

Case Report

Epididymal leiomyosarcoma: a rare case of scrotal swelling

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ABSTRACT

Sarcomas of the genitourinary tract are extremely uncommon and accounts for only 1-2% of genito urinary malignancies. Sarcomas of the para testicular region, comprising tissues such as the epididymis, spermatic cord, inguinal canal and testicular tunica are also extremely rare. epididymal leiomyosarcoma accounted only for 4 percentage of all para testicular tumours and only 16 cases are reported so far in literature and they account 4% of all Para testicular sarcomas. We are presenting a 61-year-old patient presented with a hard swelling of 1 year duration, with no other associated symptoms. On ultrasound evaluation, it was reported as extra testicular lesion, possibly from epididymal tail. We performed a high inguinal orchidectomy. Histopathological examination revealed a para testicular leiomyosarcoma arising from epididymis. This case has discussed because of the rarity of the disease and possible cure if diagnosed early and treated aggressively.

Keywords: Paratesticular tumour, Leiomyosarcoma, Sarcomas

INTRODUCTION

Para testicular sarcomas are extremely rare, with only 110 cases reported in literature.¹ The most common histological subtypes include liposarcoma, leiomyosarcoma (LMS) and rhabdomyosarcoma. LMS is thought to arise from para testicular smooth muscle tissues and they are prone for direct invasion and also early haematogenous spread. No established treatment protocol is so far established; however, general consensus comprises inguinal radical orchidectomy and high cord ligation, with some case series advocating adjuvant radiotherapy.^{2,3}

We are presenting a 61-year-old patient presented with a hard swelling of 1 year duration, with no other associated symptoms. On ultrasound evaluation, it was reported as extra testicular lesion, possibly from epididymal tail. We performed a high inguinal orchidectomy. Histo-

pathological examination revealed a para testicular LMS arising from epididymis.

CASE REPORT

61-year-old male presented with a painless right scrotal swelling of 1 year duration. No other associated symptoms. He had history of skeletal TB 13 years back and pleural effusion 2 years back for which he took antitubercular treatment. There were no other known comorbidities.

On physical examination, he has a firm to hard right scrotal swelling which is painless with well-defined margins of size 6×5 cm, right testis was not separately palpable. left testis appears normal. Patient was evaluated on a case of testicular tumour. Ultrasound examination of scrotum revealed a fairly well defined lobulated, predominantly hypoechoic mass lesion with 4×2.6 cm with significant vascularity, related to inferior aspect of

right testis, most likely extra testicular tumor, Epididymal tail lesion likely.

Tumour markers were sent, showed 1.29 IU/ml (0.5-5.5), lactate dehydrogenase 524 U/l (225-450), human chorionic gonadotropin 0.792 mIU/ml. CT thorax, abdomen, pelvis, plain and contrast study were done. No signs of metastasis. An enhancing lesion noted in posterior basal segment of left lower lobe with associated pleural thickening and was reported as Resolving consolidation/fibrosis.

Patient underwent an elective right high inguinal orchidectomy under general anesthesia and intraoperative findings were A hard 5×4 cm mass in right testis, in lower pole with no infiltration to tunica vaginalis, tunica albuginea, left testis and normal cord structures.



Figure 1 (A and B): Gross cross section.

Surgical specimen consisted of right testicular tissue with spermatic cord whole weighing 120 gm (Figure 1A). Spermatic cord alone measures 7 cm in length. Testis was enlarged diffusely measuring 7.5×6×4.5 cm. Tunica vaginalis was adherent focally to testicular mass. Epididymis show cyst measuring 1.5×1.5 cm. Cut section of para testicular mass show a well circumscribed lobulated solid lesion, having a whorled appearance, measuring 5×5×5 cm, periphery of the lesion showed normal testicular tissue. Cut sections of spermatic cord was unremarkable (Figure 1B).

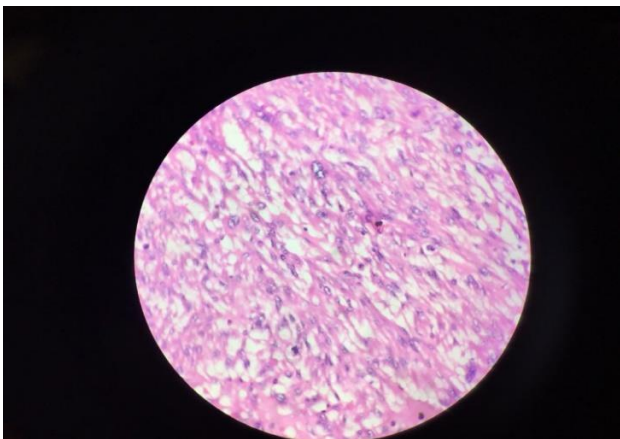


Figure 2: Histopathology.

Histologically it shows a circumscribed neoplasm composed of spindle cells arranged in interlacing bundles and fascicles. Individual cells have elongated cytoplasmic processes and mildly pleomorphic vesicular fusiform nucleus with blunt ends & conspicuous nucleoli, mitosis 1-2 per HPF seen. Few thick walled blood vessels and scattered lymphocyte infiltrates seen between tumour cells. Tunica vaginalis and albuginea free of neoplasm. In Figure 2, it was reported as paratesticular LMS. On immunohistochemistry, tumour cells were positive for SMA and desmin positive (Figure 3).

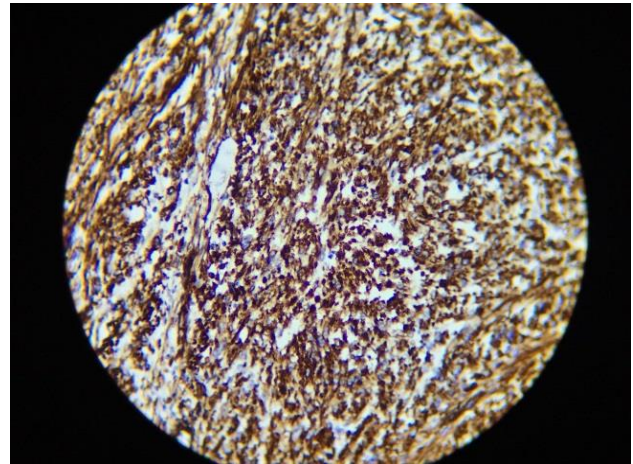


Figure 3: Immunohistochemistry.

DISCUSSION

A scrotal swelling can be testicular or para testicular, of which para testicular tumors are rare. Paratesticular region includes spermatic cord, testicular tunics, epididymis, appendices, epididymis & testis. This area contains different elements histologically like epithelial, mesothelial and mesenchymal. Hence tumours from this area has different behavioural pattern.

Primary soft tissue tumours arising from the scrotal sac can be classified into four groups, i.e. scrotal subcutis/dartos/skin, testicular, paratesticular and those arising from spermatic cord.

Chen et al found only one out of 24 had sarcoma and 23 had benign tumors in his series of 18 patients and In another study from Chandigarh, India, out of a total of 228 cases of epididymal nodules who have undergone fine needle aspiration cytology only one had LMS (0.4%) and rest had benign lesions.¹ Thus, sarcomas arising from the epididymis are rare and the most common primary epididymal soft tissue sarcoma is LMS.

LMS, liposarcoma, fibrosarcoma & undifferentiated pleomorphic sarcoma and different histological subtypes, among which liposarcoma is the most common.

Certain predisposing factors have been described in the literature leading to testicular LMS such as exposure to

large doses of anabolising steroids, chronic inflammation, or past radiotherapy.⁴⁻⁶ There are no reported predisposing factors leading to epididymal LMS with respect to its prognosis in the literature.

18 cases of primary epididymal LMS that have been published. Kwae et al, they usually occur in older patients with an average age of 60 years.^{7,8} Out of 18 cases reported, only 2 patients are below the age of 50 years (one was a pediatric patient of 6 years and the other was 28 years).

The biological behaviour of epididymal LMS may be less aggressive than para testicular LMSs. They do not spread commonly to the lymph nodes.

Initial workup includes the primary imaging method - Ultrasonography, for differentiating extra testicular lesion from intratesticular lesion. In ultrasonography a heterogenous solid mass is with increased vascularity on colour Doppler may be seen. Metastatic work-up includes CT scan of chest and abdomen.⁹ To identify tumour extension CT & MRI may be useful and PET CT for nodal involvement. But preoperatively it is difficult to diagnose paratesticular LMS.

Histological examination of excised specimen is required for a definitive diagnosis with immune-histochemical & morphological differentiation.

Specific histological findings at low power includes spindle cells organized perpendicularly with fascicular arrangement and at high power seen as eosinophilic cytoplasm containing longitudinal fibrils and hyperchromatic blunt ended nuclei.

On immunohistochemical study of LMS may show characteristics of smooth muscle differentiation like smooth muscle actin and desmin. In some CD34 and cytokeratin may also be expressed.

Because of the rare incidence of LMS, no ideal treatment has been established. Best treatment option now is radical orchidectomy with high ligation of spermatic cord. But loco regional recurrence following surgery is common, as it is difficult to get negative margins due to anatomical constraints.

Hemiscrotectomy is indicated if scrotal skin is involved. Adjuvant radiotherapy may be useful for paratesticular sarcomas which improves loco regional control, but there is no studies on the use of radiotherapy in LMS and adjuvant chemotherapy has no role in the treatment of para testicular LMS.

CONCLUSION

When an elderly male presents with a painless intrascrotal mass, diagnosis of paratesticular LMS should also be considered as a rare possibility and ideal management currently is radical orchidectomy with high ligation of spermatic cord but loco regional recurrences are high.

This patient had typical history and examination findings and underwent surgery and kept under long term follow up. For establishing an ideal treatment protocol further researches to be performed for this rare entity.

This case was presented in view of extreme rarity and to discuss the management options.

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