Dedifferentiated Liposarcoma of the Epididymis: Case Report and Literature Review

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ABSTRACT

The dedifferentiated liposarcoma of the spermatic cord is a rare tumor, and the epididymal localization is even rarer. The treatment is based on inguinal orchi-epididymectomy, with resection of adjacent structures if invaded. Adjuvant radiotherapy may be an attractive approach, given the high rate of local recurrence. Metastatic patients and dedifferentiated subtypes constitute the main indications of chemotherapy, even if her role is still debated. We describe the case of a 58-year-old man who underwent surgery for a dedifferentiated liposarcoma of the epididymis. A review of the literature for the various facets of this condition is also provided.

Introduction

Paratesticular liposarcoma is a rare mesenchymal tumor occurring mainly in the elderly [1]. Epididymal localization is exceptional. The lack of clinical or radiological features makes preoperative diagnosis difficult. Inguinal orchi-epididymectomy is the Gold Standard [2]. The correlation between the histological grade and the outcome, and the benefit of an adjuvant treatment after surgery, remain debated because of the low number of cases reported in the literature. We report here the case of a 58-year-old patient diagnosed with scrotal mass, who underwent inguinal orchi-epididymectomy. No adjuvant treatment was performed. Through our observation and data from the literature, we will discuss Diagnostic and therapeutic management, and prognostic factors of paratesticular liposarcomas.

Patient and observation

A 58-year-old male, diabetic patient consulted in our structure for a left scrotal mass which appeared 5 months ago. The physical examination found a solid, painless, left epididymal mass and no lymphadenopathy, with a mild hydrocele, and a normal spermatic cord.

The scrotal ultrasound revealed the presence of a heterogeneous, non vascularized, tissular mass, measuring 43 x 31 mm (FIG 1), in the left epididymal head, with the presence of a left mild hydrocele (FIG 2). Tumor biological markers (αFP and βHCG) were normal.

The patient underwent an inguinal orchi-epididymectomy after the spermatic cord has been clamped. The postoperative course was simple.

The gross examination showed a normal-looking testicle with an intact albuginea. The epididymis was the site of a whitish, homogeneous tumor measuring 50 x 40 x 45 mm, with a soft consistency, and having an intimate contact with the albuginea, without invading it (FIG 3). On microscopical examination, there was a proliferation of tumor cells with a fasciculated architecture. The nuclei were atypical because of their irregular shape and size.
There are many mitoses. Moreover, we note the presence of a well differentiated adipose sector, delimited by septa and sheltering some atypical nuclei, without any zone of necrosis (FIG 4).

Figure 4. Histological aspect of the orchidectomy piece (Gx400), showing the presence of tumor proliferation with atypical and irregular nuclei in shape and size.

There are many mitoses.

Thus, the diagnosis of dedifferentiated epididymal liposarcoma was made. Immunohistochemical study has not been performed. The resection limits were good, particularly in spermatic cord. The case was discussed at a multidisciplinary consultation meeting with oncologists, and the option of close follow-up was chosen.

Discussion

Paratesticular malignancies are particularly rare. Spermatic cord sarcomas are the most common tumors. They are mainly represented by differentiated liposarcomas; their frequency is estimated at 7 % [1, 3, 4].

Their rarity explains their under-diagnosis, and the lack of a well-defined therapeutic strategy. LESAUVAJE described in 1845 the first case of sarcoma of the spermatic cord [5, 6].

Approximately one hundred cases of liposarcoma of the spermatic cord are found in the literature [7]. The pathologists of the Royal Marsden and John Hopkins reported a histological description of 30 patients treated for liposarcoma. Three-quarters of the tumors were developed at the expense of the spermatic cord, 20 % at the expense of the testis and 4 % of the epididymis [8].

Age at discovery is generally between 40-70 years [7]. Clinically, paratesticular liposarcomas have no specific signs compared to other testicular or paratesticular tumors [9-11]. At physical examination, we find a nodular, firm, usually painless scrotal or inguinal mass, with variable size [10, 11]. There are no tumor markers that can help in diagnosis. Radiologically, there are no pathognomonic signs [2, 11].

Computed tomography and MRI do not appear to be superior to ultrasound in the local exploration of spermatic cord tumors [5]. Inguino-scrotal ultrasound allows the detection of solid, hyperechogenic and heterogeneous lesions, but sometimes the detection of the benign or malignant nature is difficult, especially when it is presented as small indurated nodules inside a fatty tissue of normal consistency [7]. The CT allows suspecting liposarcomas by affirming their fatty nature, but these lesions can sometimes be hypodense in comparison with the subcutaneous fat. Computed tomography also provides a topographic diagnosis and precise the locoregional extension [12]. In recurrent cases, FDG-PET scan may be useful [7].

Histologically, several types of liposarcoma are described: well differentiated, dedifferentiated, myxoid (and its high-grade variant with round cells) and pleomorphic [9-11]. The diagnosis is based on the identification of the lipoblast (lipid-vacuole cells that press back the nucleus) [10, 11, 13]. The well-differentiated type accounts for about 50% of cases [5]. The size varies from 3 to 30 cm [2, 9, 8]. On macroscopic examination, lesions are well circumscribed by a thin translucent capsule, or another more fibrous. On gross examination, these lesions have the appearance of fat, with lobules of variable size. The dedifferentiated variant contains myxoid, necrotic or hemorrhagic zones [4].

Differential diagnosis is made between well differentiated liposarcoma and lipoma, which is a delicate situation. This sometimes requires the use of immunohistochemistry (MDM2 and / or CDK4 antibodies). Their hyperexpression makes it possible to establish the diagnosis, even if it is not sensitive (MDM2) nor specific (CDK4) at 100 % [7, 14].

The presence of a dedifferentiated contingent is the main determinant of the aggressiveness of this tumor. Its development may occur "de novo" or during the recurrences of a well-differentiated liposarcoma, which gives the complete tumoral excision all its interest [7].

The treatment of choice for paratesticular liposarcoma is surgery [1, 7]. Following the general principles of surgical management of sarcomas, surgical excision with inguinal orchi-epididymectomy and wide resection of the tumor with microscopically free margins are the base of treatment [1, 7, 14]. The location of these tumors constitutes a major obstacle to complete resection, which explains the high rate of recurrence. The majority of the publications reported local recurrence rates estimated at 50 % when the treatment was exclusively surgical [7].

It is well established that the grade and size of sarcomas influence their rate of local recurrence. The sarcomas of the spermatic cord, whatever their grade and size, do not emerge from this tendency when managed by surgery alone. This raises the question of the value of adjuvant treatment [7, 13]. The role of radiotherapy seems to be more than uncertain. For Coleman et al, adjuvant radiotherapy did not significantly decrease the rate of local recurrence and did not improve overall survival [12]. For other authors, an approach combining surgery and radiotherapy seems to give more prolonged control over tumor disease [7, 15].

The role of chemotherapy remains unclear and still unclear. Chemotherapy is an interesting tool in the palliative treatment and to avoid rapid progression in the case of metastatic evolution of the disease. Chemotherapy protocols similar to those of soft tissue liposarcoma are used [7]. Well differentiated and myxoid forms have a good outcome with a 5-year survival of 85 % [9]. In contrast, 5-year survival decreases to 20 % in round-cell and pleomorphic liposarcomas. On the other hand, dedifferntiated liposarcomas have very poor outcome [2, 11].

Conclusion

The spermatic cord represents a rare localization of liposarcoma, the morphological and Immunohistochemical aspects of which are identical to those of the other localizations. Diagnosis is often delayed. Imaging can help in histological diagnosis. Therapeutic management is surgical, and requires a multidisciplinary strategy even if the place of an adjuvant treatment is not yet well specified.
The prognosis of each liposarcoma depends on its histological subtype.

**Competing interests**

The authors declare no competing interest.

**References**


