G Chir Vol. 40 - n. 6 - pp. 526-529
November-December 2019

Coexistance of a well differentiated liposarcoma and dedifferentiated low grade myxofibrosarcoma of the scrotum. A rare case and review of the literature

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SUMMARY: **Coexistance of a well differentiated liposarcoma and dedifferentiated low grade myxofibrosarcoma of the scrotum. A rare case and review of the literature.**

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Liposarcoma is a malignant soft tissue sarcoma usually located in the thigh or the posterior peritoneum in an adult. However, the occurrence of liposarcoma, and indeed a coexistance with scrotal myxofibrosarcoma, is rare. We present an interesting case of a 56-year old male who presented with an inguinal hernia. During the operation a massive fibro-elastic mass located within the left scrotum was noticed which deposited the testicle upward by displacing it into a sack rather than the penis. The mass did not come into contact with the spermatic cord but alongside it the existence of the blood vessels of the mass was found. The histopathological examination revealed a well differentiated liposarcoma along with dedifferentiated low grade myxofibrosarcoma.

KEY WORDS: Liposarcoma - Myxofibrosarcoma - Hernia.

Introduction

Liposarcoma is a malignant soft tissue sarcoma usually located in the thigh or the posterior peritoneum in an adult. Sarcomas of genitourinary region represent approximately 2% of soft tissue sarcomas (1). Almost 3% of malignant lesions of the spermatic cord are liposarcomas (6). Histological subtypes of malignant tumors are the paratesticular liposarcoma (20-56%), leiomyosarcoma (19-32%) and rhabdomyosarcoma (11-24%) (1, 3, 6). These tumors are frequently misdiagnosed as benign tumors (most commonly being interpreted as lipomas of the spermatic cord). Regarding histology, paratesticular liposarcomas are divided into four subtypes: well-differentiated, myxoid or round cell, pleomorphic and dedifferentiated. Paratesticular liposarcomas originate in mesenchymal tissue, and because of the slow growth have a low incidence and therefore clinical trials have not yet established proper treatment so far. Previous retrospective studies have suggested that adjuvant radiation therapy improves local control (6). The occurrence of liposarcoma, and indeed a coexistance with scrotal myxofibrosarcoma, is rare. The first reported case was in 1845 and only 100 cases are found in the literature (2).

Case report

A 56-year old male proceeded to our department complaining about left scrotal pain. The clinical examination revealed scrotal hernia. The patient underwent hernioplasty using the patch and plug technique. During the operation a massive fibro-elastic mass located within the left scrotum was noticed which deposited the testicle upward by displacing it

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into a sack rather than the penis. The mass did not come into contact with the spermatic cord but alongside it the existence of the blood vessels of the mass was found (Figures 1, 2).

The histopathological examination revealed a well differentiated liposarcoma along with dedifferentiated low grade myxofibrosarcoma. Grossly, tumor had a bilobated configuration formed by two uneven in size well circumscribed nodules, the biggest having a median diameter of 14 cm and the smaller one, 6 cm. The cut surface of both nodules flauntedan extensive gelatinous consistency alternating with firm white areas and foci with lipomatous appearance. Histologically, the tumor was characterized by the presence of two components, one liposarcomatous, consisting in a well differentiated, spindle cell and sclerosing, liposarcoma the former with extensive areas of mucinous degeneration and a dedifferentiated component, which demonstrated low grade myxofibrosarcomatous morphology, exhibited low cellularity and was portrayed by the presence of minimally pleomorphic spindle or stellate shaped cells, with slightly eosinophilic cytoplasm, hyperchromatic nuclei and only rare mitotic figures, arranged in fascicles or haphazardly in a myxoid matrix and frequently aligned along the elongated curvilinear capillaries of the stroma. Furthermore, sporadic fibroblasts showed cytoplasmic vacuolations {Alcian Blue (+)}, assuming a "pseudolipoblastic" appearance (Figure 3).

Tumor markers (CEA, Ca 19.9, aFP) were negative. Computed tomography of the Abdominal Cavity and the Thorax revealed no secondary metastasis. The patient underwent reoperation and the left testicle was removed as of oncological route.

The patient one year after the operation is asymptomatic.

Discussion

Liposarcoma is the most common type of soft tissue sarcoma (7). It can be divided into five histological subtypes from highest to lowest incidence: well-differentiated, dedifferentiated, myxoid, round cell, and pleomorphic (5, 6, 8). The most common of these are the well-differentiated tumors, which are

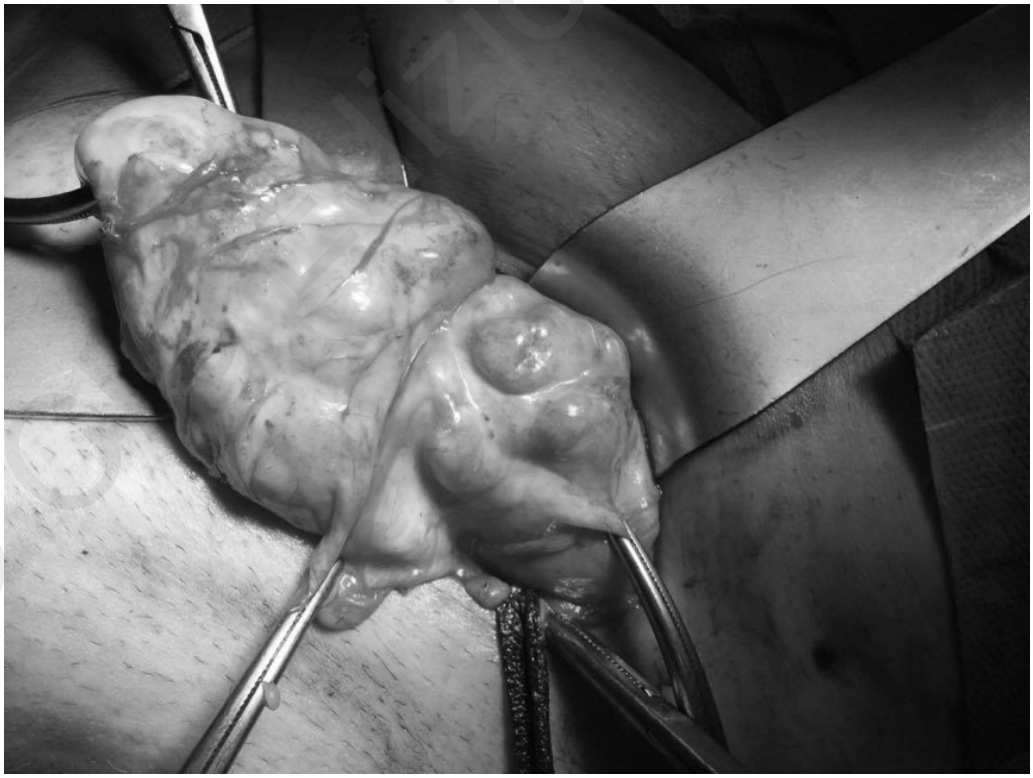

Figure 1 - Image during the operation.

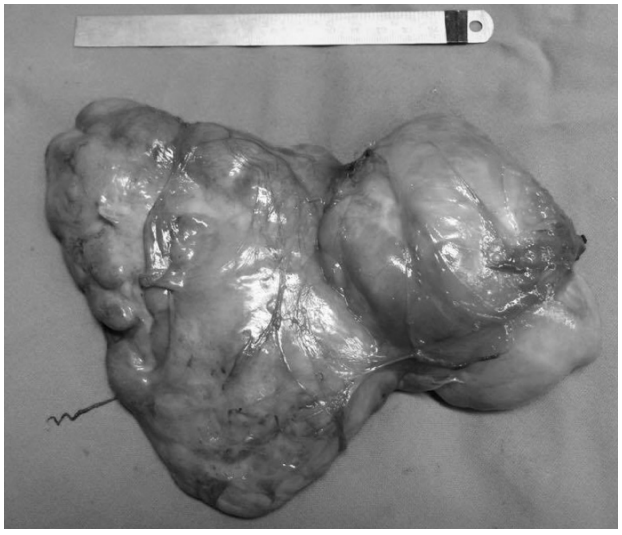

Figure 2 - Surgical specimen.

low grade tumors, followed by dedifferentiated tumors, which are higher grade tumors (9, 10). A patient typically presents with pain due to compression or invasion of the anatomical structures in the thigh or retroperitoneum (9). Not rarely, it is misdiagnosed as inguinal hernia.

Due to the anatomical location in which the liposarcoma of the spermatic cord presents and the appearance of the mass as fatty tissue on imaging studies, the diagnosis is difficult and tricky (8). Unlike germ cell tumors, ultrasound can have difficulty in distinguishing well-differentiated liposarcomas from adipose tissue. CT or MRI can be used with equal utility for retroperitoneal soft tissue sarcomas although MRI with gadolinium enhancement is the optimal imaging choice because it has demonstrated ability in extremities to better identify viable tumor tissue versus surrounding reactive tissues (8, 11). In comparison a liposarcoma of the spermatic cord may be misdiagnosed as an inguinal hernia due to its fatty composition with MRI or CT demonstrating a

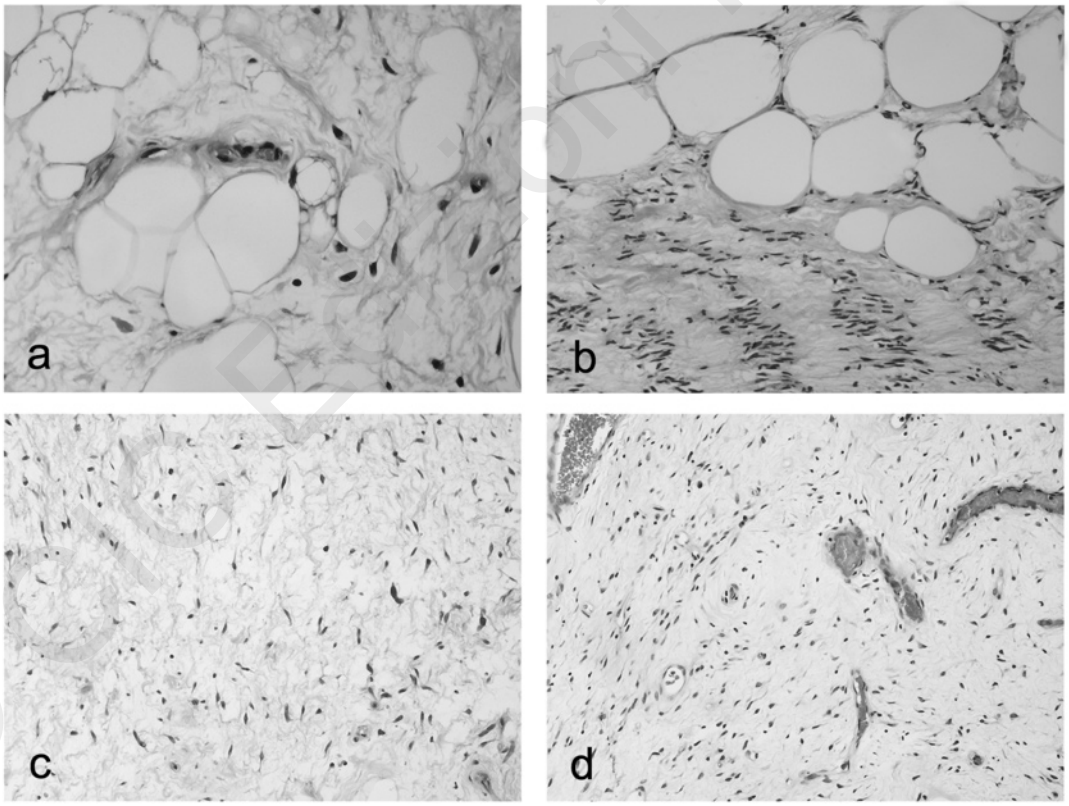

Figure 3 - Histopathological examination. The well differentiated liposarcomatous component exhibited a) sclerosing and b) spindle cell liposarcomatous morphology, while c) the dedifferentiated low grade myxofibrosarcomatous areas were composed of haphazardly arranged atypical pleomorphic, spindle and stellated, fibroblasts, some with "pseudolipoblastic" appearance d) frequently aligned along thick curvilinear stromal vessels (H/E X200).

fatty mass, especially in well-differentiated tumor (8, 12).

More dedifferentiated histological tumors are the myxoid liposarcomas. Given its location in the spermatic cord, there is no contrasting anatomy, such as muscle, that would indicate that the tumor is invasive. Hence, the mass appears to be inguinal hernia which leads to a delay in treatment (5, 8).

Radical surgical resection remains the definitive treatment for a localized well-differentiated and low grade dedifferentiated liposarcoma. Depending on the tumor size, an excisional biopsy might be performed. Optimally, radical resection with negative margins ≥1cm is possibly curative. Liposarcomas can also be treated with adjuvant radiation and chemotherapy, but this is generally reserved for higher grade tumors. Radiation therapy can be utilized in select cases of low grade, high grade, or superficial tumors >5cm or low grade deep tumors <5cm. Adjuvant radiotherapy for high grade lesions does have a benefit in decreasing local recurrence rates from 44% to 26%; however, there does not appear to be an impact on the 5-year overall survival rate. In order to provide optimal care, it is recommended that a patient with a localized liposarcoma be evaluated by a multidisciplinary committee to address consideration for possible surgical resection, radiation, or chemotherapy given consideration for the anatomical site and possible sequelae of therapy versus the pathological aggression of the liposarcoma (13-16).

Conclusion

The tumor in our case was mainly a well differentiated liposarcoma along with dedifferentiated low grade myxofibrosarcoma, thus the risk for metastatic progression is low. The coexistence of such a case is rare. Due to its composition in the imaging scans, a liposarcoma is usually misdiagnosed as hernia. Hence it must be taken into account in the differential diagnosis in patients present in with inguinal hernia symptoms.

Conflict of interest

The Authors declare that they have no conflict of interests regarding the publication of this paper. This work did not receive any specific grant from any funding agency in the public, commercial or not-for-profit sector.

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