

Myxoid liposarcoma with unusual clinical presentation: imaging diagnosis and histopathologic correlation. A case report

Liposarcoma mixoide de presentación clínica inusual: diagnóstico por imagen y correlación histopatológica. A propósito de un caso

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Summary

Myxoid liposarcoma is the second most frequent malignant neoplasm of all types of liposarcoma, usually located in the lower limbs and retroperitoneum. It is characterized as a slow-growing, painless soft tissue mass that develops especially in the intermuscular and rarely intramuscular compartments and, depending on its cellularity, can become highly metastatic. Imaging diagnosis and its histopathological correlation is essential to provide timely treatment; however, this can become a great challenge due to the lack of specificity of its findings in first line imaging studies that are generally available in hospitals up to second level. This makes the need for more advanced studies essential, such as magnetic resonance imaging (MRI), where the role of the radiologist is essential for the detection of subtle but pathognomonic findings of this pathology. We present the case of a young adult patient with an unusual presentation of a myxoid liposarcoma who presents with pain and mass at the level of the right buttock; her imaging studies are performed and analyzed and correlated with histopathological findings.

Resumen

El liposarcoma mixoide es la segunda neoplasia maligna más frecuente de todos los tipos de liposarcomas. Se localiza habitualmente en los miembros inferiores y el retroperitoneo. Se caracteriza por ser una masa de tejido blando, de crecimiento lento e indolora que se desarrolla especialmente en los compartimentos intermusculares y rara vez intramusculares y, dependiendo de su celularidad, puede llegar a ser altamente metastásica. El diagnóstico por imagen y su correlación histopatológica es fundamental para brindar un tratamiento oportuno; sin embargo, este puede convertirse en un gran desafío por la inespecificidad de sus hallazgos en estudios de imagen de primera línea —que son los que generalmente realizan en hospitales de hasta segundo nivel—. Esto vuelve primordial la necesidad de estudios más avanzados, como la resonancia magnética (RM) en la que el papel del radiólogo es imprescindible para la detección de hallazgos sutiles, pero patognomónicos de esta patología. Se presenta el caso de una paciente adulta joven con presentación inusual de un liposarcoma mixoide, quien cursa con un cuadro de dolor y masa a la altura del glúteo derecho; se practican y analizan sus estudios de imagen y se correlacionan con los hallazgos histopatológicos.

Introduction

Myxoid liposarcoma is a malignant neoplasm that occurs mainly in the lower limbs of young and middle-aged adults. It ranks second in frequency (30%-35%) of all types of liposarcomas (1) and belongs to the group of adipocytic tumors, according to the WHO classification of 2020 (2).

Its degree of cellularity predicts the possibility of metastasis; hence the importance of obtaining an adequate sample to quantify the cells. If they exceed 25%, the tumor should be considered high grade (2).

Metastases are most frequently located in serous membranes, abdominal cavity, soft tissues, bone and lung (2).

Clinically, it is characterized as a soft tissue mass with slow and subtle growth that could be confused with a benign lesion; however, any soft tissue lesion must be studied exhaustively radiologically and requires biopsy if it measures more than five centimeters in diameter (3).

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Clinical case

A 37-year-old female patient, with no significant history of trauma or pathology, allergic to iodine, came to the emergency department with pain in the right buttock, associated with a palpable mass of approximately three months of evolution, which intensified 72 hours before admission. She was diagnosed with cellulitis and treated; however, the lesion progressively increased in size in the last two weeks and the pain made ambulation difficult.

On physical examination, a fluctuant mass was palpable at the level of the upper third of the right buttock measuring approximately 15 cm.

Paraclinical tests showed no signs of inflammatory response, with results of acute phase reactants within normal parameters.

Soft tissue ultrasound identified an image of heterogeneous appearance, predominantly hypoechogenic, ovoid, with lobulated edges, internal septa, with diameters of 12.1 × 9.6 × 8.3 cm, volume of approximately 514 mL, avascular on color Doppler, which was classified as a probable hematoma (Figure 1).

A simple computed axial tomography (CT) scan of the pelvis with extension to the lower limbs showed, in the gluteal region, in the outer quadrants, an image suggestive of an intramuscular collection in the right gluteus maximus muscle, with diameters of 6.4 × 3.9 × 6.5 cm, estimated volume of 84 mL, with density of 8 HU in simple phase, per fluid. Contiguously, without defining an adequate cleavage plane with the previous lesion described, another organized collection was observed in the subcutaneous cellular tissue, which impressed with low density septa in the interior, measuring 10.9 × 11.8 × 6.9 cm, approximate volume of 461 mL (Figure 2).

The patient was evaluated by the surgery department, who requested simple and contrasted magnetic resonance imaging (MRI) of the pelvis, which showed an intramuscular lesion in the right gluteus maximus muscle, well circumscribed, measuring 13.9 × 9.1 × 12.6 cm, with extension into the subcutaneous cellular tissue, predominantly low signal with areas of high signal with respect to the muscles in T1-weighted sequences, very high signal in T2 with low signal internal septa and areas of restricted diffusion. Dynamic sequences after contrast medium administration showed progressive and heterogeneous enhancement. Additionally, mild edema of adjacent soft tissues. In view of the above, a myxoid lesion was suggested as the first diagnostic possibility (Figure 3).

After the findings obtained, the treating department decided to perform a core biopsy guided by ultrasound. The result showed connective tissue fragments with moderate scattered lymphocytic inflammatory infiltrate and congestive vascular structures; it was considered inconclusive. Excisional biopsy of the mass was performed and sent for histopathological analysis with a confirmatory diagnosis of myxoid liposarcoma.

Discussion

Myxoid liposarcomas are intermediate-high grade malignant neoplasms. They occupy the second place among liposarcomas in adults and the preferential locations of these tumors are the extremities and the retroperitoneum. Generally, they grow in the intermuscular compartment and not so frequently intramuscular. They are slow-growing tumors that characteristically present as a large, painless lump (4, 5).

Imaging diagnosis represents a challenge, since both ultrasound and CT findings are non-specific, so MRI could contribute better to the diagnosis. Ultrasonographically, this type of lesions appear as an avascular, complex, heterogeneous, predominantly hypoechogenic and well-defined mass with posterior acoustic reinforcement (6). In CT they are usually lesions of intermuscular location, multilobulated and well defined, in which the high water content gives them their characteristic low attenuation (6). In MRI, these lesions also appear as well-defined masses with heterogeneous signal in T1-weighted sequences. In T2, they look "pseudocystic", with high signal, with small fat foci both linear and punctiform, almost imperceptible within the tumor, as well as in its contours. These lesions can coexist with edema of the adjacent tissue that appears by compression or infiltration; however, it is not always present (7). The case documented here is an unusual presentation, in a young adult female patient who presented with pain and the appearance of a lump in the right buttock. Initially the lesion was classified as a possible hematoma due to its ultrasound characteristics and its rapid evolution in the last few days, so it was complemented with simple CT, which showed the liquid nature of the lesion. Since there was a clinical-radiological discordance, an MRI was performed which corroborated the lesion with an intramuscular component in the gluteus maximus and subcutaneous, of liquid content, multiseptated with low signal foci in fat suppression sequences which were not detailed in the first review and which represent pathognomonic findings of this lesion, which should be better analyzed in the face of a large lesion of liquid nature (4, 5).

It is important to remember that characteristics such as size, depth and density/intensity can guide the determination of benignity or malignancy of a lesion. As general features, large, deep and heterogeneous lesions are usually seen in malignant tumors, although some benign tumors can also present in such a way, which implies making histopathological correlation for its diagnosis (8).

Specifically, if certain characteristics are found in MRI, such as a tumor >10 cm, located in deep areas, irregular margins, without lobulations and with thick septa (>2 mm) it is much more likely to be associated with an intermediate or high grade liposarcoma (9).

Surgical resection continues to be the treatment of choice for this type of tumor, trying to maintain clean margins, since this will influence the patient's survival. Being very radiosensitive tumors, radiotherapy should be considered before and after surgical treatment. Finally, it is suggested to add chemotherapy, especially in tumors larger than 10 cm, without forgetting to perform consecutive controls, since these tumors tend to metastasize (3, 10).

Nevertheless, the treatment of these lesions is not only based on the characteristics of the tumor, but also takes into account factors such as size, age, location and sex, which influence the treatment protocol to be followed, as well as the patient's prognosis (10).

Finally, it has been observed that there is a better survival rate (96%) of patients at five years compared to other soft tissue sarcomas, with a disease-free rate of 76%, the latter considered a direct factor of clinical benefit, which shows a favorable prognosis for this type of sarcoma (11).

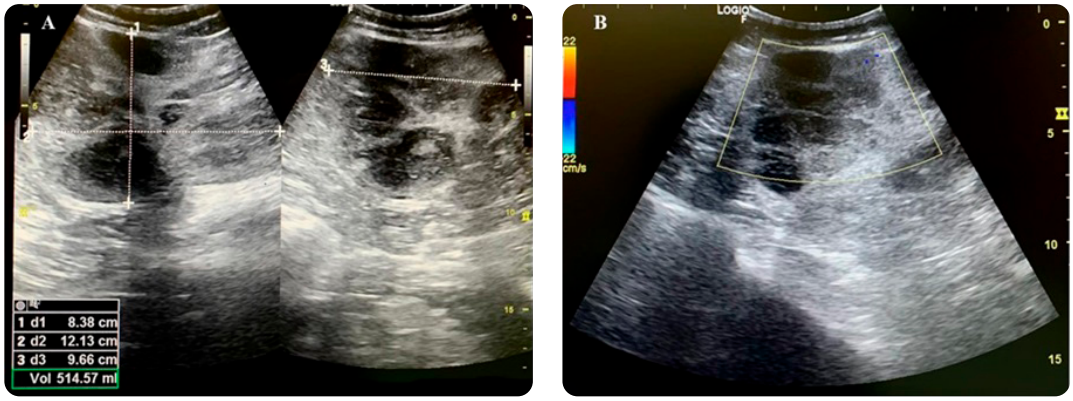


Figure 1. Ultrasound of soft tissues of the right gluteal region. a) Lesion of 12.1 × 9.6 × 8.3 cm for an approximate volume of 514 mL of mixed content, characterized by hypoechoic and anechoic areas and septa. b) Avascular lesion on color Doppler.

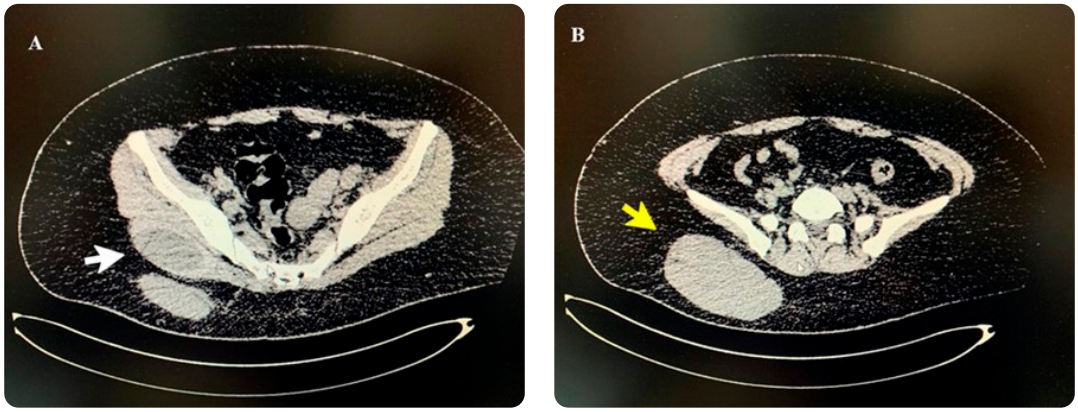


Figure 2. Simple CT of pelvis in axial section. a) Intramuscular lesion of the right gluteal muscle with density of 8 HU, in relation to liquid. b) Organized collection in the subcutaneous cellular tissue that appears to have dense septa in the interior.

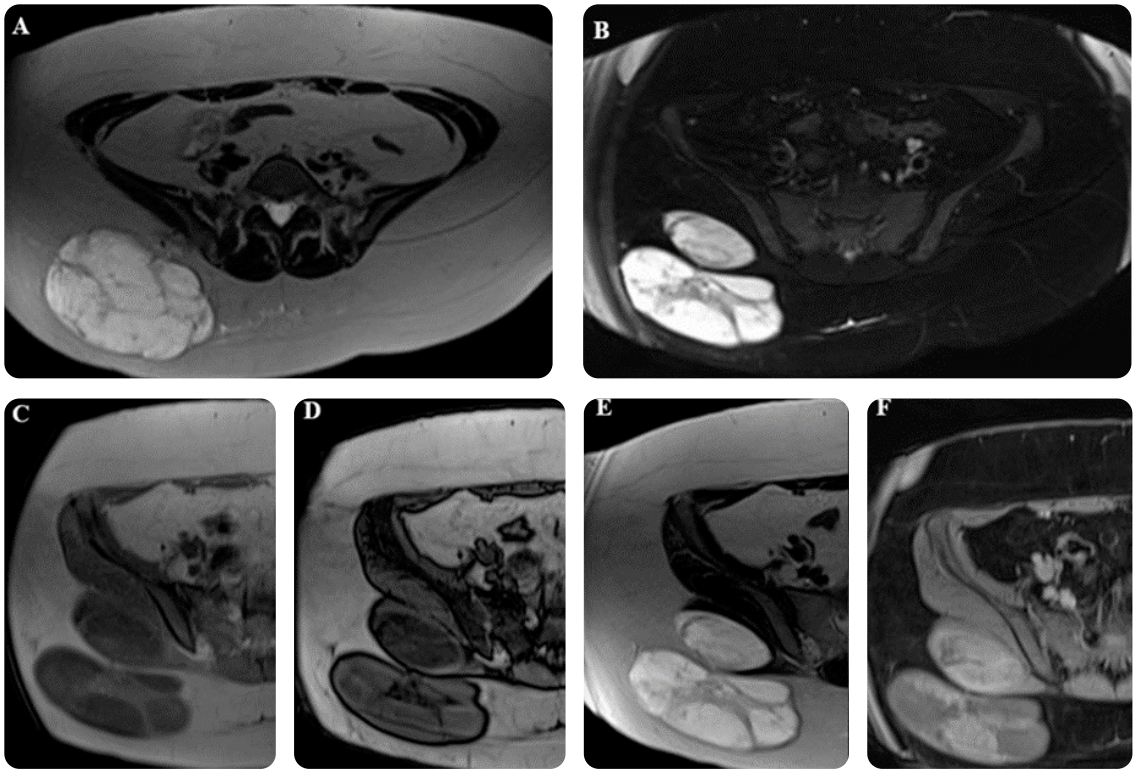


Figure 3. Simple and contrasted MRI in axial sections. a) T2-weighted view, high signal lesion located in the subcutaneous cellular tissue, polylobulated with septa. b) T2 FAT-SAT. Intramuscular component of the lesion. c and d) Axial slices in DIXON in phase and out of phase, in which linear images with signal drop corresponding to fat content are perceptible. e) T2-weighted sequences at the same level. Linear images of similar intensity to subcutaneous fat. f) T1-weighted sequences FAT-SAT with gadolinium. Heterogeneous enhancement of the contrast medium.

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