

## RARE MALIGNANT TUMORS OF THE NECK IN CHILDREN

### TUMORES MALIGNOS RAROS DO PESCOÇO EM PEDIATRIA

Inês Santiago Martins<sup>1</sup>, Maria José Noruegas<sup>2</sup>, Paulo Coelho<sup>2</sup>, Rui Catarino<sup>2</sup>, Fátima Heitor<sup>3</sup>,  
Maria Conceição Sanches<sup>2</sup>

<sup>1</sup>Serviço de Imagiologia, Hospital Distrital de Santarém

Directora: Dra. Isabel Sapeira

<sup>2</sup>Serviço de Radiologia, Hospital Pediátrico de Coimbra, Centro Hospitalar e Universitário de Coimbra

Directora: Prof. Doutor Filipe Caseiro-Alves

<sup>3</sup>Serviço de Oncologia Pediátrica, Hospital Pediátrico de Coimbra, Centro Hospitalar e Universitário de Coimbra

Directora: Dra. Fátima Heitor

#### Correspondência

Inês Santiago Martins  
Hospital Distrital de Santarém  
Serviço de Imagiologia  
Av. Bernardo Santarém  
2005-177 Santarém  
e-mail: sm\_ines@hotmail.com

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#### Abstract

Neck masses are common findings in pediatric patients, and are benign in the majority of cases. In addition to the more common tumors (lymphoma, rhabdomyosarcoma), rare tumors are also encountered in the pediatric head and neck. The imaging and clinical findings usually are nonspecific in these tumors, but some of these clinical and imaging characteristics may aid in narrowing the differential diagnosis. Imaging studies are important in determining the location of the tumor, its relation to adjacent structures, and for staging and follow-up. Ultrasound is the first imaging study performed in pediatric patients, but Magnetic Resonance Imaging is the method of choice, because of its excellent soft-tissue contrast and lack of ionizing radiation.

The aim of this article is to review some malignant tumors that are rare in the neck of children, namely synovial sarcoma, primitive neuroectodermal tumor, rhabdoid tumor, myoepithelial carcinoma, dermatofibrosarcoma protuberans, and malignant peripheral nerve sheath tumor. Special emphasis is given to the imaging findings of each of these tumors.

#### Key-words

Head and Neck, Pediatric, CT, MR, Ultrasound.

#### Resumo

As tumefacções cervicais são frequentes em pediatria. A maioria são benignas, no entanto os tumores malignos também podem ocorrer. Para além dos mais comuns (linfoma, rabdomiossarcoma), há tumores raros que por vezes envolvem a cabeça e pescoço em idade pediátrica. Os achados clínicos e imagiológicos destes tumores são, na maioria dos casos, inespecíficos, no entanto algumas características clínicas e imagiológicas podem ajudar no diagnóstico diferencial. Os exames de imagem são muito importantes na avaliação da localização do tumor, a sua relação com as estruturas adjacentes, no estadiamento e no seguimento dos doentes. Em pediatria, a ecografia é muitas vezes o primeiro exame a ser realizado, mas a Ressonância Magnética é o exame de eleição na avaliação destes tumores pela sua resolução tecidual e ausência de radiação.

Este artigo tem como objectivo rever alguns dos tumores malignos que são raros no pescoço em pediatria, nomeadamente o sarcoma sinovial, tumor neuroectodérmico primitivo, tumor rabdoide, carcinoma mioepitelia, dermatofibrosarcoma protuberans e o tumor maligno das bainhas nervosas periféricas, dando ênfase aos seus achados imagiológicos.

#### Palavras-chave

Cabeça e Pescoço, Pediatria, Tomografia Computorizada, Ressonância Magnética, Ecografia.

#### Introduction

Neck masses are common findings in pediatric patients and can be classified as congenital, vascular, inflammatory, benign and malignant tumors. In some series lymphoma was the most common malignant tumor involving the cervicothoracic area. The head and neck region is also the most common site of rhabdomyosarcoma, which accounts for more than 50% of soft-tissue sarcomas in children<sup>1-3</sup>. In addition to the more common tumors, there are rare tumors that affect the neck of children, and tumors that although not so infrequent, are rare in the pediatric age. In this article we propose to review a few of these malignant tumors that are rare in the neck of children. The tumors to be reviewed here are synovial sarcoma, primitive neuroectodermal tumor, rhabdoid tumor, myoepithelial carcinoma, dermatofibrosarcoma protuberans, and malignant peripheral nerve sheath tumor.

Imaging studies are usually performed in the workup of these patients. Ultrasound should be the first imaging modality for evaluation of suspected masses in children. In cases of large lesions or in case of malignancy, magnetic resonance imaging (MRI) is the modality of choice because of its excellent soft-tissue contrast and lack of ionizing radiation<sup>4</sup>. Cross sectional imaging is essential for tumor staging and planning surgical resection, and biopsy is necessary for an accurate diagnosis.

#### Synovial sarcoma

Synovial sarcoma is the fourth most common soft-tissue sarcoma. It affects adolescents and young adults, most often in the extremities, particularly the knee<sup>5</sup>. The head and neck are rarely affected, and even less in the pediatric population<sup>6</sup>. Despite its name, this tumor does not usually arise in an intraarticular location but rather occurs adjacent

to a joint or within 5 cm of it. In the head and neck, they appear to originate from the paravertebral connective tissue spaces<sup>7</sup>.

Patients present with a slow growing soft-tissue mass, associated to pain and tenderness, and the long duration of symptoms may simulate a benign process<sup>5</sup>.

On ultrasound this tumor appears as a solid, round or lobulated, hypoechoic and heterogeneous soft-tissue mass (Fig 1 a). Hypoechoic areas representing hemorrhage or necrosis may occur<sup>5</sup>. On CT the tumor is isodense or slightly hypodense to muscle, with areas of low attenuation representing necrosis or hemorrhage. Synovial sarcoma frequently demonstrates a multinodular appearance, and heterogeneous enhancement (Fig 1 b). Calcifications may occur<sup>5</sup>. MRI (Fig 1 c-f), shows a heterogeneous, well-defined, multilobulated soft-tissue mass with signal intensity similar to or slightly higher than muscle on T1-weighted imaging (WI), and high signal intensity on T2-WI. The signal heterogeneity has been termed the “triple sign” (Fig 1 c,d), represented by intermixed areas of high, intermediate and low signal intensity, and representing areas of hemorrhage or necrosis, solid tumor, and calcified or fibrotic regions, respectively. This sign is also present in other neoplasms. Intervening septa are frequently present, as well as fluid levels. This combination of features creates a “bowl of grapes” appearance. Smaller lesions are more homogeneous. Prominent, heterogeneous and early enhancement is usually present<sup>5</sup>. A high-grade synovial sarcoma may be suggested by

some imaging features, including the absence of calcification, presence of cystic components, hemorrhage, and the triple sign. Synovial sarcomas frequently invade adjacent muscle<sup>5</sup>.

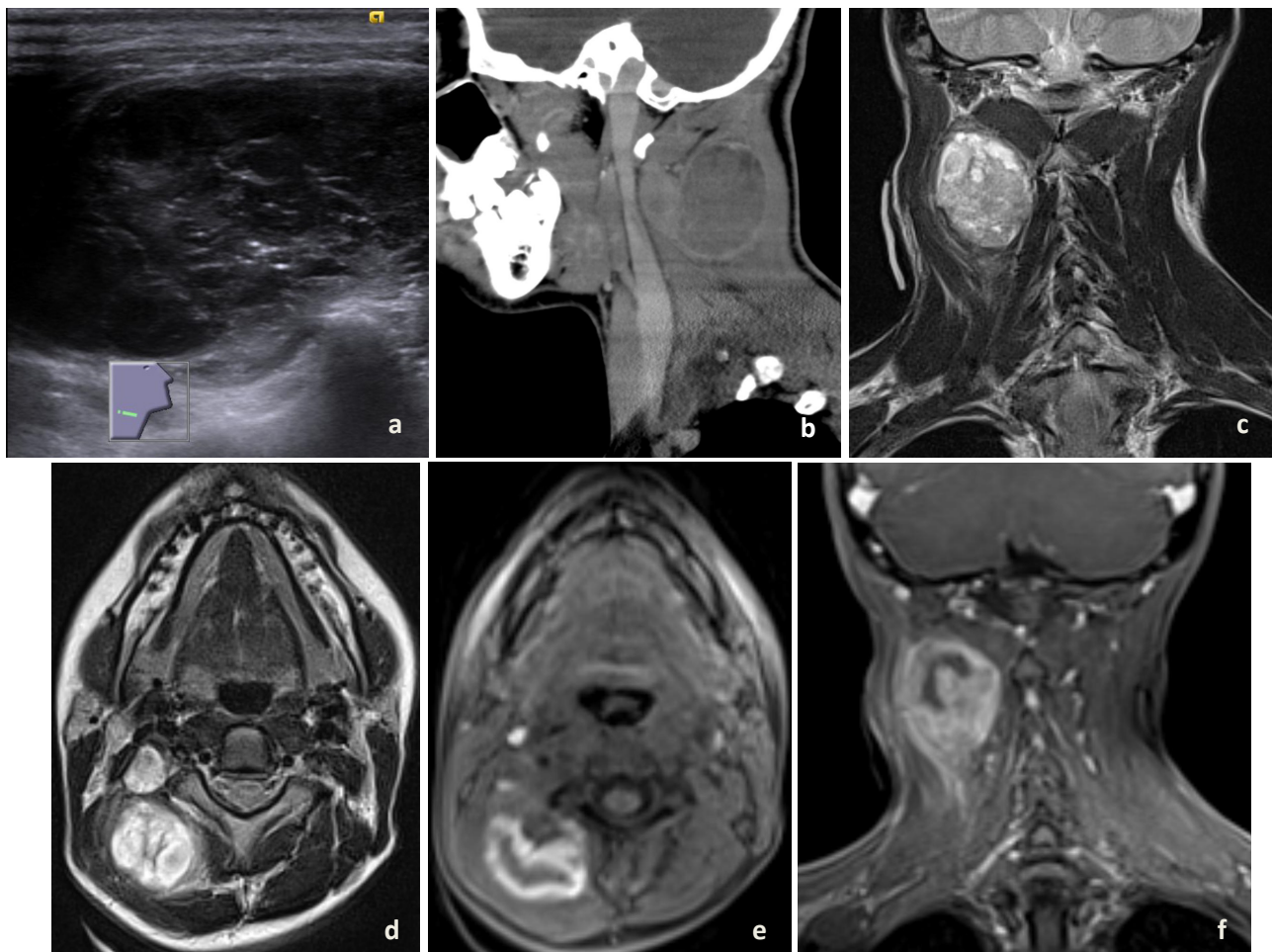
Surgery is the treatment of choice, being the role of adjuvant therapy controversial. Synovial sarcoma is an intermediate-to high-grade lesion, and, despite aggressive surgical resection, local recurrence occurs in up to 50% of patients and metastatic disease develops in 41%, to the lung, lymph nodes and bone<sup>5</sup>.

### Rhabdoid tumor

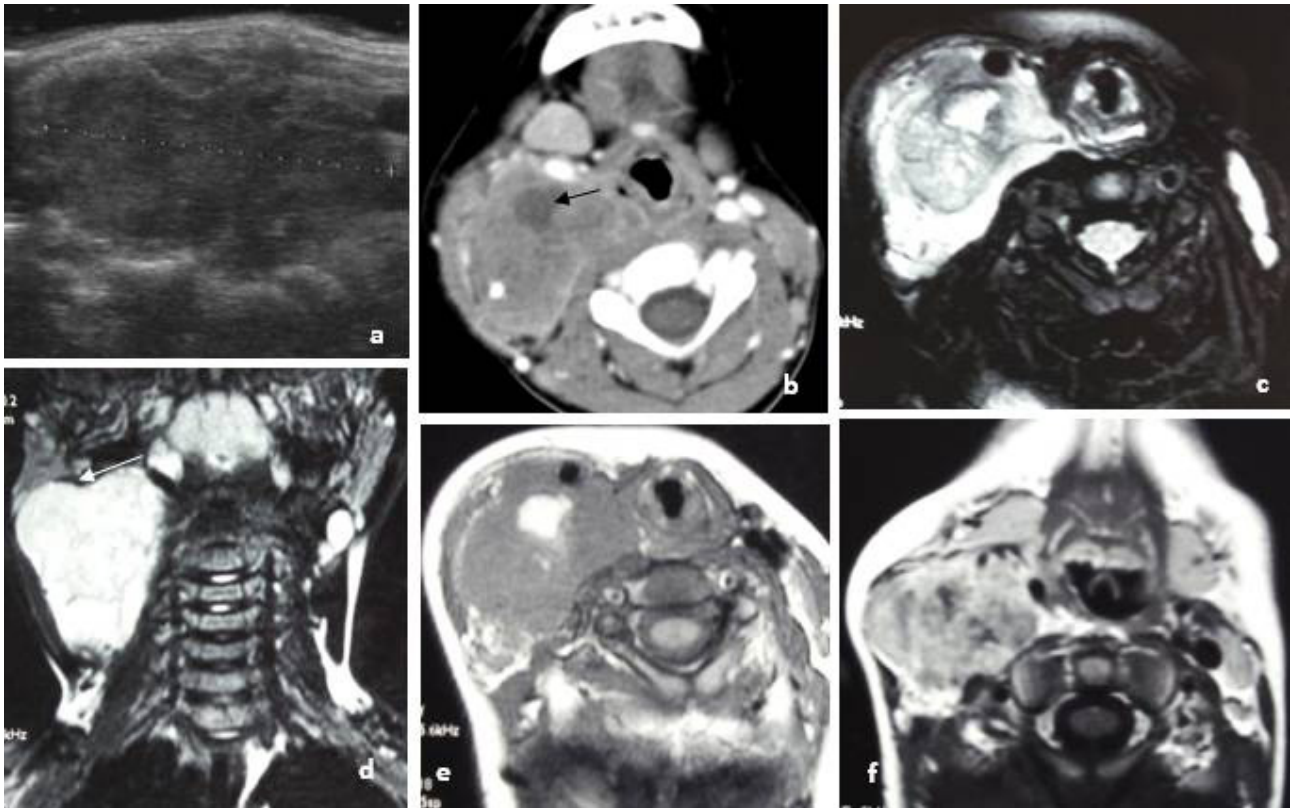
Malignant rhabdoid tumors are highly aggressive neoplasms that most commonly arise in the kidneys of young children<sup>8</sup>, most cases in the first year of life<sup>9</sup>. Extrarenal malignant rhabdoid tumors are extremely rare, and have been described in other organs, including the neck<sup>8</sup>.

Malignant rhabdoid tumors usually present with palpable, rapidly growing masses, often at an advanced stage<sup>10</sup>.

The imaging characteristics are nonspecific, most cases presenting with large, ill-defined and heterogeneous masses. A large heterogeneous solid mass is found on ultrasound (Fig 2 a). CT may show a large, heterogeneous, hypodense lesion (Fig 2 b), with or without calcifications. MRI (Fig 2 c-f) shows an ill-defined mass lesion with heterogeneous hyperintensity on T2-WI with areas of necrosis, with contrast enhancement, restricted diffusion and reduced ACD values, reflecting the aggressive biological behavior of the tumor.



**Figure 1** - Synovial sarcoma in the right postero-lateral region of the neck in a 15-year-old male. (a) Ultrasound. (b) Contrast-enhanced sagittal CT. (c) Coronal T2-WI MRI. (d) Axial T2-WI MRI shows the juxtaarticular location of the tumor, adjacent to the cervical vertebrae C2 and C3. (e) High signal intensity on T1-WI represents hemorrhage (f) Coronal contrast-enhanced T1-WI with fat saturation.



**Figure 2** - Malignant rhabdoid tumor in the posterior cervical space of the right lateral region of the neck in a 2-year-old boy with a 1-month history of rapidly growing neck swelling. (a) Ultrasound (b) Contrast-enhanced axial CT with a central non-enhancing area that might represent hemorrhage / necrosis (arrow). (c) Axial T2-WI MRI with fat saturation. (d) Coronal T2-WI with fat saturation. The mass contacts superiorly the parotid gland (arrow). (e) Axial T1-WI. An hyperintense central area represents hemorrhage. (f) Axial contrast-enhanced T1-WI shows heterogeneous enhancement of the lesion. Pathologic specimen revealed invasion of the carotid artery, internal jugular vein and vagus nerve.

These tumors can be locally invasive<sup>8</sup>.

Various combinations of multimodal therapy are needed, including surgical resection, chemotherapy and radiation<sup>10</sup>. Metastatic disease may occur in the lung and liver<sup>9</sup>. The course of extrarenal malignant rhabdoid tumors is short, and patients die 6 months after diagnosis<sup>8</sup>.

### Myoepithelial carcinoma

Myoepithelial carcinoma of the head and neck is a rare lesion, in most cases arising in salivary glands in adults. Myoepithelial carcinomas may also arise from other soft tissues, and have rarely been reported in children. Patients present with a painless, slowly growing mass<sup>11-14</sup>.

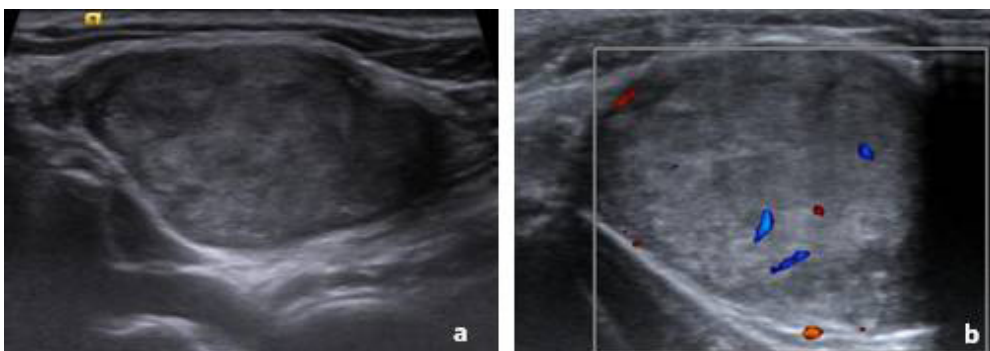
On CT this tumor is reported to show heterogeneous enhancement with partial calcification<sup>15</sup>. MRI shows a lobulated, heterogeneous mass, hyperintense on T2-WI and hypointense on T1-WI, with well-circumscribed margins.

It may show necrotic areas<sup>12,14</sup>. In our patient, ultrasound findings were of a heterogeneous solid lesion with well-defined and lobulated contours, and vascularization on color-Doppler (Fig 3).

Wide surgical excision is the treatment of choice, and predicts a better prognosis. The behavior of head and neck myoepithelial carcinoma is very variable. Frequently it recurs locally and metastases are common from several months to many years after the initial resection, to the lungs, brain, bone, kidneys and skin<sup>11</sup>. Myoepithelial carcinomas in children seem to have a more aggressive clinical course than in adults<sup>12</sup>.

### Malignant Primitive Neuroectodermal Tumor

Primitive neuroectodermal tumor (PNET) is a term used to describe a group of highly malignant small round cell tumors of neuroectodermal origin<sup>16</sup>. This broad family can be subdivided into three groups: central PNET, referring to



**Figure 3** - Intermediate grade myoepithelial carcinoma in a 9-year-old male, appears as a mass in the posterior left side of the neck. (a) Ultrasound. (b) Color-Doppler ultrasound



tumors arising from the central nervous system; tumors arising from the autonomic nervous system; and peripheral PNET (pPNET), referring to PNET arising outside the central and autonomic nervous system. Malignant pPNET encompasses various tumors, such as osseous and extraosseous Ewing sarcoma, malignant peripheral neuroepithelioma of bone and soft tissues, etc<sup>17</sup>. For practical purposes in this text it is termed as “malignant pPNET”.

Extraskelletal malignant pPNET is rare, and is rarely encountered in the head and neck<sup>16,17</sup>. It usually manifests between 20 months and 30 years of age. Patients present with a painful, rapidly growing, superficial or deep soft-tissue mass usually measuring 5-10 cm. Cervical masses can be large enough to cause compression of adjacent structures<sup>18</sup>. On ultrasound the lesions are hypoechoic, with anechoic areas representing hemorrhage or necrosis, and with increased Doppler blood flow<sup>18,19</sup> (Fig 4 a).

CT demonstrates a poorly defined soft-tissue mass, isodense to muscle, and may have low density areas corresponding to hemorrhage or necrosis (Fig 4 b). Calcification may be present. Osseous involvement of the bone surface with cortical erosion or periosteal reaction is seen in 40% of cases<sup>18</sup>.

MRI demonstrates a soft-tissue mass with heterogeneous signal intensity similar to that of muscle on T1-WI and intermediate to high signal intensity on T2-WI. Fluid levels, necrosis or hemorrhage may be evident<sup>18</sup> (Fig 4 c-f). An additional feature of extraskelletal malignant pPNET is the presence of serpentine high-flow channels, which have low signal intensity in all pulse sequences. Although this finding can be also seen in other tumors, the presence of this feature in a young person with a large intramuscular mass should raise the possibility of extraskelletal malignant pPNET. A pseudocapsule with well-defined margins may be seen at MRI, but in other cases an infiltrative pattern of growth

with ill-defined margins is present. Prominent contrast enhancement is seen at both CT and MRI<sup>18</sup>.

Treatment of the malignant pPNET family of tumors is usually a combination of neoadjuvant chemotherapy and surgical resection, which may be supplemented with radiation therapy and adjuvant chemotherapy<sup>18</sup>.

These tumors are aggressive with a propensity for locoregional recurrence and rapid metastatic spread to lung, liver, bone, brain and lymph nodes. The prognosis is poor and there is a high mortality<sup>16,17</sup>.

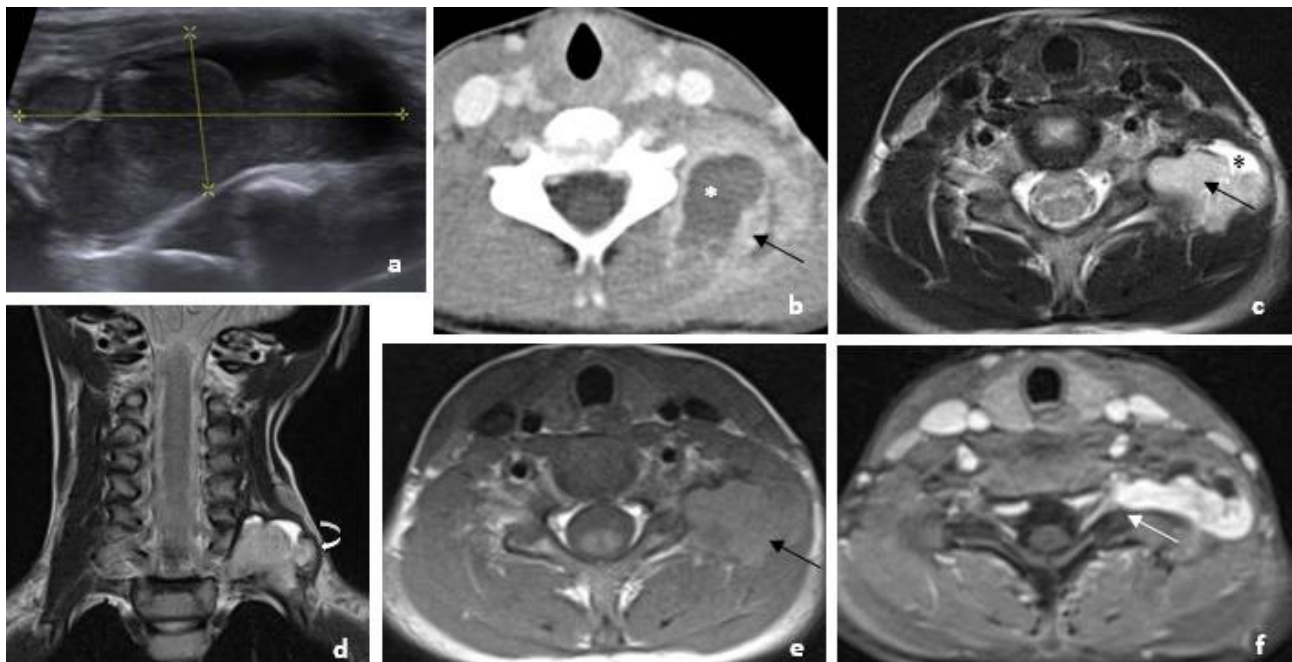
### Dermatofibrosarcoma protuberans

Dermatofibrosarcoma protuberans (DFSP) is a rare superficial soft-tissue sarcoma affecting the skin. It is an infiltrative tumor of low to intermediate-grade malignancy<sup>20,21</sup>. It affects more frequently young to middle-aged adults, and very rarely the pediatric population<sup>20,22,23</sup>.

The trunk is the most common site of involvement, followed by the proximal extremities and less commonly the head and neck<sup>21,24</sup>. When DFSP affects the head and neck, it involves the scalp, cheek, and supraclavicular areas<sup>25</sup>.

DFSP presents as a painless, firm and well defined superficial nodule or polypoid protuberance measuring from 1 cm to more than 25 cm<sup>21,24</sup>. The lesions can be multiple and may coalesce to form an indurated plaque, which on clinical examination is often violaceous<sup>24</sup>. As a result, it can be mistaken clinically for a vascular malformation or a hemangioma. The tumor originates in the dermis but spreads locally into the subcutaneous tissues, fascia, muscles, and bones<sup>25</sup>. The growth rate is variable; some lesions remain stable in size for many years, whereas others demonstrate slow progressive growth<sup>24</sup>.

It is often diagnosed by its typical clinical manifestations. Imaging methods, mainly MRI, are used to evaluate atypical primary lesions and to define the extent of the tumors, thus



**Figure 4** - Extraskelletal malignant pPNET in the left posterior cervical / perivertebral space in a 9-year-old boy. (a) Ultrasound shows a heterogeneous lesion with solid and cystic areas. (b) Contrast-enhanced axial CT shows a mass with cystic (asterisk) and solid areas, with prominent enhancement of the solid component (arrow). (c,d) Axial and coronal T2-WI show a heterogeneous lesion with a hyperintense solid component (arrow) and a cystic component (asterisk). A hypointense pseudocapsule is seen (curved arrow in d). (e) Axial T1-WI reveals an isointense lesion (arrow). (f) Axial contrast-enhanced T1-WI. The mass is seen to insinuate between the transverse processes of C7 and D1 vertebrae (arrow).

allowing accurate preoperative assessment<sup>21-25</sup>. The imaging findings of DFSP are nonspecific, but the combination of imaging findings, location of the lesion, and clinical features help narrow the differential diagnosis<sup>24</sup>.

In our patient, ultrasound showed a hypoechoic and heterogeneous lesion, with oval shape and well-defined borders, located in the subcutaneous tissue, with moderate vascularization on color-Doppler (Fig 5 a). CT findings (Fig 5 b) include isodense to hypodense lesions without calcifications, and uniform (small lesions) to heterogeneous (large lesions) enhancement<sup>24</sup>.

MRI shows a well-defined lesion (Fig 5 c,d) with low to intermediate signal on T1-WI and intermediate to high signal on T2-WI, with uniform and marked enhancement. Areas of high signal on T1-WI may occur, compatible with hemorrhage<sup>21,24,25</sup>.

Treatment includes complete surgical excision, and/or adjuvant radiotherapy. After complete resection the prognosis is excellent, otherwise it has a high propensity for local recurrence. Recurrence after five years has been reported, and therefore it is important to follow these patients over a

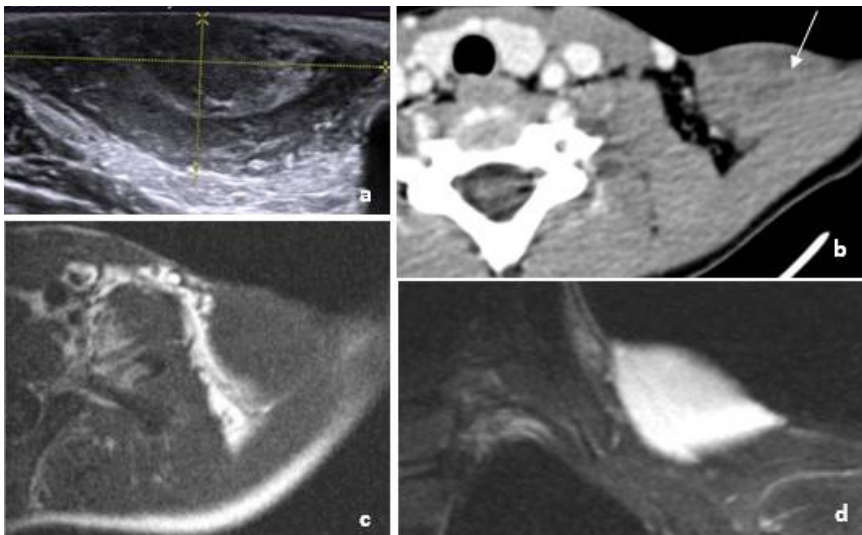
long period of time. Distant metastases are infrequent, with the lungs and bones being the most common sites<sup>21,22,24,25</sup>.

### Malignant peripheral nerve sheath tumor

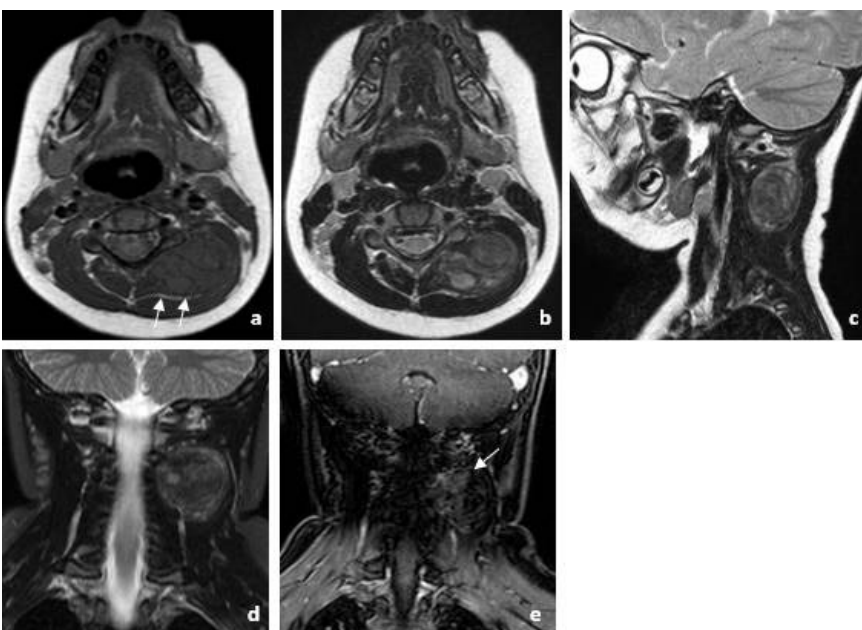
Malignant peripheral nerve sheath tumors (MPNST) is the current term used by the World Health Organization for a group of tumors of neurogenic origin with the appearance of any nerve sheath cell. These tumors are very rare and are high-grade sarcomas. The past literature referred to MPNST as malignant schwannoma, malignant neurilemmoma, neurogenic sarcoma, and neurofibrosarcoma<sup>26-29</sup>.

They are solitary lesions that occur in ages 20–65 and are found infrequently in children. About 50% occur in patients with neurofibromatosis type 1, who develop these tumors at an earlier age<sup>26,30</sup>.

MPNST often arise in the deep soft tissues in proximity to peripheral nerves, usually in the extremities and the trunk and are seldom found in the head or neck. It presents clinically as a rapidly enlarging soft tissue mass (larger than 2–6 cm), with irregular borders, with or without pain and dysesthesia; most of the lesions are deeply located<sup>27-30</sup>.



**Figure 5** - DFSP in the left supraclavicular region in a 8-year-old boy, presenting as a slow growing mass with 4 years of evolution. (a) Ultrasound (b) Axial contrast-enhanced CT scan demonstrates a hypodense lesion (arrow). The lesion is isointense on T1-WI (c), and uniformly hyperintense on T2-WI (d).



**Figure 6** - MRI of a low grade MPNST in the left perivertebral cervical space in a 2-year-old boy. (a) Axial T1, (b) axial T2, (c) sagittal T2, (d) coronal T2 with fat saturation and (e) coronal contrast-enhanced T1 with fat saturation. The mass is heterogeneous possibly due to necrosis and/or hemorrhage, and is mainly isointense on T1 and hyperintense on T2. The borders are well defined and the "split-fat" sign is seen (arrow in a). There is slight enhancement after administration of gadolinium (arrow in e).

MRI is the imaging study of choice<sup>28,31</sup> (Fig 6). Some imaging characteristics may suggest a peripheral nerve sheath tumor such as a location in the region of a major nerve, depiction of the nerve entering or exiting the mass, and the presence of certain signs: split fat sign (a rim of fat surrounding the lesion), fascicular sign (appearance of the fascicular bundles), target sign (low signal intensity centrally and high signal intensity peripherally). The target and fascicular signs are typically seen in benign lesions, although they may occasionally be seen in MPNST<sup>26,27</sup>. Signs of necrosis and hemorrhage and patchy contrast enhancement are seen in MPNST<sup>28</sup>.

Aggressive biologic behavior may be suggested by rapid growth, indistinct margins, heterogeneity and its infiltrative nature within the nerve and adjacent structures.

Complete surgical resection is the mainstay of successful treatment. Postoperative radiotherapy is recommended. MPNST has a high rate of local recurrence via direct perineural invasion, and has a propensity to distant metastases to lungs, bone and pleura. MPNST is one of the most aggressive tumors in the head and neck area, with relatively poor outcomes in young patients<sup>29-33</sup>.

## Conclusion

The imaging and clinical findings of these tumors are nonspecific in the majority of cases. However, imaging

studies, particularly MRI, are useful in determining the site of tumor origin, its extent and relation to adjacent structures, and for follow-up. In some of these tumors, the combination of findings may aid in narrowing the differential diagnosis. Anyway, histological analysis after tissue biopsy is required for final diagnosis of these tumors.

A soft-tissue mass, particularly if calcified, near but not in a joint of a young patient, may suggest the diagnosis of synovial sarcoma. A malignant rhabdoid tumor should be included in the differential diagnosis list in the presence of an extremely aggressive tumor of early childhood, regardless of tumor location. In a young person, the presence of a large intramuscular mass, containing serpentine high-flow channels, which have low signal intensity with all pulse sequences, should raise the possibility of an extraskeletal malignant pPNET, although it may also be seen in other tumors. Often, the diagnosis of DFSP is clinical because of its typical appearance and superficial location. Imaging studies may help in atypical cases, demonstrating a well-defined, subcutaneous lesion. The presence of MPNST may be suggested by signs indicating the neurogenic origin of the lesion, and signs that suggest an aggressive biologic behavior.

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