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Superficial Myxoid Liposarcoma of the Thigh

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Abstract

Myxoid liposarcoma is the most common form of myxoid sarcoma. Their primary and superficial localizations are very rare compared to the deeper forms. We report the case of myxoid liposarcoma in a 57-year-old patient. The clinical picture marked by painless swelling of the right thigh had evolved over a period of about ten years. The diagnosis was guided by ultrasound and magnetic resonance imaging. A pathological examination of the surgical specimen after removal of the tumor helped to clarify the diagnosis. We discuss the clinical presentation and therapeutic management.

Keywords

Liposarcoma, Myxoid, Superficial Thigh, Surgery

1. Introduction

Tumors of fat tissue are the most common soft tissue tumors. Liposarcoma account for 6% of these tumors, half of which are myxoid liposarcoma, formerly known as round cell liposarcoma. They constitute a common spectrum of tumours, ranging from pure myxoid liposarcoma with good differentiation to tumors with a high proportion of round cells, which are dedifferentiated [1]. Myxoid liposarcoma come in two forms, superficial and deep, but the latter has a worse prognosis. Myxoid liposarcoma preferentially occurs in the lower limbs. The commonly accepted negative prognostic factors are age (>45 years), size (>10 cm), sub aponeurotic location, a high grade of sub fascial location, high histological grade, high round cell count and positive resection margins [2] [3] [4] [5] [6]. Magnetic resonance imaging (MRI) is an essential diagnostic tool for these tumours. Although there is a pathognomonic image, the diagnosis can be difficult if there is a high round cell component [3]. The aim of this study is to present

a case of superficial liposarcoma and to discuss its management.

2. Observation

A 57-year-old patient without pathological medical history consults for a painless swelling of the right thigh that had been progressing for about ten years. Physical examination revealed a voluminous mass of approximately 21 × 24 cm, pedunculated of firm consistency on the upper and inner surface of the right thigh, mobile with respect to both planes with healthy-looking skin, without collateral venous circulation or satellite adenopathy (Figure 1).

Standard radiographs showed osteoarticular integrity and ultrasonography of the thigh, a heterogeneous solid mass with hypo-echogene and hyper-echogene range. Magnetic resonance imaging revealed a solid, heterogeneous tumor mass, heterogeneous, with superficial topography and fat foci with T1 and T2 sequences, involving the upper-medial third of the right thigh, adjacent to the gracilis muscle without bone extension (Figures 2-4).

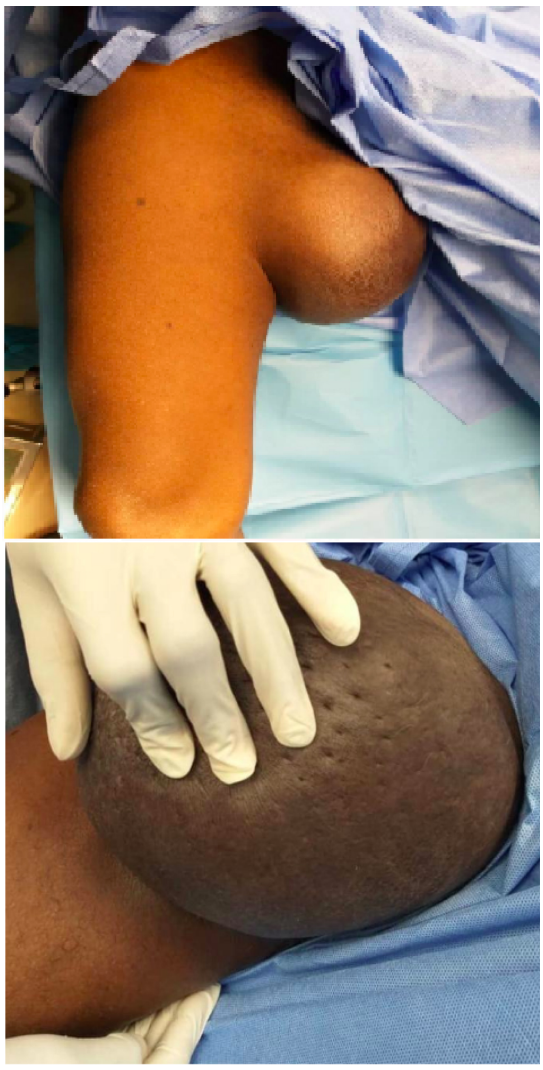

Figure 1. Clinical aspect of the tumor.

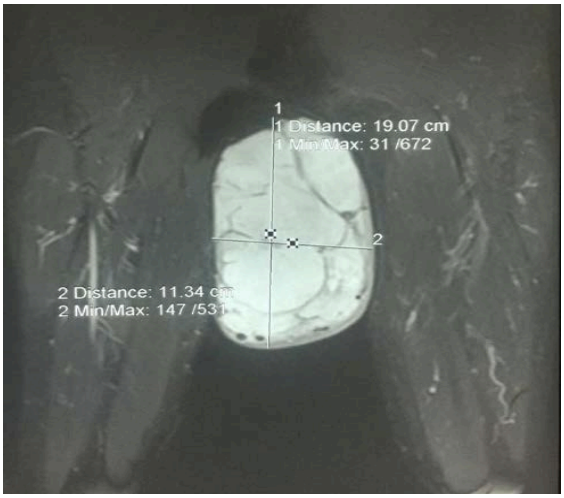

Figure 2. MRI coronal section in T1gado sequence.

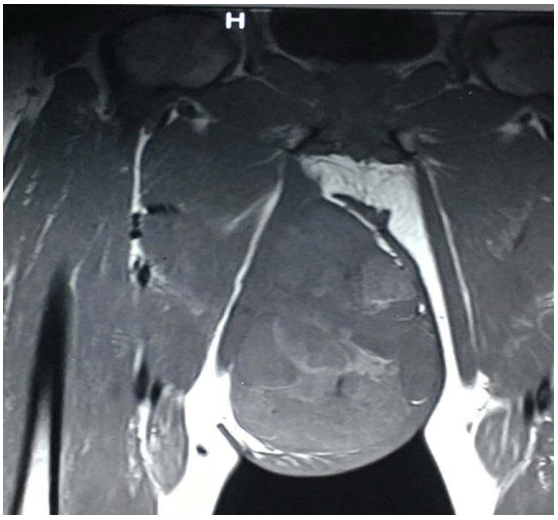

Figure 3. MRI coronal section in T1 sequence fat sat removal.

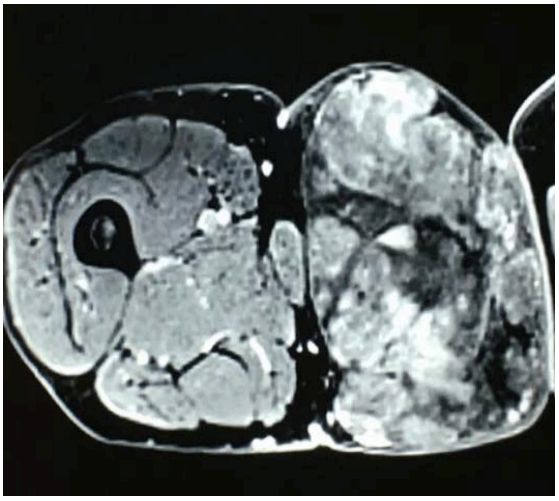

Figure 4. MRI axial section in injected T1.

No secondary localization of the tumor extension was found.

Complete excision was indicated and performed (Figures 5-7). The cleavage plane was subcutaneous without muscle invasion. Skin closure was performed by arciform plasty. Post-operative follow-up was favorable.

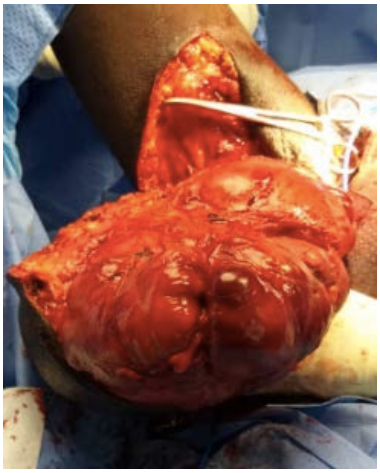

Figure 5. Image of intraoperative exeresis.

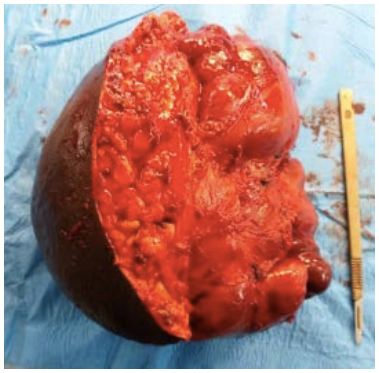

Figure 6. Appearance of the tumor after resection.

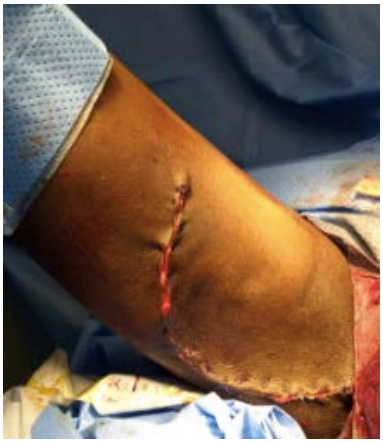

Figure 7. Closure of the skin plane.

On anatomopathological, macroscopic examination showed a mass weighing 2700 gr, measuring 27 × 26 × 12 cm, covered by a skin flap measuring 29 × 20 cm, greyish, firm, with an irregular surface, macro-nodular, well limited. Microscopic examination identified a sarcomatous tumor proliferation consisting of round, spindle-shaped cells whose more or less abundant cytoplasm, sometimes basophilic, contains an optically empty vacuole conferring adipocyte differentiation. The nuclei are rounded and ovoid, irregular, vesicular, nucleolated with moderate anisokaryosis and fairly numerous mitoses. The stroma is marked by a predominantly myxoid aspect with pseudocystic lymphatic dilatations. The tumor is developed in the dermis of the skin, delimited by pseudo-capsular fibrosis which appears to be intact.

After a 15-month follow-up, the patient showed no evidence of recurrence at the locoregional level.

3. Discussion

Myxoid liposarcoma is classified as soft tissue malignancies of adipocyte origin; they account for about 50% of all liposarcoma and are therefore relatively common [2] [3]. They occur most often between the ages of 40 and 50 years with male predominance [2] [5] [6].

Liposarcoma is preferentially located in the deep intramuscular regions below the superficial fascia, in the thigh or hollow popliteal [2] [4] [5] [6]; they are sometimes present in the retroperitoneum. However, in rare cases, they may be superficial just under the skin [4] [5]. This was observed in our 57-year-old patient whose tumor developed in the dermis. Moreover, this superficial character could explain the rare pedunculated aspect of the tumor in our case [5].

These tumors are revealed in more than 90% of patients by a large painless mass [3]. In most cases, as in our patient, superficial myxoid liposarcoma is painless; however, they represent the most aggressive histologic profile with potential for soft tissue metastasis of the bone before reaching the lungs, which is not the case in our patient after a 15-month follow-up [4] [5]. The use of traditional medicine still remains a diagnostic and therapeutic delay factor in our environment [4] [5]. The delay was ten years in our case.

From a diagnostic point of view, the ultrasound performed as first line is an accessible, inexpensive, non-invasive but operator-dependent examination. It makes it possible to orient the diagnosis and to specify whether the tumoris superficial or deep, vascular or not [3]. MRI, gold standard, performed as a second line of treatment, shows a heterogeneous tumor with a fat component in hypersignal T1 and hyposignal T2; the myxoid component is suspected in front of a very intense signal in T2 [2] [4] [7] [8]. It is relatively typical of myxoid sarcomas and allows simultaneous assessment of regional and locoregional extension. The pathological anatomy of liposarcoma corresponds to a mesenchymal malignant tumor corresponding to a proliferation of atypical adipocytic cells which may resemble mature adipocytes but are most often made up of immature adi-

pocytes called lipoblasts [5] and relatively small ovoid cells arranged in a plentiful myxoid matrix with a sophisticated capillary network sometimes reminiscent of a fence [9].

Therapeutically, the management of this tumor consists of complete tumor resection with safety margins greater than or equal to 1cm, followed by adjuvant or even neo-adjuvant radiotherapy [9]. The latter is described and recommended in the deep forms, but its interest remains controversial in the superficial forms [4] [10]. In our patient, we performed a complete resection of the tumor with a safety margin of 1 cm without adjuvant radiotherapy. Despite the good prognosis of these tumors, a close follow-up will be carried out with a survival rate of about 90% at 5 years [10].

4. Conclusion

Myxoid liposarcoma is the most common form of tissue sarcoma. However, its superficial form is rare. This localization could explain its pedunculated aspect and its less aggressive character, contrary to the deep form. Our observation shows that when faced with an indolent mass of soft tissue in the pedunculated and superficial thigh in an adult, one should think of a possibly myxoid liposarcoma and confirm it by histological evidence. Its treatment is resolutely surgical; the adjuvant treatment being specified only in deep forms with a poorer prognosis.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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