), all these findings suggested progression disease.

The toddler underwent to a second surgery to remove the posterior mediastinal tumor, thoracic wall, spinal canal, partial vertebrectomy, resection of the posterior wall, transverse processes and pedicles, and partial resection of the articular processes of T6 and T7. Pathology reported a malignant peripheral nerve sheath tumor with compromise of

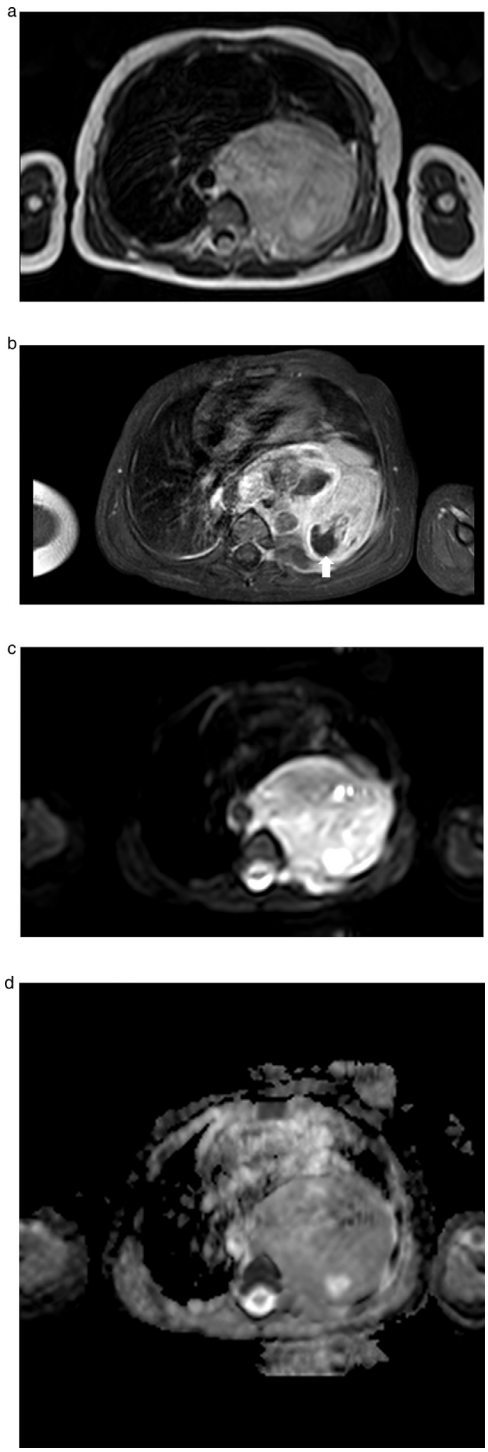

Fig. 1 – Chest magnetic resonance imaging (MRI) (a) Axial T2-weighted spin-echo (SE) image, mass hyperintense in posterior mediastinal. (b) Axial contrast-enhanced fat-saturated T1-weighted, heterogeneous with cystic/necrotic degeneration (arrow). (c) Axial diffusion-weighted imaging (DWI) b 1000 sec/mm^2, mass markedly increased signal intensity and (d) axial apparent diffusion coefficient (ADC) maps, decreased signal intensity, restricted diffusion

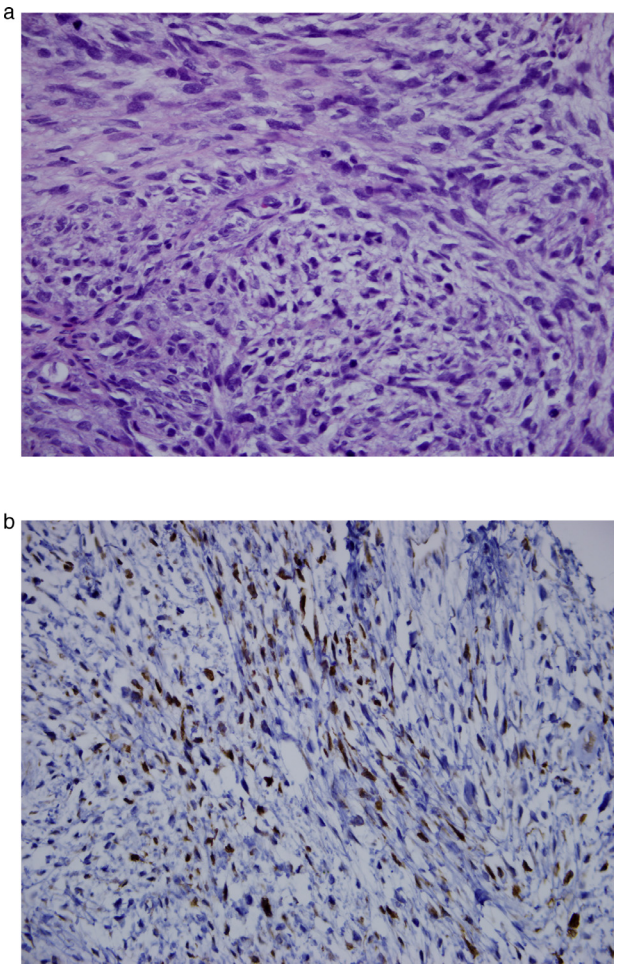

Fig. 2 – Pathological analysis. (a) Hematoxylin and eosin staining at 40x: pleomorphic tapered cells with multiple mitotic areas and some S-shaped nucleus. (b) Immunohistochemical staining for S100 protein: Focal nuclear reactivity of tumor cells

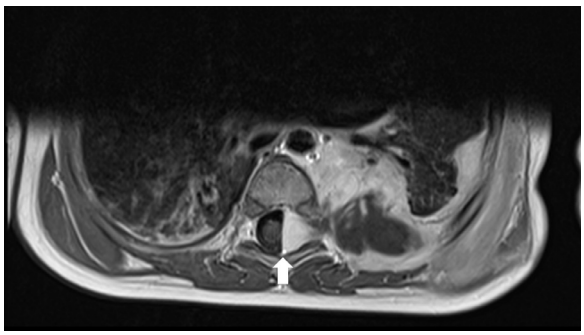

Fig. 3 – Chest magnetic resonance imaging (MRI). Chest magnetic resonance imaging (MRI). Axial contrast-enhanced T1-weighted, lesion adjacent to the intervertebral foramen of spinal nerve T7, with invasion of the spinal canal. The lesion occupied 50% of the epidural space and displaced the spinal cord to the right (arrow), without evidence of spinal injury. Several lobulated masses were observed on the lateral and anterior pleural spaces

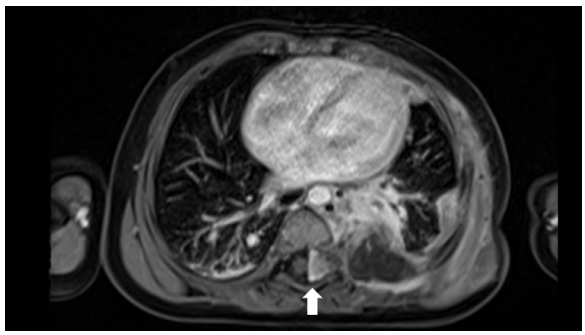

Fig. 4 – Chest magnetic resonance imaging (MRI). Axial contrast-enhanced fat-saturated T1-weighted, mass invading the spinal canal (arrow). Several pleural necrotic lobulated implants, following the surgical incision's path. All these findings suggested disease progression

the resection edges, tumoral necrosis and a high mitotic rate.

The toddler was evaluated by pediatric geneticists, who found macrosomia, postnatal macrocephaly and a family history of type 1 neurofibromatosis (NF1). They performed a sequencing panel for hereditary cancer, which included gene analysis for NF1, type 2 neurofibromatosis (NF2), NSD1, TP53, and PTEN, with nonpathological results found among the analyzed genes.

The clinical case was widely discussed in a multidisciplinary group as an initially low-grade tumor with an almost complete resection, which later had a local recurrence with spinal canal, pleural and diaphragmatic infiltration, histological change to a high-grade tumor, an extremely rare presentation in pediatric population, and poor prognosis due to the age of the patient, the high mitotic rate, rapid locoregional progression, "R2" resection surgery, and histological type. The chemotherapy treatment with palliative intention was rejected by the parents. A treatment with chemotherapy with an initially palliative intention was rejected by the parents.

The last image control with an MRI of the chest with contrast, when the toddler was 25 months old, showed postoperative changes with edema and extensive areas of enhancement in the left posterolateral chest wall, and resection of the posterior segment of the fifth, sixth, and seventh ribs. In the left epidural space of T7, in the same place where the previously known and later resected lesion of the spinal canal was located, a small nodule was observed in contact with the spinal cord, with no apparent signs of myelopathy. In addition, multiple pleural implants were found, with greater size and number compared to the previous MRI (Fig. 5).

Palliative robotic radiotherapy was proposed but it was ruled out due to evidence of tumor progression with multiple voluminous pleural masses, tumor lesions in contact with the great vessels, and a nodular lesion in the T7 epidural space. A new evaluation of the surgical group considered that the patient was not a candidate for a new surgical resection. Adjuvant radiotherapy was ruled out due to the high dose of radia-

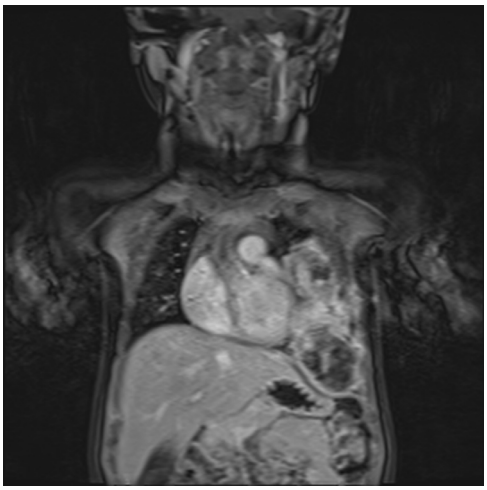

Fig. 5 – Chest magnetic resonance imaging (MRI). Coronal contrast-enhanced fat-saturated T1-weighted. Several pleural implants were found, with an increase in size and number showing peripheral enhancement

tion, and Cyberknife treatment was rejected due to the extension of the tumor.

Finally, in consensus with the parents, it was decided to offer the patient life support measures and symptomatic management. The toddler died 2 months later.

Discussion

Soft tissue sarcomas are a group of neoplasms originating from primitive mesenchymal tissue which represent 7% of all childhood tumors [1]. MPNSTs, previously called neurofibrosarcomas or malignant schwannomas [2,3], are rare malignant neuroectodermal neoplasms, arising de novo or from preexisting benign neurofibromas or schwannomas. However, only a small proportion of patients with neurofibromatosis type 1 (about 5%) develop malignant peripheral nerve sheath tumors [1].

MPNSTs account for 5%-10% of all soft tissue sarcomas and usually occur between 20 and 50 years of age and sporadic MPNSTs typically occur in the fifth decade, with no significant sex predilection [4–8]. Only 1.7% of them have been reported to occur in children less than 5 years old of age [9]. Studies in childhood MPNSTs report similar malignant behavior as in adults [1,9]. Genetic data suggest the loss of a tumor suppressor gene on chromosome 17 is important in the pathogenesis of MPNST [1].

Primary tumors most commonly occur in the extremities (45%-59%), followed by the trunk (17%-34%), and head and neck (19%-24%). It is an important diagnosis because it is one of the most aggressive malignant soft tissue sarcomas based on historically high rates of local recurrence and rapid disease progression [5,10].

We expose the case of a child with a MPNST to highlight the importance of an early diagnosis in this age. This kind of tumor is most commonly found in the extremities, but it can occur in the mediastinum. MPNSTs are typically seen in patients between 20 and 50 years of age, although occasional cases may be seen in the pediatric population being more common in children over 10 years old. They are very aggressive tumors and survival is poor [4,5].

A high proportion (about 25%-70%) is associated with NF1 (2,6), an autosomal dominant disorder resulting from the defective tumor suppressor protein neurofibromin 1 located on chromosome 17, but can also occur sporadically in the general population, just like our patient. Rapid tumor enlargement is more common in patients with NF1-associated MPNSTs and the long-term prognosis for them becomes even worse for them [1,6,7,10,11]. NF1-associated MPNSTs decrease 5-year disease-specific survival when compared with sporadic MPNSTs (16%-38% vs 42%-57% in sporadic disease) [2,4,5].

Intrathoracic primaries are often large at the time of diagnosis with complete surgical resection being difficult due to tumor surrounding vital vessels as in our patient, increasing the risk of local recurrence [1].

A MPNST clinically presents as a soft tissue mass that involves the major nerves and presents with pain and neurologic symptoms, such as paresthesia, motor weakness or radicular pain, related to the tumor's anatomic location as it enlarged [1,2,4,11]. A secondary MPNST may arise after radiation treatment, with a latent period that exceeds 10 years [7].

Microscopic and immunohistochemical analysis are imperative to establish a correct diagnosis of MPNST [9]. Pleomorphism of these tumors is wide, and they engender some of the most controversial, difficult differential diagnoses. Histologically, MPNST shows fasciculated growth pattern with hyperchromatic spindle cells. Nuclei are elongated, and perivascular hypercellularity is often seen. Marked mitotic activity and geographic tumor necrosis are also commonly present. These findings contribute in assessing the prognosis of the tumor [10]. Rare tumors show unusual histologic features with epithelioid, glandular, primitive, pleomorphic, schwannian, rhabdomyosarcomatous (triton tumor) differentiation or myxoid like our patient, it becomes our case in a rare presentation of a rare entity [4,12]. Low-grade MPNSTs arising from neurofibroma is diagnosed when there is generalized nuclear atypia, increased cellularity, and unusually low levels of mitotic activity [10].

Differential diagnosis should be done with neurofibromas and schwannomas. It is difficult to differentiate MPNSTs from neurofibromas. Neurofibromas are cellular, have low or no mitotic activity and show minimal nuclear atypia. Schwannomas are encapsulated tumors of nerve sheath and their progress commonly does not implicate the nerve intrinsically, they have nuclei arranged in stacked alignments knows as palisades [7,10].

Such as soft tissue neoplasms, MPNSTs are more easily characterized with advanced imaging techniques, but radiologic features are nonspecific. Diagnosis of a lesion suspicious for a MPNST should never be relied on imaging alone, as a result all suspicious tumors should be biopsied [5,9,13]. All scans should be reviewed with an experienced radiologist to glean as much information as possible concerning anatomic relations and possible pathologic findings, and to prevent unanticipated results in the operating room [13].

Radiographs of MPNSTs commonly are normal. Involvement of adjacent bone is rare, and then if it is found may suggest malignancy. On ultrasonography they look as well-defined, large, usually elongated, fusiform-shaped and hypoechoic masses that are oriented longitudinally in the nerve distribution, with tapered ends adjacent to the involved nerve. It may show posterior acoustic enhancement and simulate a cyst [5,7].

CT is not the preferred imaging technique, but it is useful to valuating the tumor extension and eventual metastasis, most frequently to bone and lungs [2,14]. On CT, MPNSTs are seen as a soft-tissue mass with a sudden increase in size or with heterogeneous attenuation due to internal necrosis and hemorrhage [3]. Neurofibromas are observed as a well-defined soft-tissue mass that has a low density relative to muscle, we also can observe low attenuation and little or no enhancement [1,5,7]. Angiography is not used routinely but it may show nonspecific imaging changes as displacement of major vascular structures around the involved nerve, enlarged vascularity and the presence of tortuous corkscrew nutrient feeding vessels at the upper and lower poles of the MPNST [7].

MRI is currently the modality of choice for revealing the anatomical location, contour, nerve origin, fat planes involvement and relationship of a MPNST with adjacent neural, vascular, and muscular structures because is superior to ultrasound and CT in demonstrating features of MPNST because it distinguishes the lesion from the fat tissue better than CT and its excellent tissue contrast and multiplanar capability and better contrast resolution [5,7,8,14,15]. It is always necessary to order contrast scans to evaluate the enhancing quality of the mass. Information on the enhancing qualities of the mass, combined with its appearance on T1 and T2 images can confer valuable clues as to the histopathology findings that may be encountered [1,13].

The MRI signal intensities are variables in MPNSTs but tumors with a high myxoid or water content would have hyperintensity on T2-weighted images, but tumors with more cellular components might be relatively hypointense [15]. Fat suppression sequences should be used to determinate the nerve involved; however, this technique can suffer from spatial and contrast resolution limitations in addition to motion artifacts as a result of longer imaging times. Then it is possible to require newer techniques using magnetic resonance neurography which can offer improved visualization and definition of MPNSTs, but magnetic resonance neurography requires available radiologists familiar with the short tau inversion recovery sequences necessary to obtain the desired images [13].

It is important to note the difference between a benign peripheral nerve sheath tumor (BPNST) and a MPNST [2]. Peripheral nerve sheath tumors (PNSTs) are usually isointense to adjacent muscles on T1-weighted images and hyperintense on T2-weighted images. BPNSTs and MPNSTs are spindle or ovoid shaped, and we found an ovoid shaped tumor in our patient by MRI. Target sign (central core of hypointensity surrounded by hyperintensity on T2-weighted and enhanced T1-weighted images) has always been described as being nearly pathognomonic for neurofibromas, but it can be seen in schwannomas and less frequently in MPNSTs; this finding corresponds pathologically to a more cellular Antoni type A region centrally and a more myxoid Antoni type B region peripherally. It is also more frequently to find the split fat sign (a rim of fat) or a fascicular appearance in a BPNST than in a MPNST, particularly in more differentiated BPNSTs; that appearance is because a layer of connective tissue or an epineurium surrounds the entire nerve trunk. Infiltrative margin, bone destruction, larger size (>5 cm), areas of hemorrhage, peritumoral edema, intratumoral lobulation, heterogeneous enhancement and necrotic or cystic areas are suggestive of malignancy; loss of the target sign in neurofibroma also indicate a MPNST [1–3,5,8,13,15,16]. No single sign is enough for diagnosis, except for bone destruction, and a combination of 2 or more features can help us in early diagnosis [16].

Positron-emission tomography (PET) with Fluorine-18-fluorodeoxyglucose (FDG) (FDG-PET) is a potentially useful, noninvasive method for detecting malignant changes in plexiform neurofibromas. PET-CT has shown to be of use in the identification of NF1-associated MPNSTs and detection of unsuspected and unusual metastatic sites of disease in childhood MPNST. Additionally, the surgeon may biopsy the area of maximum uptake of FDG and ensures that the biopsy reflects the highest grade of the tumor in a heterogeneous lesion [1,17].

Similar to most soft-tissue sarcomas, once the diagnosis of MPNST is suspected, surgery is the first treatment option. Radical surgical excision resection with the goal of achieving complete removal with negative margins is the treatment of choice in MPNST [8,10,13,14]. Due to a high risk of recurrence with incomplete resection, postoperative irradiation and chemotherapy may be necessary [2–4,11].

Treatment response should be evaluated using the same imaging technique as the initial assessment, except if the first imaging modality used was a CT to avoid the use of ionizing radiation, especially in children. On follow-up with MRI should be used the same planes and sequences [1].

A thoracoabdominal location, positive margin status and large tumor size (>5 cm) of MPNST is associated with increased incidence of local recurrence and then with adverse survival [4,5,7]. Predictors of distant metastases include large tumor size, high tumor grade and local recurrence [5]. The best outcome for patients with malignant peripheral nerve sheath tumor is expected after complete removal of tumor combined with adjuvant radiotherapy and chemotherapy [6,10]; however, prognosis of MPNSTs remains poor [11].

Teaching points

- MPNST is an uncommon entity in the children which have poor prognosis. Radical surgery is the treatment of choice; however, MPNST has a high incidence of local recurrence. A high suspicion for this diagnosis should be maintained
- MRI is the preferred imaging technique in children to limit the use of ionizing radiation and because has a higher contrast resolution; however, all suspicious tumors should be biopsied because distinction between BPNST and MPNST is difficult.

- Infiltrative margins, bone destruction, larger size (>5 cm), areas of hemorrhage, peritumoral edema, intratumoral lobulation, heterogeneous enhancement, absence of target sign and necrotic or cystic areas are suggestive of MPNST.
- MRI is useful in diagnosis, staging, surgery and radiotherapy planning and follow-up of children with MPNST. CT may be used to determinate metastasis.

Authors' contributions

Conception and design: Lina Cadavid–A, Daniel Norena–R, Patricia Gil–S

Acquisition of data: Patricia Gil–S, Daniel Norena–R

Analysis and interpretation of data: Lina Cadavid–A, Gabriel Varela–A, Daniel Norena–R

Drafting or revising the article: Lina Cadavid–A, Gabriel Varela–A, Daniel Norena–R, Patricia Gil–S

Questions

Question 1. Which of the following answer choices is not a malignant characteristic in a peripheral nerve sheath tumor (PNST)?

Answer choice 1. Positive target sign (central core of hypointensity surrounded by hyperintensity on T2-weighted and enhanced T1-weighted images)

Answer choice 2. Larger size (>5 cm)

Answer choice 3. Presence of necrosis and hemorrhage

Answer choice 4. Infiltrative margins

Answer choice 5. Bone destruction

Explanation for question 1 "[Infiltrative margin, bone destruction, larger size (>5 cm), areas of hemorrhage, peritumoral edema, intratumoral lobulation, heterogeneous enhancement, absence of target sign and necrotic/cystic areas are suggestive of MPNST]"

Question 2. Which of the following answer choices is false?

Answer choice 1. MPNSTs are not associated with neurofibromatosis type 1

Answer choice 2. MPNSTs is commonly associated with neurofibromatosis type 1

Answer choice 3. Neurofibromatosis type 1 (NF1)-associated MPNST usually has a lower mortality.

Answer choice 4. Rapid tumor enlargement is more common in patients with NF1-associated MPNST

Answer choice 5. NF1-associated MPNSTs decrease 5-year disease-specific survival (DSS) when compared with sporadic MPNSTs

Explanation for question 2 "[A high proportion (about 25%-70%) is associated with neurofibromatosis type 1 (NF1)]"

Question 3. Which of the following answer choices is true?

Answer choice 1. MPNSTs most commonly occur in the extremities

Answer choice 2. MPNSTs most commonly occur in the head

Answer choice 3 MPNSTs most commonly occur in the neck

Answer choice 4. MPNSTs most commonly occur in the trunk

Answer choice 5. MPNSTs most commonly occur in the mediastinum

Explanation for question 3 "[Primary tumors most commonly occur in the extremities (45%-59%), followed by the trunk (17%-34%), and head and neck (19%-24%)]"

Question 4. Which of the following answer choices is true?

Answer choice 1. MPNSTs account for 5%-10% of all soft tissue sarcomas

Answer choice 2. MPNSTs account for 20% of all soft tissue sarcomas

Answer choice 3. MPNSTs account for 30% of all soft tissue sarcomas

Answer choice 4. MPNSTs account for 50% of all soft tissue sarcomas

Answer choice 5. MPNSTs account for more than a half of all soft tissue sarcomas

Explanation for question 4 "[MPNSTs account for 5%-10% of all soft tissue sarcomas]"

Question 5. Which of the following answer choices is false?

Answer choice 1. Only 1.7% of them have been reported to occur in children less than 5 years old

Answer choice 2. Most of the MPNSTs occur in children less than 5 years old

Answer choice 3. Surgery is the first treatment option in MPNSTs

Answer choice 4. Radiographs are the preferred imaging technique for the diagnosis of MPNSTs

Answer choice 5. Bone destruction is always found in MPNSTs

Explanation for question 5: Answer choice 1 and 2 "[Only 1.7% of them have been reported to occur in children less than 5 years old]," Answer choice 3 "[Similar to most soft-tissue sarcomas, once the diagnosis of MPNST is suspected, surgery is the first treatment option]," Answer choice 4 "[Radiographs of MPNSTs commonly are normal]," Answer choice 5"[Involvement of adjacent bone is rare, and then if it is found may suggest malignancy]."

REFERENCES

[1] Park K, Van Rijn R, Mchugh K. The role of radiology in paediatric soft tissue sarcomas. Cancer Imaging 2008;8:102–15.
[2] Hrehorovich PA, Franke HR, Maximin S, Caracta P. Malignant peripheral nerve sheath tumor 1. RadioGraphics 2003;23(3):790–4.
[3] Gladish GW, Sabloff BM, Munden RF, Truong MT, Erasmus FF, Chasen MH. Primay thoracic sarcomas. RadioGraphics 2002;22(3):621–37.
[4] Stucky CH, Johnson KN, Gray RJ, Pockaj BA, Ocal IT, Rose PS, et al. Malignant peripheral nerve sheath tumors (MPNST): the mayo clinic experience. Ann Surg Oncol 2012;19:878–85.
[5] Kamran SC, Shinagare AB, Howard SA, Hornick JL, Ramaiya NH. A–Z of malignant peripheral nerve sheath tumors. Cancer Imaging 2012;12(3):475–84.
[6] Lee JH, Lee HK, Choi CG, Suh DC, Lee KS, Khang SK. Malignant peripheral nerve sheath tumor in the

parapharyngeal space : tumor spread through the eustachian tube. Am J Neuroradiol 2001;22(4):748–50.
[7] Chee DWY, Peh WCG, Shek TWH. Pictorial essay : imaging of peripheral nerve sheath tumours. Can Assoc Radiol J 2011;62(3):176–82.
[8] Kar M, Deo SVS, Shukla NK, Malik A, Dattagupta S, Mohanti BK, et al. Malignant peripheral nerve sheath tumors (MPNST) – clinicopathological study and treatment outcome of twenty-four cases. World J Surg Oncol 2006;4(55):1–8.
[9] Kudesia S, Bhardwaj A, Thakur B, Kishore S, Bahal N. Primary MPNST in childhood- a rare case report. Pathol Sect 2014;8(11):1–2.
[10] Gogate BP, Anand M, Deshmukh SD, Purandare SN. Malignant peripheral nerve sheath tumor of8 facial nerve : presenting as parotid mass. J Oral Maxillofac Pathol 2013;17(1):129–31.
[11] Mullins BT, Hackman T. Malignant peripheral nerve sheath tumors of the head and neck : a case series and literature review. Hindawi Publ Corp 2014;2014(368920):1–6.
[12] Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. Acta Neuropathol 2012;123(3):295–319.
[13] Gupta G, Mammis A, Maniker A. Malignant peripheral nerve sheath tumors. Neurosurg Clin NA 2008;19(4):533–43.
[14] Sperandio M, Di Poce I, Ricci A, Di Trapano R, Costanzo E, Di Cello P, et al. Malignant peripheral nerve sheath tumour : CT and MRI findings. Hindawi Publ Corp 2013;2013(517879):1–6.
[15] Li C, Huang G, Wu H, Chen W, Shih L, Lii J-M, et al. Differentiation of soft tissue benign and malignant peripheral nerve sheath tumors with magnetic resonance imaging. Clin Imaging 2008;32(2):121–7.
[16] Yu Yong-hui, Wu Jing-tao, Ye Jing, Chen Ming-xiang. Radiological findings of malignant peripheral nerve sheath tumor: reports of six cases and review of literature. World J Surg Oncol 2016;14:142. doi:10.1186/s12957-016-0899-0.
[17] Ferner RE, Golding JF, Smith M, Calonje E, Jan W, Sanjayanathan V, et al. [18F]2-fluoro-2-deoxy-D-glucose positron emission tomography (FDG PET) as a diagnostic tool for neurofibromatosis 1 (NF1) associated malignant peripheral nerve sheath tumours (MPNSTs): a long-term clinical study. Ann Oncol 2008;19(2):390–4.