

An Unusual Case of Giant Myxoid and Round Cell Liposarcoma of the Thigh: Case Report and Literature Review

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Case Report

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Abstract: Liposarcomas represent the second most frequent soft tissue sarcoma in adults. Their diagnosis can be delayed because of the silent evolution of those malignant tumors, and their nonspecific symptoms make them even more difficult to clearly identify. Doctors should think of this pathology in front of asymptomatic and painless mass of rapid growth, so that they can provide effective care, avoid non conservative surgery and optimize the prognosis. In this report, we describe an unusual case of a giant myxoid and round cell liposarcoma of the thigh that benefited from conservative surgery with good evolution, with a review of literature.

Key words: Liposarcoma, myxoid, round cell, giant, conservative surgery.

INTRODUCTION

Liposarcoma is a malignant soft tissue tumor that takes origin in primitive mesenchymal cells [1, 2]. It is the second most common sarcoma of deep soft tissues in adults, right after malignant fibrous histiocytoma [3, 4]. The preferential sites of those tumors are the deep soft tissues of the extremities [3].

The diversity of histological types of liposarcomas implies numerous variations of clinical, biological and radiologic expressions and the assessment of all these aspects is crucial to conduct appropriate diagnosis and therapy [3].

That being said, they often remain asymptomatic and the diagnosis is thus frequently belated [5], making their therapeutic care even more challenging.

In the following case, we report the aspects of an unusual giant myxoid and round cell liposarcoma of the thigh that evolved well after conservative surgery.

CASE REPORT

A 35-year-old male patient with no history of substantial pathology presented to us with a painless voluminous mass of the posterior surface of the right thigh, evolving for 18 months in an afebrile context and conservation of his general condition. There was no

reported episode of trauma incident or physical injury prior to the appearance and development of the swelling.

Clinical examination revealed a mass of about 35 centimeters of major axis, invading the entire posterior region of the thigh. This mass was soft, fixed relative to superficial and deep plans, with no sign of cutaneous reaction (no ulcerations and no fistula) and no motor or sensitive deficit [figures 1,2 and 3].



Fig-1



Fig-2



Fig-3

Fig-1, 2 and 3: Clinical aspect of a voluminous mass of the posterior region of the thigh

We realized an MRI that showed a voluminous tumor formation occupying the posterior lodge of the thigh, with lobulated appearance and lipomatous signal.

The mass was in contact with the posterior femoral cortical with no sign of infiltration or invasion of the adjacent structures [figures 4,5,6,7,8].



Fig-4

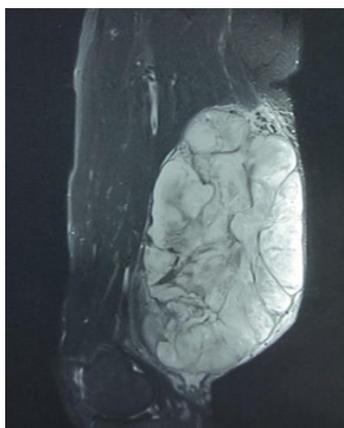


Fig-5



Fig-6

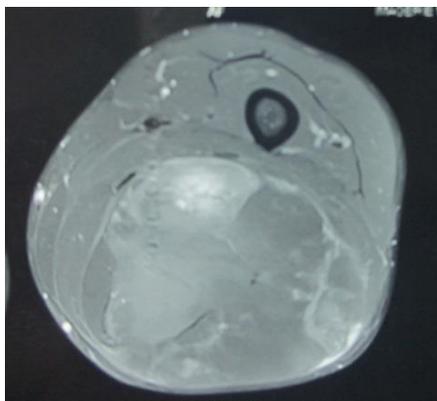


Fig-7

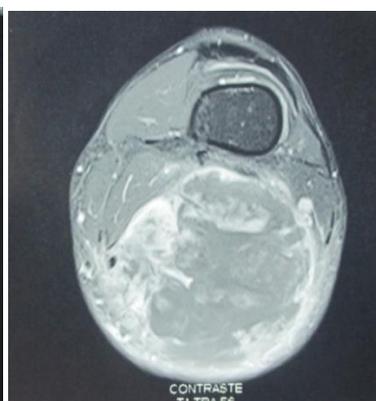


Fig-8

Fig-4, 5, 6, 7, 8: MRI images of the tumoral formation

The examination regarding tumor extension- that included an abdominal echography, a C.A.P CT (thoraco-abdomino-pelvic computed tomography) and a bone scintigraphy- revealed no secondary localization. Biological laboratory tests showed no abnormalities.

Histopathological examination found multinodular masses composed of a myxoid matrix with plexiform capillary vasculatures. Stellate and round cells were noticed with cytoplasmic atypia and some figures of mitosis. Atypical proliferating lipoblasts with variable degrees of proliferation were also present [figures 9,10,11].



Fig-9: Image showing the skin scar of the biopsy

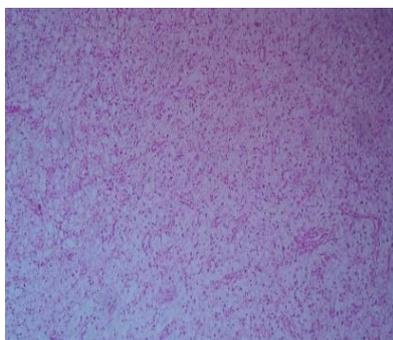


Fig-10

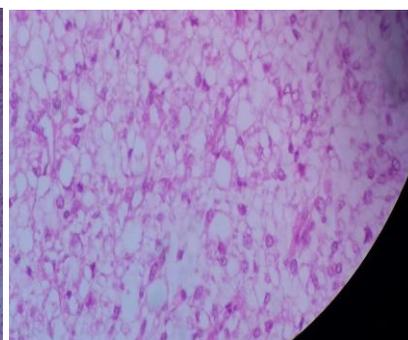


Fig-11

Fig-10, 11: histological aspect of the tumor

We concluded to a myxoid and round cell liposarcoma. The case was discussed in a multidisciplinary meeting, and the patient has benefited from a marginal excision of the tumor in healthy

boundaries [figures 12,13,14]. The mass measured 32x25 centimeters and weighted 4500 grams [figure 15].



Fig-12



Fig-13



Fig-14

Fig-12, 13, 14: Images of the complete surgical excision of the mass



Fig-15: Intraoperative macroscopic aspect of the resected tumor

Surgery was followed by local radiation therapy. We obtained good evolution with no functional

impairment and no recurrence after five months of the excision [figures 16, 17, 18].



Fig-16



Fig-17



Fig-18

Fig-16, 17, 18: Images of good clinical evolution after 5 months of surgical resection

DISCUSSION

Liposarcoma represents 10 to 35% of all soft tissue sarcomas [6], it mostly affects middle-aged adults with a peak from 40 to 60 years old with a slight male predominance [3].

The nomination of liposarcoma is due to its lipomatous differentiation. It does not signify that the tumor takes origin from the fat [3]. The World Health Organization (WHO) first classified it as five histologic subtypes: well-differentiated, dedifferentiated, myxoid, pleomorphic, and mixed type [2, 4, 7]. Then, a new

classification divided liposarcomas into three types: myxoid /round cell (1), well-differentiated / dedifferentiated (2), and pleomorphic (3) [2, 8, 9].

The myxoid type is the second most frequent one after well-differentiated type, with a rate of 20 to 50% of all liposarcomas [3, 4, 8]. Round cell liposarcoma commonly coexists with myxoid one. They have the same clinical and morphological properties, and tumors integrating both of the types are very frequent, so well that they were merged under a single category in the latest WHO classification [2].

Clinically, telltale signs are not specific [5]. Often remaining asymptomatic for a long time [5], it usually reveals as voluminous painless soft tissue mass, occurring in the thigh in more than two thirds of the times, and measuring up to 19,5 centimeters [2,10]. In our case, we qualified the mass as “giant” since we measured it 32x25 cm after surgical excision.

MRI can be very specific. It exhibits the rate of lipomatous constituent and shows peculiar images that could lead to identify the tumor's nature and its histologic type, even before realizing the biopsy [3, 8]. In the case of myxoid liposarcoma, MRI shows a mass of high signal intensity on T2-weighted images, with lipomatous component producing a characteristic appearance of marbled and little heterogeneous textural pattern on T1-weighted sequences [3]. The diagnosis is then strongly suggested, but myxoid liposarcoma can still be confused with other entities as cystic masses [3, 4, 8, 11]. On the other hand, myxoid liposarcoma with round cells has a less specific appearance as the lipomatous constituent is less present, making it more difficult to distinguish from other solid tissue masses [3]. Beyond its role in the diagnosis, MRI enables us to assess locoregional extension of the tumor, along with CT scan, abdominal ultrasound and scintigraphy [5].

Histopathologically speaking, myxoid liposarcomas are composed by a rich myxoid stroma of small lobulated cells with different degrees of differentiation, associated to a vascular plexiform network. Round cell types contain primitive round or oval cells contained in a lean myxoid background [2, 3, 5], and usually coexist with myxoid variants [2].

The treatment of localized liposarcomas, regardless the type, consists of a surgical excision associated to adjuvant local radiation therapy [12]. Chemotherapy has little interest [5]. Healthy resection margins-that should be confirmed on histopathological examination- are a primordial condition for complete recovery [3, 5]. Postoperative radiotherapy helps preventing the recurrence of the tumor [13], knowing that myxoid liposarcoma is the most radiosensitive type [5].

The evolution depends of the type of liposarcoma, and the myxoid one is considered as the major factor for poor prognosis [14]. Other factors also known for conditioning the evolution, such as the advanced age of the patient, the precocity of diagnosis and treatment, the quality of the resection and the existence of secondary localization [2,5,14].

CONCLUSION

Diagnosis of liposarcomas can be challenging due to their usual asymptomatic evolution and unspecific revealing signs. It is essentially based on MRI imaging and histologic data. Proper therapeutic

management relies on adequate surgery and local radiotherapy. If well conducted, total recovery could be obtained.

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