Case report on paratesticular sarcoma and a focused review of the literature

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SUMARY

Insidious, usually painless, and rare inguinoscrotal masses arising from paratesticular elements (spermatic cord, epididymis, tunica or the stroma) are known as paratesticular tumors. The overall incidence is less than 5%, and the total number of giant (>10cm) paratesticular liposarcomas is less than 300 cases recorded since 2020. We report a similar clinical dilemma of a giant scrotal mass managed via a wide local resection and close surveillance in a 61 year old male. However, owing to its rarity, there is no fixed treatment protocol; hence, a supplementary review of similar cases is discussed here.

Keywords: *case report; liposarcoma; spermatic cord; testis; orchidectomy; sarcoma*

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INTRODUCTION

Barring the testis, tumors arising within the scrotum from its contents are labeled as paratesticular tumors. The source of origin can be varied, like the epididymis, spermatic cord, tunica layers, and other supporting stroma (1). The rarity of these tumors is well known, with the majority being benign in nature. Fewer than 30% are malignant, the most common being sarcomas (1). Almost 30% of all genitourinary sarcomas arise from the spermatic cord (2,3). When the tumor exceeds 10cm, it is labeled as a "giant" paratesticular mass, of which approximately less than 300 cases have been recorded since 2020 (4,5). Pre-operative investigations may not always point to a final diagnosis, and herein, we report a similar clinical conundrum, in a 61-year-old male, with an insidiously increasing scrotal mass suspected to harbor testicular malignancy but a varying final pathology.

CASE PRESENTATION

A 61-year-old hypertensive male presented with a history of swelling in the left hemiscrotum over the past two years. It had gradually increased in size and started causing him pain and discomfort while walking, a few months prior to his presentation to us.

Examination revealed an 18cm x 15cm x 7cm scrotal mass, hard and not associated with tenderness on palpation. No distinct testicle was palpable adjacent to this mass, and the transillumination test was negative. Serum tumor markers (LDH, Beta-HCG, AFP) were normal. The ultrasound of the scrotum showed a hyperechoic mass with lobulated margins and small cystic areas within it.

A CT scan suggested a large (14.1 cm x 12.2 cm) complex solid mass and cystic areas in the left hemiscrotum wherein the left testis was not separately visualized (Figure 1).

No lymphadenopathy or distant metastases were noted. The patient underwent a high inguinal orchiectomy with cord ligation and mass excision. Intraoperatively, the testis appeared grossly normal, abutted by the mass, which was seen to arise from the spermatic cord. The entire specimen was excised with adequate margins (Figure 2).

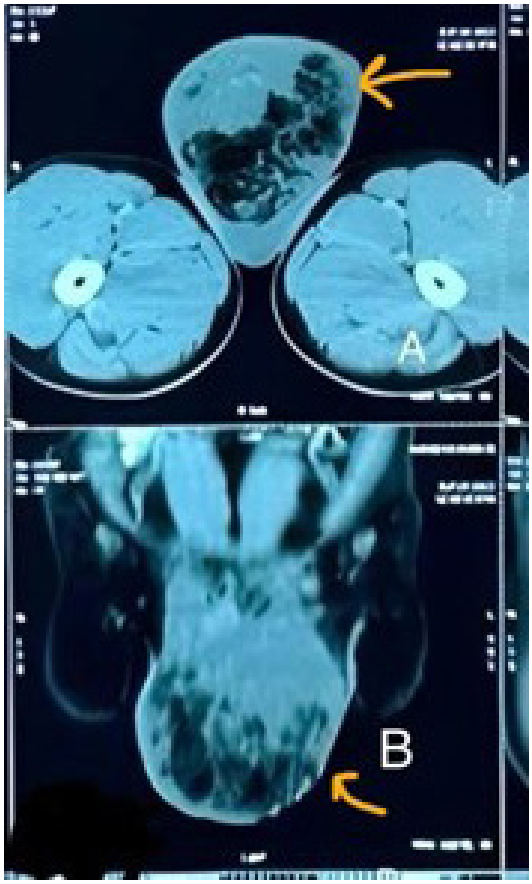

Figure 1. CT scan of the left hemiscrotum. A) Axial section; B) Coronal section. The arrow indicates the scrotal mass.

The patient's postoperative course was uneventful, and he was discharged on day 2. Two weeks after the procedure, the histopathology slides were reviewed, which suggest a low grade well-differentiated liposarcoma, possibly from the spermatic cord elements (Figure 3). Given tumor-free margins and FNCLCC (Federation Nationale des Centers de Lutte Contre le Cancer histological grade I (6,7), the multidisciplinary tumor board decided to keep the patient on close follow-up and a three-month imaging for the first year with CT and annual fluorodexoglucose positron emission tomography

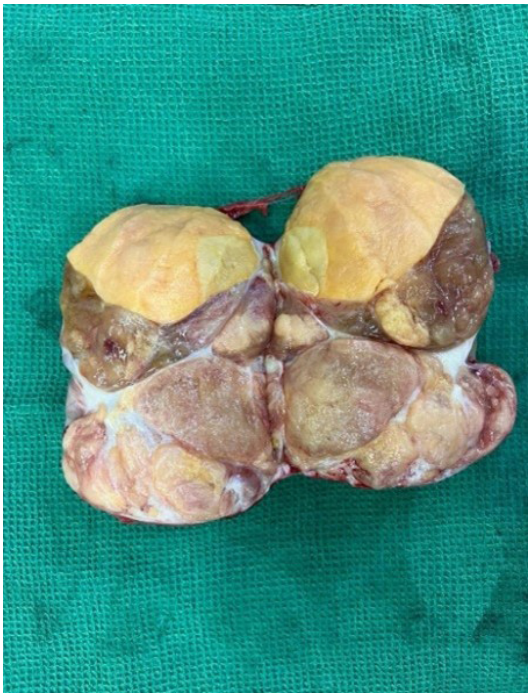

Figure 2. Material obtained during the surgical procedure: cut section of the gross specimen of the mass.

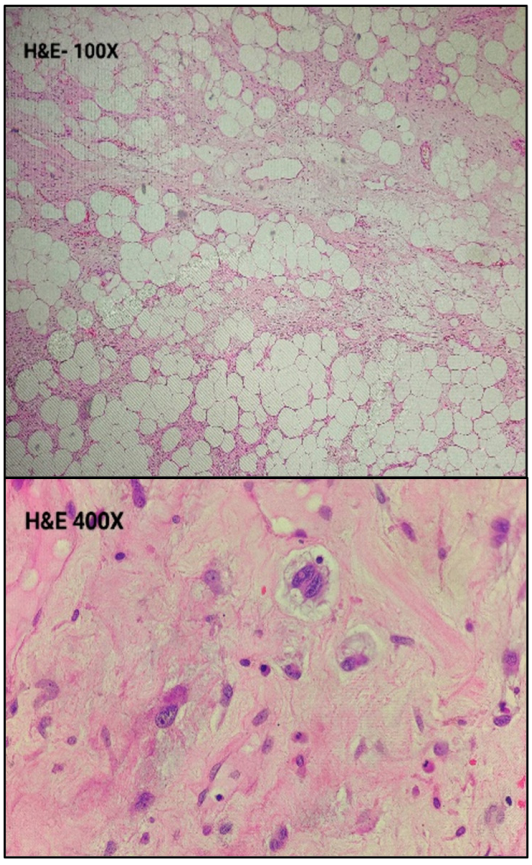

Figure 3. Histopathology hematoxylin-eosin (H&E) slides of the obtained material. **Above:** Adipocytic tumor with fibrous spindle septa. **Below:** Atypical stromal multinucleated forms with scattered mast cells.

(FDG PET). The last visit of this patient was in the 6th month postoperatively; the patient is doing well and has no surgical site morbidity.

DISCUSSION

Le Sauvage, in 1845, was first credited with reporting on paratesticular sarcoma, and Herbert, in 1952, documented the first paratesticular liposarcoma (8). Most paratesticular tumors are benign, with a handful being malignant, i.e., sarcomatous change. Of the malignant variety, the common varieties include liposarcoma, leiomyosarcoma, and rhabdomyosarcoma. Dedifferentiated sarcomas and malignant fibrous histiocytomas (MFH) are uncommon varieties. The paucity of literature on these tumors makes a formal management protocol challenging.

Rhabdomyosarcoma, an aggressive tumor, is seen commonly in the first two decades of life, whereas liposarcoma, leiomyosarcoma, and fibrosarcoma are seen in adults. Geographically, there is a higher incidence of these tumors in Japanese men (3).

The management of this disease starts with an efficient investigative protocol. Investigations help not only point toward a diagnosis but also help plan a wide local resection of the tumor so as to avoid recurrence and rule out metastasis. In our case, we relied on CT, which helped us rule out metastatic disease. However, it was insufficient in differentiating a paratesticular from a testicular mass.

Akbar et al reviewed the role of ultrasonography in paratesticular masses and differentiated them from other scrotal mass mimics. Although hetero-echoic masses with differential vascularity may point to a spermatic cord tumor, it is unreliable in differentiating the subtypes and in such large masses identifying a normal testis. CT and MRI are more specific in identifying the location, extent, and possible character of the mass (9). However, there are no pathognomic CT features for differentiating benign from malignant masses (3). A systematic review undertaken to establish the role of FDG PET in musculoskeletal soft tissue masses provided some evidence to support a standardized uptake value (SUV) threshold of 2.4 to distinguish between benign and malignant lesions. No similar study has been undertaken on a large scale specifically for paratesticular lesions (10). Better survival and local recurrence are noted in patients with SUVmax less than 10.3, and this study established low uptake by myxoid and synovial sarcoma types (11).

It is well established that the treatment of soft tissue sarcomas begins with complete surgical resection (12). Hence, initial management of paratesticular masses warrants a radical orchiectomy with high cord ligation. Eventually, it is the tumor biology, local clearance, and adjuvant treatment that dictate the clinical course of our patients. In our case, orchiectomy seems sufficient for establishing adequate treatment for our patient. However, this requires a strict and close follow-up regimen. The role of the multidisciplinary team effort is regarded as a primary approach to such cases, with the occasional need for adjuvant radiotherapy or chemotherapy. Post-operative radiotherapy is debated due to the recur-

rence of surgically aggressive lesions (13).

The overall survival of 64% and five-year survival of up to 70-75% have been reported in some larger series (3,14). The threshold for wide local resection during re-resection should be quite low, and attaining a negative surgical margin significantly influences disease-free survival. Coleman et al, based on their retrospective data, reported the inadequacy of local resection and the need for wide local re-resection (3). Resecting of inguinal scars, abdominal muscle layers, hemiscrotectomy, and spermatic cord remnant excision are some maneuvers that should be carried out to establish an efficient wide local clearance. Retroperitoneal lymphadenectomy (RPLND) should be reserved for cases of lymphatic spread rhabdomyosarcoma and specifically embryonal rhabdomyosarcoma.

CONCLUSION

Paratesticular sarcomas are more prevalent now due to better investigative and pathological expertise. They are notorious for recurrence, local spread, and occasional aggressive behavior. Hence, it becomes important to advance our findings in tumor biology and carry out multidisciplinary efforts to establish better long-term survival outcomes in our patients. This warrants a coordinated effort at a multi-institutional level. Due to the preoperative toxicity of chemoradiation, the mainstay remains a wide local resection for negative margins, with adjuvant radiation control after adequate surgical wound healing.

Ethical Considerations: Signed and written consent received from the patient before proceeding to formation of manuscript and for image use.

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