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PARATESTICULAR LEIOMYOSARCOMA: AN UNUSUAL INTRASCROTAL TUMOR

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ABSTRACT
Paratesticular leiomyosarcomas are very uncommon tumors. If not properly diagnosed or treated, these tumors result in significant morbidity and mortality. We report a case of a 50 year old male presenting with painless left sided scrotal swelling, gradually increasing in size since two months. On examination, a hard mass was felt in left hemiscrotum, located above and separate from testis. Patient was subjected to left orchidectomy with high ligation of the spermatic cord and the specimen was sent for a histopathological examination. A histopathological diagnosis of leiomyosarcoma- grade 2 by French Fédération Nationaledes Centres de Lutte Contre le Cancer (FNCLCC) Grading System. We are presenting this case on account of the rarity of its occurrence.

INTRODUCTION

Leiomyosarcomas are malignant mesenchymal soft tissue tumors arising from any tissue containing smooth muscle. Soft tissue sarcomas contribute to 1-2 % of urological malignant tumors. Urological soft tissue sarcomas account for 2.1% of soft tissue sarcomas in general [1]. Paratesticular leiomyosarcomas contribute to only 10% of paratesticular sarcomas. Till date, over 100 cases of leiomyosarcoma of spermatic cord and a handful of cases of leiomyosarcomas of epididymal origin have been reported in literature [2]. They are usually seen in middle age or old age men presenting as a discrete, slow growing painless mass separate from the testis [3]. Diagnosis is made on histopathological examination. Radical orchidectomy with high ligation of the spermatic cord is the standard treatment.We report a case of paratesticular leiomyosarcoma- grade 2 (FNCLCC System) which was treated by orchidectomy and high ligation of the spermatic cord and followed up with surveillance.

CASE HISTORY

A 50 year old male presented with left sided scrotal mass which was painless and gradually increasing since two months. On examination, a hard mass measuring 7x4x2 cm was felt in left hemiscrotum located above and separate from testis. Ultrasonography revealed an extra testicular tumor in the left hemiscrotum. Computerised Tomography scan was suggestive of left extratesticular adenomatoid lesion [Figure 1]. Patient underwent left high inguinal orchidectomy for the tumor. Grossly, a well circumscribed, creamy white, lobulated, firm 70x40x20 mm mass was seen above the testis with focal areas of haemorrhage. Testis and epididymis were separate and unremarkable. Spermatic cord was measuring 6 cm in length [Figure 2]. On microscopy, a malignant spindle cell tumor was seen with tumor cells arranged in parallel bundles and fascicles with large intervening areas of coagulative necrosis. Nuclei showed blunt ends and were typically cigar shaped. Marked nuclear pleomorphism and

tumor giant cells seen. There were more than 20 mitoses per 10 high power fields [Figure 3a & 3b]. Thrombosed vessels were also seen in the tumor. Testis and epididymis were unremarkable. Surgical margins were free of tumor. A differential diagnosis of leiomyosarcoma, fibrosarcoma and undifferentiated pleomorphic sarcoma was considered on light microscopy. On immunohistochemistry**,** smooth muscle actin was strongly positive and desmin showed good positivity [Figure 3c & 3d]. CD68 and S100 were negative. A final diagnosis of high grade leiomyosarcoma-grade 2 (FNCLCC System) of paratesticular region was given based on tumor cells resembling normal smooth muscle cells, mitoses more than 20/10 hpf and necrosis less than 50% of tumor. The patient is currently free of disease and there is no evidence of metastasis after two years post surgery.

Fig 1. CT scan showing a 70x40x20 mm extra testicular tumor in left hemiscrotum

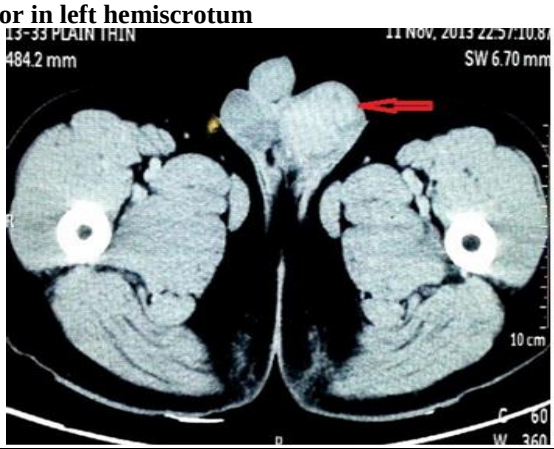


Fig 2. Gross photograph showing a well circumscribed, creamy white, lobulated, firm 7x4x2 cm mass above and separate from the testis

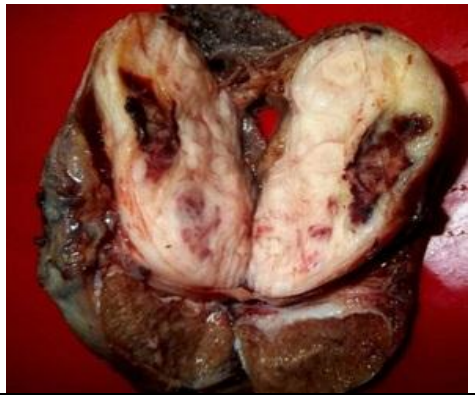
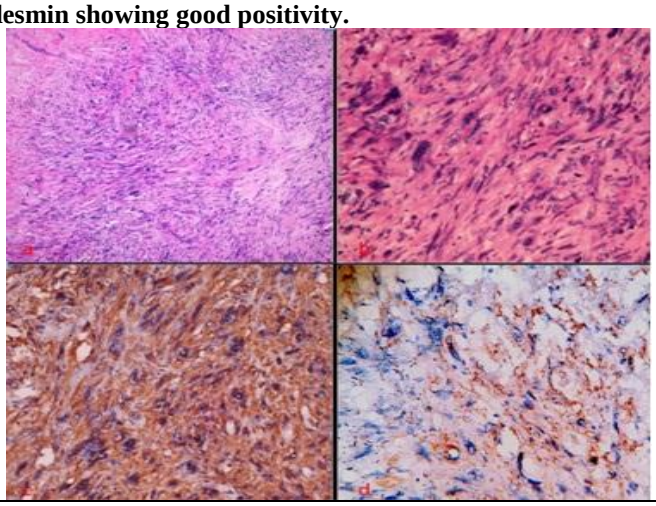


Fig 3. (a) Malignant spindle cells arranged in fascicles with intervening areas of necrosis. Marked nuclear pleomorphism & tumor giant cells seen. (H&E x100), (b) High power magnification showing malignant spindle cells arranged in fascicles with intervening areas of necrosis. Marked nuclear pleomorphism & tumor giant cells seen. (H&E x400), (c) Immunohistochemistry with smooth muscle actin showing strong positivity, (d) Immunohistochemistry with desmin showing good positivity.



DISCUSSION

Majority of masses in the scrotal sacs are neoplastic and are usually of testicular origin. A small proportion of these masses are extratesticular and arise from the paratesticular tissue. Paratesticular region is a complex anatomical area consisting of spermatic cord, testicular tunics, epididymis and vestigial remnants like appendices epididymis and appendices testis. The tumors arising in the paratesticular region form a heterogenous group with varying behavioural patterns. Rarely, metastatic tumors occur in paratesticular region [4].

The tumors of the paratesticular region include adenomatoid tumor, benign and malignant mesotheliomas, adenocarcinoma and papillary cystadenoma of epididymis, melanotic neuroectodermal tumor, desmoplastic small round cell tumor and mesenchymal tumors. Mesenchymal tumors are relatively rare and leiomyosarcomas are still rarer. Paratesticular leiomyosarcomas may arise from spermatic cord, scrotum or epididymis [1]. Leiomyosarcomas contribute to only 10% of paratesticular sarcomas. Over 100 cases of leiomyosarcoma of spermatic cord and a handful of leiomyosarcomas of epididymal

origin have been reported in literature [2]. Etiological factors are not well documented. In a cytogenetic analysis study conducted by Boghosian et al. it was concluded that at least three subtypes may be identified chromosomally within leiomyosarcomas [5]. Some authors assign the possible cause to be local irradiation in childhood [6].

Paratesticular leiomyosarcomas are usually seen in the sixth and seventh decades. Our patient was younger and was 50 years of age. The most common clinical finding is painless, firm, progressively growing intrascrotal, extratesticular mass as was seen in our case. It is difficult to diagnose them preoperatively and they are frequently misdiagnosed as benign tumors and require histopathology for diagnosis.

The typical histopathological features of leiomyosarcoma are fascicles of spindle cells with hyperchromatic cigar shaped nuclei and eosinophilic cytoplasm. On immunohistochemistry, these tumors often show positivity for smooth muscle actin, muscle specific actin and desmin and less commonly for CD-34, myogenin, Ki-67, S-100 protein [6]. In our case the histopathological features were classical and immunohistochemistry showed strong positivity for smooth muscle actin and good positivity for desmin.

Differential diagnoses on histology include fibrous mesothelioma, fibrous tumors and pseudotumors, fibromatosis and other soft tissue sarcomas. Immunohistochemistry is an effective tool to rule out these and to arrive at a definitive diagnosis of spindle cell sarcomas.

Paratesticular leiomyosarcomas are usually low grade tumors, but high grade tumors do occur and may behave more aggressively. Prognosis of the commonly occurring low grade tumorsis usually good if there is no local recurrence. Around 40% of cases are known to show local recurrence. The important prognostic factors are size, presence of tumor free surgical margins, histological grade of tumor and the presence of nodal or distant metastasis. Distant metastasis to lungs, liver, bone or lymph nodes have also been reported.[6] In our case, the leiomyosarcoma was grade 2,with free surgical margins and no lymph node or distant metastasis.

The standard treatment offered in the present case was radical orchidectomy with high ligation of spermatic cord and wide local excision of all nonvital structures. Hemiscrotectomy is indicated if scrotal skin is involved. There is no ideal treatment protocol established in view of the limited number of cases seen world over. Adjuvant radiotherapy as well as chemotherapy have been used in the treatment [2]. In our case, no radiotherapy or chemotherapy was given because of economic constraints. Surveillance for low and intermediate grade leiomyosarcomas of the paratesticular region is advocated.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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