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Leg myxoid liposarcoma: correlation between image and anatomopathological tests

Lipossarcoma mixoide da perna: correlação entre os exames de imagem e anatomopatológico

Marcio Luís Duarte[1]; Élcio Roberto Duarte[2]; Edgard B. Solórzano[3]; Jael B. A. Ferreira[3]

1. Webimagem, Santos, São Paulo, Brazil. 2. Hospital Irmã Dulce, Praia Grande, São Paulo, Brazil. 3. Brasil Imagem Medicina Diagnóstica, Santos, São Paulo, Brazil.

ABSTRACT

A 41-year-old male patient refers to a painful tumor on the lateral side of the right leg. Ultrasonography visualized a predominantly hypoechoic well-delineated large heterogeneous mass in the subcutaneous tissue, showing an internal hyperechoic area. Magnetic resonance imaging (MRI) demonstrated an oval and lobulated tumor with well-defined margins at the anterolateral part of the leg, with contrast enhancement. The anatomopathological evaluation revealed a neoplasm with an extensive chondromyxoid matrix. The immunohistochemical study demonstrated positive vimentin (V9 clone), positive S-100 protein and negative desmin (D33 clone), smooth muscle actin (1A4), CD68 (KP1 clone) and caldesmon. The set of findings is compatible with myxoid liposarcoma.

Key words: liposarcoma myxoid; ultrasonography; magnetic resonance imaging.

RESUMO

Paciente do sexo masculino, 41 anos de idade, relata tumor doloroso na face lateral da perna direita. A ultrassonografia demonstrou massa volumosa, hipoecogênica, heterogênea e bem delineada no tecido subcutâneo, com área hiperecogênica em seu interior. A ressonância magnética (RM) revelou um tumor ovalado e lobulado, com margens bem definidas na parte anterolateral da perna, apresentando realce pelo contraste. O estudo anatomopatológico constatou neoplasia com extensa matriz condromixoide. O estudo imuno-histoquímico apresentou vimentina (clone V9) e proteína S-100 positivas e desmina (clone D33), actina de músculo liso (1A4), CD68 (clone KP1) e caldesmon negativos. O conjunto de achados é compatível com lipossarcoma mixoide.

Unitermos: *lipossarcoma mixoide; ultrassonografia; imagem por ressonância magnética.*

RESUMEN

Paciente masculino de 41 años de edad, reporta tumor doloroso en la parte lateral de la pierna derecha. La ultrasonografía demostró una masa voluminosa, hipoecogénica, heterogénea y bien delimitada en el tejido subcutáneo, con área hiperecogénica en su interior. La resonancia magnética (RM) reveló un tumor ovalado y lobulado, con márgenes bien definidas en la parte anterolateral de la pierna, presentando realce de contraste. El estudio anatomopatológico constató neoplasia con abundante matriz condromixoide. La inmunohistoquímica presentó vimentina (clon V9) y proteína S-100 positivas, y desmina (clon D33), actina de músculo liso (1A4), CD68 (clon KP1) y caldesmón negativos. El conjunto de hallazgos es compatible con liposarcoma mixoide.

Palabras clave: *liposarcoma mixoide; ultrasonografía; imagen por resonancia magnética.*

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INTRODUCTION

Liposarcoma is a malignant soft-tissue tumor and the most common sarcoma in adult life[1, 2], present in the lower limbs in about 75% of the cases[2, 3].

The World Health Organization (WHO) ranked the soft-tissue liposarcoma into five different histological subtypes: well differentiated, myxoid, dedifferentiated, pleomorphic or mixed[4].

The myxoid liposarcoma accounts for 30%-35% of all liposarcomas[1] and occurs predominantly in young adult extremities[5], between the fourth and the fifth decades of life, with no sex predilection[3].

As a rule, it is seen in the thighs and has a low propensity for metastasis[3, 6], being, more commonly, extrapulmonary[4]. The rate of metastatic disease is significantly higher for patients whose tumor contains a greater proportion of round cell component[4].

CASE REPORT

A 41-year-old male patient refers to a painful tumor on the lateral side of the right leg after a local injury five years ago. The lesion has been growing in size since then. He denies functional and labor limitations. Refers to systemic hypertension and diabetes mellitus in treatment. The patient is a smoker and social drinker. Physical examination shows a tumor on the right side of the leg, without skin change. Mobility in the leg, hip, knee, and ankle on the right side is not limited. Ultrasonography visualized a large well-delineated heterogeneous mass of subcutaneous tissue, predominantly hypoechoic, showing an internal hyperechoic area (**Figure 1**).

Magnetic resonance imaging (MRI) demonstrated an oval lobulated tumor with well-defined margins at the anterolateral part of the leg, located among the soleus, peroneus longus, extensor digitorum longus, and lateral gastrocnemius muscle bellies, following the track of common/superficial peroneal nerve, insinuating itself into the subcutaneous fat through the fibular tunnel, cranially. The lesion has a close relationship with the lateral cortex of the fibula, with heterogeneous paramagnetic contrast impregnation (**Figure 2**).

Surgery to remove the tumor was performed, and the anatomopathological evaluation revealed a neoplasm with an extensive chondromyxoid matrix. The immunohistochemical study demonstrated positive vimentin (V9 clone), positive S-100 protein and negative desmin (D33 clone), smooth muscle actin (SMA – 1A4), CD68 (KP1 clone) and caldesmon. The set of findings is compatible with myxoid liposarcoma (**Figure 3**).

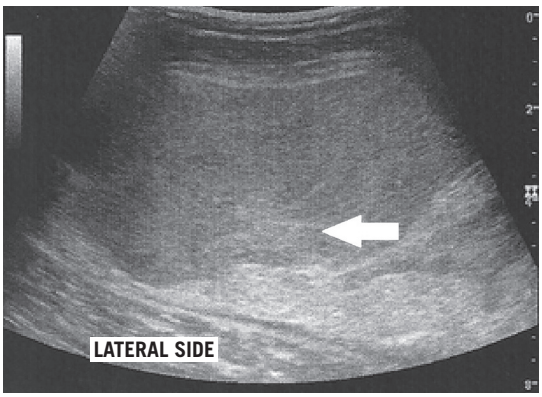

FIGURE 1 – *Ultrasonography demonstrating heterogeneous well-delineated mass, with hyperechogenic area inside (white arrow)*

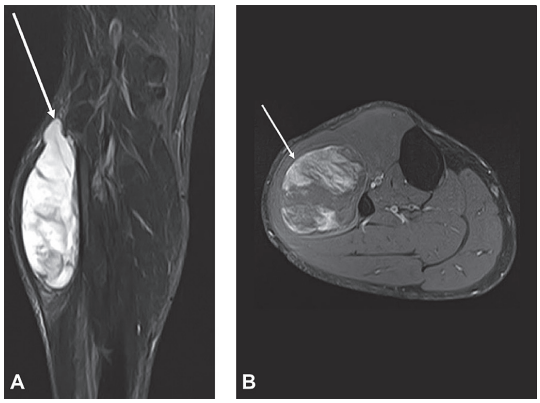

FIGURE 2 – *A) MRI in T2 STIR weighted image without contrast in the coronal section revealing a high signal tumor, well-delimited, with septations (white arrow); B) MRI in T1 FAT SAT weighted image with contrast in the axial section demonstrating heterogeneous enhancement of the lesion (white arrow)*

MRI: magnetic resonance imaging; STIR: short TI inversion recovery; FAT SAT: fat saturation.

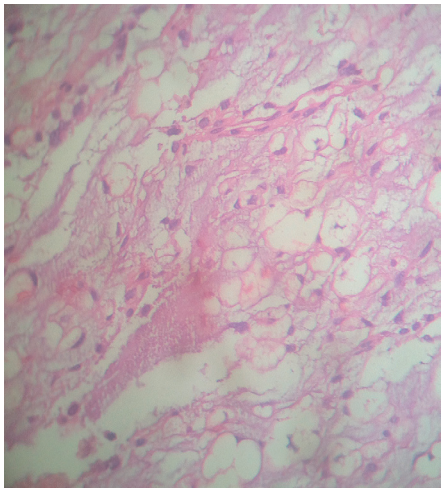

FIGURE 3 – *Photomicrograph (HE stain): presence of neoplasm with an extensive chondro-myxoid matrix*

HE: hematoxylin and eosin.

DISCUSSION

Ultrasonography (US) findings consist of detecting a cystic complex mass with Doppler vascularization[3]. Computed tomography (CT) scan without contrast shows a well-defined mass with a density similar to water, while the contrast CT scan shows progressive reticular enhancement[7]. Calcifications are an uncommon finding[3].

The MRI allows for adequate identification of specific histological liposarcoma subtypes[8]. It also proves to be viable and effective for detecting bone and soft tissue metastases[6]. The myxoid liposarcoma, in MRI, has low or isointense signal intensity to muscle on T1-weighted image and high signal on T2[9]. The tumor may look like a benign cystic tumor when an MRI is performed without contrast[10].

In the contrast study, enhancement areas inside the tumor represent increased cellularity and vascularity, while areas without contrast represent necrosis, reduced cellularity and accumulated mucinous material[10]. The presence of amorphous or linear hyperintense foci with adipose tissue linear aspect on T1-weighted images has been reported as a pattern suggestive of myxoid liposarcoma[2]. Accentuated contrast enhancement in MRI images is considered the most important adverse prognostic factor[2].

The high water content in myxoid liposarcoma seen in the histopathological analysis and constituting the major part of the lesion is reflected in US, CT scan and MRI[4]. However, the detection of a small amount of adipose tissue in the septum of the lesion or small nodular foci overlapping with myxoid tissue base allows the prospective diagnosis in 78%-95% of myxoid liposarcomas[4]. Histological analysis shows a myxoid matrix as the predominant component and small amounts of fat, which creates a typical appearance on MRI[5].

The treatment of choice for myxoid liposarcoma is extensive surgical excision[4]. In cases with incomplete resection due to lesion size or proximity to the neurovascular bundle, radiotherapy is often used to reduce local recurrence[4]. Adjuvant chemotherapy may also be beneficial[4].

Additional factors associated with worse prognosis are[4]:

- patients aged over 45 years;
- the histological presence of spontaneous necrosis.

The overall five-year survival rate ranges from 47% to 77%[4].

CONCLUSION

Imaging and surgical biopsy are of great value in establishing the most appropriate therapeutic program, highly adapted to the histopathological findings. However, the interpretation of the diagnostic imaging modalities requires huge experience and carries a risk of ignoring a critical part of malignancy. It is of fundamental importance to understand the principles of musculoskeletal oncology in the evaluation of any soft tissue mass that can be malignant.

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CORRESPONDING AUTHOR

Marcio Luís Duarte ID 0000-0002-7874-9332
e-mail: marcioluisduarte@gmail.com