

Myxoid Liposarcoma: When the Diagnosis is Not Obvious: Case Report and Literature Review

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Abstract

Sarcomas are a rare and heterogeneous group of malignant tumors of mesenchymal origin that comprise less than 1% of all adult malignancies and 12% of pediatric cancers. Liposarcoma is defined as a malignant mesenchymal neoplasm that is composed of lipogenic tissue, and is a common malignant soft tissue tumor, accounting for 10% to 16% of all sarcomas. This case is about a 47-year-old male, who visited his family doctor with complaints of asymmetrically enlarged left thigh mass with a week of evolution, associated with light left thigh pain that appeared 1 month before presentation. Physical examination showed a difference of 11.5 centimeters (cm) in the measurement between both thighs. An ultrasound was requested and showed a massive neof ormation on the posterior aspect of the left thigh, compatible with liposarcoma. The patient was referred to the General Surgery Service where he performed magnetic resonance imaging (MRI), which confirmed the presence of bulky neof ormation, compatible with undifferentiated liposarcoma. As part of the protocol, the patient was then referred to the Portuguese Oncology Institute in Oporto, where he performed genetic tests and therapeutic guidance. Soft tissue sarcomas are a heterogeneous group of tumors with a large spread in biological behavior, prognosis, and requested treatment modalities. Myxoid liposarcoma show a good prognosis in most cases. However, it is biologically different from other liposarcomas, with the presence of the t (12;16) translocation, high radio and chemosensitivity and a high prevalence of extrapulmonary metastases.

Keywords: Sarcoma; Myxoid liposarcoma; Family medicine

Introduction

Sarcomas are a rare and heterogeneous group of malignant tumors of mesenchymal origin that comprise less than 1% of all adult malignancies and 12% of pediatric cancers [1-3]. Approximately 80% of sarcomas originate from soft tissue, being the main complaint a gradually enlarging and painless mass [3]. The World Health Organization (WHO) classifies most soft tissue neoplasms according to the presumptive tissue of origin [2].

Liposarcoma is defined as a malignant mesenchymal neoplasm that is composed of lipogenic tissue, and is a common malignant soft tissue tumor, accounting for 10% to 16% of all sarcomas. It typically affects patients between the fifth and seventh decade of life, and usually develops in the extremities or retroperitoneum [4]. According to WHO classification of soft tissue tumor and bone (2013) there are four liposarcoma subtypes: atypical lipomatous tumor/well-differentiated, dedifferentiated, myxoid/round cell and pleomorphic liposarcomas [5]. There is a great range of biologic behavior amongst these subtypes, spanning from well-differentiated liposarcomas with low metastatic potential to the high-risk round cell or pleomorphic types, which tend to be higher grade and are associated with a high rate of distant metastases [6].

Myxoid liposarcoma represents approximately 5% of all soft tissue sarcomas and about one third to one half of all liposarcomas [5,7,8]. It primarily affects younger adults, with a peak incidence during the fifth decade [9]. Myxoid liposarcoma is a clinicopathologically and genetically distinct subtype, characterized by its classical involvement of the deep soft tissue of the lower extremity (75%), especially the medial thigh and popliteal region, and the presence of the t (12;16) translocation [10].

Methods

Interview and family evaluation with the patient. Consultation of the clinical process of the hospital.

Case Report

Personal background

A 47-year-old male, divorced, house-builder. Duvall cycle not applicable, low class according to Graffar scale, Apgar score corresponding to medium dysfunctional family.

1. Pathological background: obesity, peripheral venous insufficiency and dyslipidemia.
2. Surgical background: irrelevant.
3. No medicine or food allergies known.
4. Up-to-date vaccination.
5. Denied smoking, alcohol or drug abuse.
6. No risky sexual behavior

Description

Presented on February 2017 for evaluation of asymmetrically enlarged of left thigh mass with a week of evolution. The patient had noticed the asymmetry himself and report light left thigh pain 1 month before presentation. His mobility was not limited by pain, but he began to have difficulty on wearing the pants and folding the referred limb in the workplace. He reported a minor trauma in that area weeks earlier.

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Received January 08, 2018; Accepted February 09, 2018; Published February 15, 2018

Citation: Marcalo SC, Vilarinho T, Oliveira M (2018) Myxoid Liposarcoma: When the Diagnosis is Not Obvious: Case Report and Literature Review. J Clin Case Rep 8: 1080. doi: 10.4172/2165-7920.10001080

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The physical examination showed a difference of 11.5 centimeters (cm) in the measurement between both thighs (Figure 1).

Considering the pathological background of obesity and peripheral venous insufficiency, the hypothesis of differential diagnosis with a venous thrombosis was placed, an ultrasound was requested. However, it showed a massive neof ormation on the posterior aspect of the left thigh, compatible with liposarcoma.

The patient was referred to the General Surgery Service where he performed magnetic resonance imaging (MRI), which confirmed



Figure 1: Difference between both thighs of patient on the first evaluation.



Figure 2: MRI assessment using coronal T2 without fat suppression.



Figure 3: MRI assessment using axial T2 without fat suppression.



Figure 4: MRI assessment using axial T1 without fat suppression.

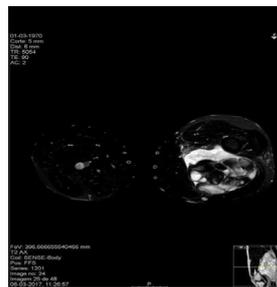


Figure 5: MRI assessment using axial T2 with fat suppression.



Figure 6: MRI assessment using sagittal T2 without fat suppression.



Figure 7: Ultrasound image of the lesion.

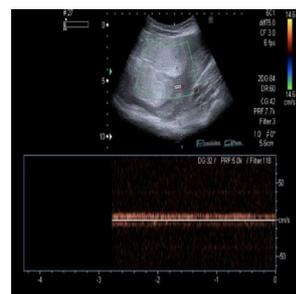


Figure 8: Ultrasound image showing a vascular lesion.

the presence of a bulky neof ormation with well-defined contours but with marked tissue heterogeneity with fat component, in the posterior compartment (distal third) of the left thigh, measuring 18 cm of longitudinal diameter, 11 cm of transverse axis and 9 cm of anteroposterior diameter, compatible with undifferentiated liposarcoma. No other locoregional structural changes were observed (Figures 2-8).

As part of the protocol, the patient was then referred to the Portuguese Oncology Institute in Oporto, where he performed genetic

tests and therapeutic guidance. Regarding to the genetic tests, the amplification of the *MDM2* oncogene, which is characteristic of well differentiated liposarcoma, has not been demonstrated. However, a translocation involving the *FUS* gene (16p11) was observed.

Discussion

The differential diagnosis of a soft tissue mass includes benign soft tissue tumors, such as a lipoma, as well as malignant tumors [2]. Thus, imaging is a crucial tool in the evaluation of a lipomatous mass. Usually, characterization with computed tomography (CT) and MRI is sufficient to allow the distinction between a lipoma and liposarcoma [11].

Given that benign soft tissue masses are at least hundred times more common than malignant soft tissue sarcomas, it can be difficult to determine which soft tissue masses warrant further evaluation [2]. Knowing this, the United Kingdom Department of Health has published criteria for urgent referral of a patient with a soft tissue lesion [12]:

1. Soft tissue mass >5 cm;
2. Painful lump;
3. Lump that is increasing in size;
4. A lump of any size that is deep to the muscle fascia;
5. Recurrence of a lump after previous excision.

Soft tissue sarcomas most commonly present as an enlarging, painless mass in the extremities or trunk [13]. The presence of distant metastatic disease at the time of initial diagnosis is uncommon but more likely in large, deep, high-grade sarcomas [4]. In the particular case of myxoid liposarcoma, proximately 80% of metastases are located in the lungs, but retroperitoneum, mesentery, bone and soft tissue of the trunk are other regions for metastasis [4]. This subtype of liposarcoma the primarily treatment includes complete surgical excision as there is a high concordance between a negative margin status, local recurrence and disease-specific survival [14]. Neoadjuvant or postoperative radiation therapy has been used very successfully in this tumor [15]. Chemotherapy is usually reserved for patients with metastatic, locally advanced and/or unresectable disease [14].

In this particular case, the genetic tests were negative for the t (12;16) and patient showed no metastatic disease. After decision of cancer group consultation, the patient was submitted to surgery for extensive excision of the neof ormation, with preservation of the sciatic nerve, on July 2017. Histological examination revealed a myxoid liposarcoma.

The patient then underwent radiotherapy treatments, which ended in November 2017. He is currently under chemotherapy, showing a favorable evolution.

Conclusion

In conclusion, soft tissue sarcomas are a heterogeneous group

of tumors with a large spread in biological behavior, prognosis, and requested treatment modalities. Myxoid liposarcoma show a good prognosis in most cases. However, clinicians and pathologists should be aware because myxoid liposarcoma is biologically different from other liposarcomas, with the presence of the t (12;16) translocation, high radio and chemosensitivity and a high prevalence of extrapulmonary metastases.

Acknowledgement

The authors would like to thank the colleagues of the Portuguese Institute of Oncology in Oporto, for their collaboration in providing the images and clinical information about this case, as well as for all the diagnostic and therapeutic follow-up of the patient described.

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