CASE REPORT

Spermatic cord leiomyosarcoma in a young male: A case report and literature review

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Abstract

Spermatic cord Leiomyosarcoma is an extremely rare intrascrotal tumour. Owing to its rarity, no definitive management guidelines have been formulated as yet. The majority of published literature comprises of case reports or case series and show varying outcomes depending upon multiple patient- and disease-related factors. Almost all cases are older adults with majority in the sixth or seventh decades of life. It is commonly labelled as an indolent curable tumour if treated early by radical orchiectomy. The role of lymphadenectomy, adjuvant radiotherapy or chemotherapy is unclear.

This case report concerns a young 38-year-old man who suffered from a painless firm left hemiscrotal mass for the past two years. Ultrasonography showed an intrascrotal paratesticular mass. Metastatic workup was negative. Left radical orchiectomy was performed and histopathology of the surgical specimen revealed leiomyosarcoma of the spermatic cord. The patient is on post-surgery follow-up and disease-free for six months. A literature review is also presented.

Keywords: Leiomyosarcoma, Para-testicular, Spermatic-cord, Sarcoma, Testicular.

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Introduction

Soft-tissue sarcomas of the genitourinary tract are extremely rare, accounting for only 1-2% of all urinary malignancies and less than 5% of soft-tissue sarcomas in general.[1] Primary Para testicular tumours are even more rare, comprising less than one tenth of all intra scrotal tumours and in itself are a heterogeneous group of tumours of various histologies. These tumours are often grouped according to their paratesticular tissue of origin, which may be tunica albuginea, epididymis, or spermatic cord. Among them, primary spermatic cord tumours are the commonest, with a collective prevalence of less than 0.3 per million population. Paratesticular tumours can also be classified histologically; frequently reported malignant subtypes are liposarcomas, leiomyosarcomas, rhabdomyosarcomas and fibrosarcomas,[2] while common benign histologies are haemangiomas, lymphangiomas, lipomas, and leiomyomas. Although the exact number of cases is difficult to estimate due to its rarity and scant literature, an estimated figure of 110 cases of leiomyosarcoma of the spermatic cord have been quoted in literature, almost all of them in older adults.[1,3] The disease usually presents as firm, painless gradually enlarging mass in the scrotum. However, it may sometimes be painful. Diagnostic modalities include ultrasonography, computed tomography, or magnetic resonance imaging. Due to its rarity, there are no robust, evidence-based guidelines for its management and follow-up. Usual treatment recommendations are based on case reports, small case series, and literature reviews. Protocols defined in SCARE[4] guidelines were used while reporting this case.

Our study will add to the knowledge that primary Leiomyosarcomas of spermatic cord can also occur in younger adults and should be considered in differential diagnosis. Moreover, it can be managed with definitive surgery only, if we are able to get negative surgical margins. Adjuvant radiotherapy and/or chemotherapy can be reserved for any future metastasis.

Case Report

A male patient, 38 years of age, presented to the Combined Military Hospital, Rawalpindi, in November 2020 with a painless lump in the left hemiscrotum for the past two years. He had no history of scrotal pain, lower urinary tract symptoms, testicular trauma and fever or chills. Physical examination revealed a palpable mass of 5cm that was firm, non-tender and appeared to arise from the left spermatic cord. It was not attached to Dartos or the surrounding tissues. Blood counts and chemistry were normal. Germ cell tumour marker levels, including alpha feto-protein, beta-hcg and lactate dehydrogenase were within normal limits.

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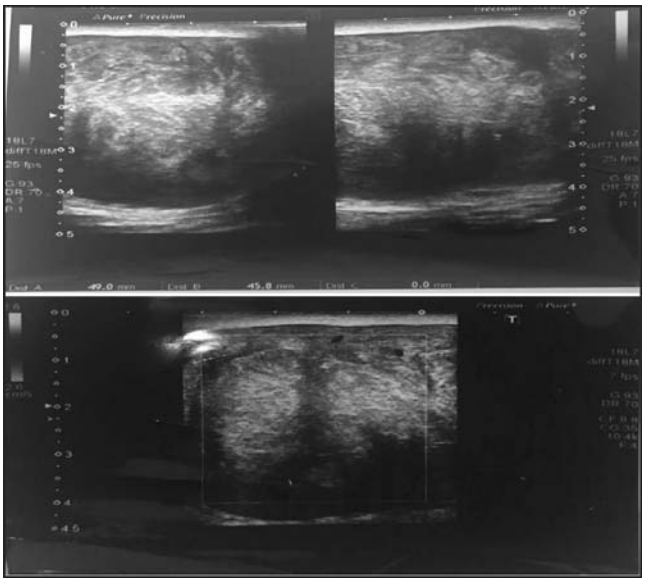

Figure-1: Ultrasonogram showing a well-defined solid heterogeneous paratesticular mass. Mass showed increased vascularity on Doppler ultrasound.

Ultrasonography of the scrotum (Figure-1) showed 4.9 × 4.5 cm, well-defined solid heterogeneous mass located adjacent to the left testicle with increasing vascularity as shown by Doppler ultrasonogram, suggesting a neoplastic mass. Contrast enhanced CT-scan of the thorax, abdomen, and pelvis showed no locoregional or distant metastasis. Surgery was planned and en masse left radical orchiectomy with high ligation of spermatic cord was performed. Peroperative findings were consistent with the clinical and radiological findings, with the mass seen arising from the base of the spermatic cord but not involving Dartos or testes. On microscopic examination (Figure-2), the mass consisted of elongated spindle-shaped cells in interlacing bundles with variably eosinophilic cytoplasm and blunt-ended hyperchromatic nuclei on Haematoxylin and Eosin (HE) stain. Tumour was well-differentiated with <50% necrosis and 9 mitoses per high power field (9/HPF). On immunohistochemistry, Smooth Muscle Actin (SMA) and Caldesmon tested positive. Special Masson trichrome stain was negative for collagen. Definitive pathological diagnosis of this patient was a grade 1 Leiomyosarcoma (LMS) of the left spermatic cord according to National Federation of French Cancer Centre Institute (FNCLCC) grading system, with stage cT2b cNo cMo as per TNM staging. Follow-up with CT scan of the thorax, abdomen, and pelvis after six months showed no evidence of any local or distant recurrence. Repeat germ cell tumour markers are also within normal limits.

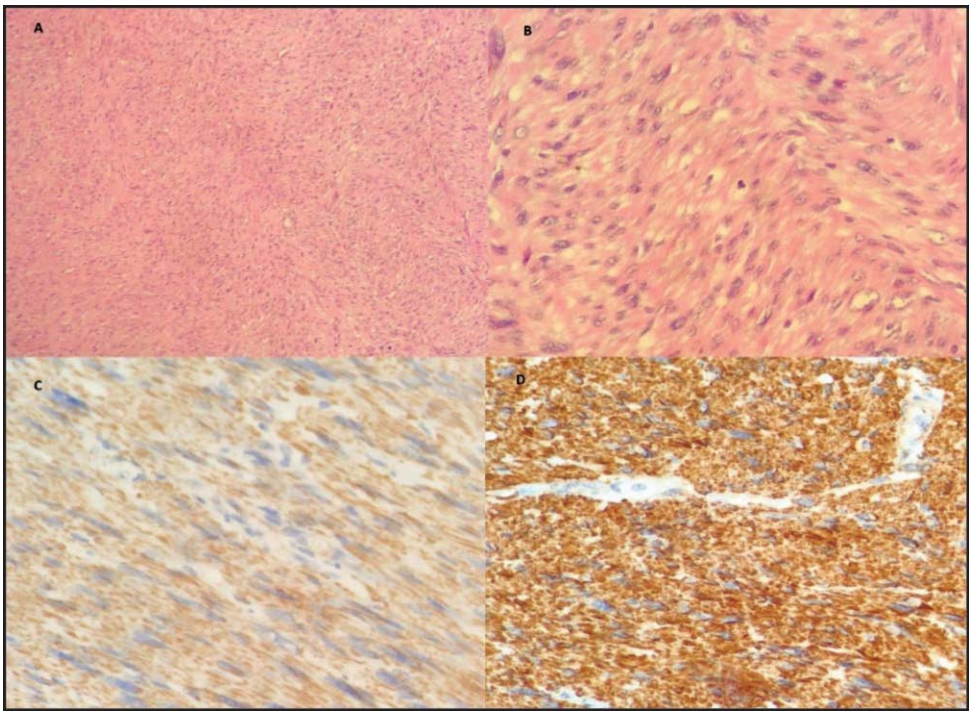

Figure-2: Microscopic and immune-histochemical pattern of studied tumour (A) HE staining at 10x magnification (B) HE staining at 40x magnification (C) Staining for smooth muscle actin (D) Staining for h-Caldesmon.

Discussion

Paratesticular tumours are a rare entity comprising less than 5% of all scrotal masses; the remaining are testicular tumours.[5] Of all the paratesticular tumours, nearly one third are malignant. A study conducted at the Massachusetts General Hospital[2] described the largest cohort of spermatic cord tumours (SCTs) studied to date, comprising 362 cases of all types of SCTs. The annual prevalence of SCTs was noted to be 0.3 cases per million that did not change over time. Common malignant histiotypes were liposarcoma (46%), Leiomyosarcoma (20%), histiocytoma (13%) and rhabdomyosarcoma (9%) with leiomyosarcoma and histiocytoma being the most aggressive subtypes. Median age of all types of SCTs was described to be 64.7 years. Tumour stage, grade, histotype, and lymph nodal involvement were independent prognostic factors on multivariate analysis.

Three main subcategories of

leiomyosarcoma, depending on the topography, have been described. These are LMS of deep soft tissue, cutaneous/subcutaneous tissue (including the Dartos muscle) and vascular origin. Spermatic cord has been classified as deep soft tissue by the American Joint Committee on Cancer (AJCC). The primary origin of LMS of spermatic cord is undifferentiated mesenchymal cells of cremaster muscle and ejaculatory duct. Its grading, like other soft tissue sarcomas, is based on the FNCLCC Criteria.[6] These tumours have variable behaviours and prognosis. Painless, gradually enlarging, firm intra-scrotal or para-testicular mass is the common presentation and maybe associated with hydrocoele. Ultrasonography is the imaging modality of choice in the initial workup of any intra-scrotal mass as it can identify location as well as differentiate benign versus malignant tumours with very high sensitivity. Generally, benign tumours are homogeneously hyperechoic, while malignant ones are either hypoechoic or heterogeneously hypo-hyperechoic, usually with irregular and high vascularity. Some malignant SCTs have also been reported to be hyperechoic and these should be evaluated further by magnetic resonance imaging.[7] CT and MR imaging is also helpful to assess local tumour extension while PET-CT scan is useful to assess nodal or distant metastases, if any. Definitive diagnosis is given on histology with immunohistochemistry for smooth muscle differentiation. This is done by staining for its markers, like alpha smooth muscle actin, muscle-specific actin, and desmin. Staining can also be done for CD34 and h-caldesmon.

Commonly practiced treatment is radical orchiectomy with high spermatic cord ligation. No standard guidelines are available due to the rarity of the disease. The paucity of cases has also made it difficult to understand the pattern of spread. The commonest means of dissemination are lymphomatous to iliac and retroperitoneal group of lymph nodes, haematogenous to the lungs, and by local infiltration of adjacent scrotal skin/testes.[8] The value of prophylactic lymphadenectomy in preventing recurrence or improving prognosis is undetermined. Data indicates that lymph node dissection is unnecessary unless enlarged lymph nodes are detected on imaging or seen during surgery.[9] A recently conducted study by US Sarcoma Collaborative Group regarding the role of radiotherapy in retroperitoneal sarcomas concluded that use of radiotherapy, both in adjuvant or neoadjuvant setting, was not associated with improved survival or reduced recurrence rate.[10] However, it has been used in select few patients with paratesticular sarcomas.[11]

This particular case of LMS of spermatic cord in a young adult patient of 38 years is extremely rare. Almost all of the related cases reported in literature involved older adults with median occurrence in the sixth and seventh decades of life. So LMS should be one of the differentials in a gradually enlarging firm mass of spermatic cord/para-testicular region even in young male patients. Treatment would usually, in the absence of metastasis, be radical orchiectomy with funicolectomy only. Adjuvant radiotherapy may, however, be offered in selective high-risk patients. The patient may have prolonged recurrence-free survival.

Conclusion

The rarity of primary spermatic cord leiomyosarcoma precludes any evidence-based guidelines regarding definitive treatment, making management decisions often exacting. Yet, this condition should be considered among differentials of all adult males presenting with a painless scrotal mass, regardless of age. Treatment offered in our case, being localized, involved radical orchiectomy and 3 monthly follow-up till first year. This might be the sufficient treatment option till more literature is available to devise optimal management criteria.

Disclaimer: The abstract has not been presented or published in a conference, or published in an abstract book and is not part of any PhD thesis.

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