RARE PARATESTICULAR LEIOMYOSARCOMA: A CASE REPORT

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ABSTRACT: The 56-year-old man had been aware of a painless scrotal swelling for about six years. The swelling was becoming larger and painful with backache and sciatica over the past six months before he visited our clinic. It was diagnosed as hydrocele radiologically elsewhere. The man had no history of medical diseases. Physical examination revealed a non-tender, solid mass on the left hemi-scrotum, measuring 10x5x3cms in diameter with no inguinal lymphadenopathy. Subsequent ultrasound abdomen revealed liver metastasis. Radical orchidectomy with high cord ligation was performed with a tumour free margin of 2 cm. The histopathology of the specimen revealed a paratesticular leiomyosarcoma of grade II. On immunohistochemical staining, expression of SMA, muscle specific actin, and desmin was observed, while expression of CD34, S-100 protein, vimentin has been negative. A Stage 4 Grade II leiomyosarcoma was advised radiotherapy and systemic chemotherapy for palliation. Not willing for same was discharged, 3month follow up was uneventful and disease progression was not seen. Role of radiotherapy and chemotherapy more than palliation has not been established due to small sample sizes.
KEY WORDS: Paratesticular tumor; Leiomyosarcoma; Orchidectomy.

INTRODUCTION: Para testicular leiomyosarcoma is very rare and standard treatment consists of radical orchiectomy and high cord ligation[1,2]. We report a case of a Para testicular leiomyosarcoma successfully treated by enucleation. The tumor usually presents with a painless, slow-growing scrotal mass in middle-aged or older men. The diagnosis is always based on histological examination[3,4]. Complete and radical local excision with a tumor-free margin is necessary to achieve the best outcome. Local recurrence and/or distant metastasis with dismal prognosis have been reported[1,3,5], though the tumors are often mistaken as benign tumors. Long-term follow-up is necessary for these patients. We report on a case of para testicular leiomyosarcoma.

Case Presentation: The 56-year-old man had been aware of a painless scrotal mass for about eight years. The tumor was becoming larger and painful with backache and sciatica over the past six months before he visited our clinic. The man had a history of hypertension but no other medical diseases. Physical examination revealed a non-tender, solid mass on the left hemi-scrotum, measuring 10x5x3cms in diameter. It was mobile under the scrotal skin and adhering to the testis. There was no inguinal lymphadenopathy. Ultrasonogram of the scrotum opined a hyperdense mass. Subsequent ultrasound abdomen and sipe work-up revealed no lesions. We performed a Radical orchidectomy with high cord ligation a tumour free margin of 2 cm was obtained [Figure 1]. Grossly, the tumor was pinkish, oval shaped with irregular surface, and elastic. The pathological diagnosis was compatible with a paratesticular leiomyosarcoma. The tumor was composed of highly cellular fascicles of spindle shaped cells with cigar-shaped nuclei and eosinophilic cytoplasm [Figure 2/Figure 3]. Some bizarre giant tumor cells with prominent pleomorphic nuclei were present. And

the mitotic count was high (about 7 mitoses/10 HPF) with mild focal necrosis, suggestive of a grade-2 tumor according to the NCI grading system, probably from the spermatic cord.[2] Immuno-histochemical studies showed the tumor stained positive for actin and desmin, but negative for CD-34 and S-100 [Figure 4]. Postoperative abdominal computed tomography showed no obvious intra-abdominal lesions. We proposed postoperative radiotherapy, but the patient refused. The patient was doing well at nine months of follow-up, with no signs of local recurrence or distant metastasis.

DISCUSSION: Leiomyosarcoma is one of the types of smooth muscle neoplasms. Smooth muscle of skin area is present only in the walls of blood vessels, along hair follicles (arrectores pilorium), and the dartos muscle in the scrotum[1,3,6]. There are two types of leiomyosarcomas, cutaneous leiomyosarcomas arising from the dartos muscle or errector pilorum, and subcutaneous leiomyosarcomas arising from the muscle lining of arterioles and veins in the subcutaneous tissue. Leiomyosarcomas of the scrotal wall usually arise from the dartos muscle, but it is difficult to differentiate the tumor from other leiomyosarcomas of the skin by histology alone[3,7,8]. This case could be either a case of subcutaneous leiomyosarcoma from the spermatic cord or the vessels. Up to 20011, only less than 27 cases of leiomyosarcomas arising from the spermatic cord had been reported[4,8,9,12].There are three typical histological features of leiomyosarcomas, including perpendicularly arranged fascicles of spindle cells with eosinophilic cytoplasm, hyperchromatic blunt-ended nuclei, and scattered paranuclear vacuoles.[5,6,9,14]

On immunohistochemical staining, most leiomyosarcomas express smooth muscle actin, muscle-specific actin, and desmin; while CD-34, myogenin, Ki-67, S-100 protein, and cytokeratin had also been reported in some cases.[1,7,8,12,15] This case was positive only for actin and desmin. The etiology of leiomyosarcomas remains unclear, though some authors suggested local irradiation at childhood could be a potential cause.[5] Clinically most paratesticular leiomyosarcomas present in men of middle or older age as painless, slow-growing scrotal tumors.[3,4,7,9,16] The diagnosis is always based on histological examination. Other rare tumors, including benign leiomyoma, fibrous mesothelioma, various benign fibroustumors and pseudotumors, and fibromatosis, should be considered in the differential diagnosis of paratesticular leiomyosarcomas.[12,13,15,18] Paratesticular leiomyosarcomas are often mistaken as benign tumors preoperatively. The prognosis is usually good if without local recurrence,[4,10,18]though a local recurrence rate of 40% has been reported.[4,8,16] Three prognostic factors have been proposed: the presence of a tumor-free margin, the subtype, and the grades of the tumors. A positive margin at initial excision greatly increased the risk of local recurrence[3,5,9,19,20]. Therefore, a complete radical local excision and high cord ligation with a tumor-free margin is necessary. Postoperative radiation therapy with or without chemotherapy have been reported in some cases, but the benefits remain inconclusive due to smaller number of cases.[1,5,19,20]Regional lymphadenectomy is indicated for those with clinically suspected or proven lymphatic spread. Distal metastasis to the lungs, liver, bone,or lymph nodes has also been reported. Though still controversial, it is generally accepted that distant metastasis more common for leiomyosarcomas of subcutaneous type than for cutaneous type.[3,5,9,13,18]

According to the NCI system,[2,8,10,12,18,20] leiomyosarcomas were classified into three grades according to the degree of mitoses, pleomorphism of nuclei, and focal necrosis.

Grade 1: Tumors with no necrosis, less than 6mitoses in 10 HPF, and mild disseminated pleomorphism.

Grade 2: Tumors with focal necrosis (<15%), more than 6 mitoses in 10 HPF, or obvious nuclear pleomorphism.
Grade 3: Tumors with vast necrosis (>15%), regardless of the number of mitoses or the severity of pleomorphism.

Paratesticular leiomyosarcomas are mostly of low-grade tumors, but high-grade tumors may behave more aggressively. In the large series of 24 cases of paratesticular leiomyosarcomas, none of the cases with grade-1 or grade-2 tumors was dead of disease, while the three cases of grade-3 tumors were all dead of disease during the follow-up period.[12,14,17,19,20]Therefore, long-term follow-up is necessary for patients with this rare tumor.

CONCLUSION: A Paratesticular sarcomas are rare and most of the available information derives from small series or case reports. In most cases, information about tumor grade is lacking, follow-up is short and different types of sarcomas are analyzed together. Therefore, standard treatment for all paratesticular sarcomas consists of radical orchiectomy with high cord ligation. The role of adjuvant therapy is not clear. Overall, 5-year disease-specific survival was 75% [1,3,4] and median interval to local regional relapse was 36 months[2,6,8,9]. Dissemination occurs by hematogeneous metastasis, regional lymph node spread and local extension. This case of paratesticular tumor was a grade II leiomyosarcoma and has a disease-free follow-up period of 9 months.

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FIGURES:

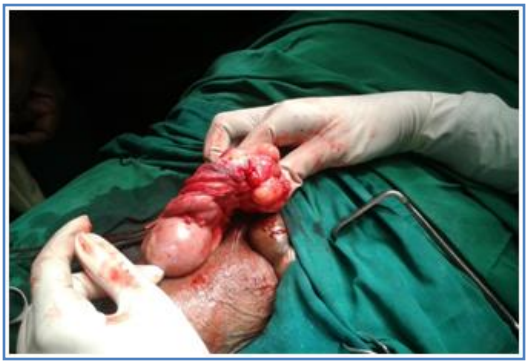

Fig. 1- Tumor in the paratesticular region, proceeded for radical orchidectomy with high Ligation.

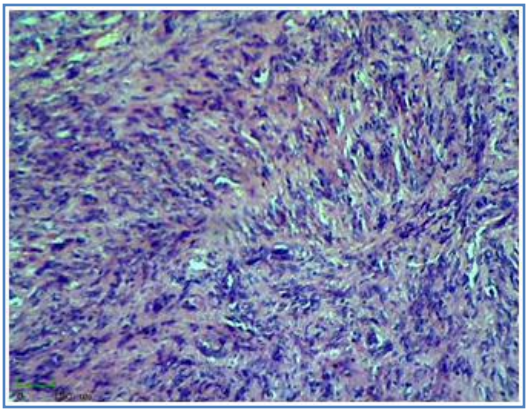

Fig. 2- Low Power: Pallisading pattern

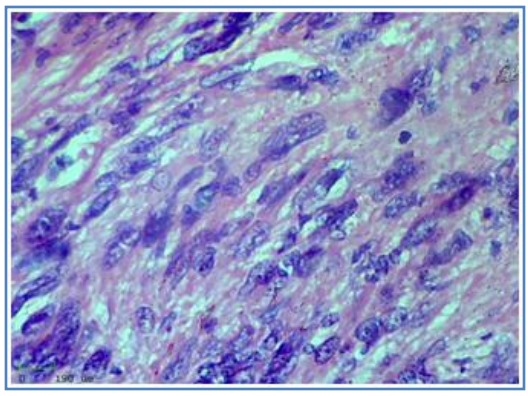

Fig. 3- High Power: Cigar shaped pattern.

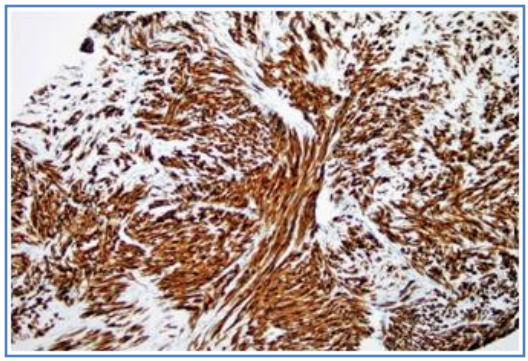

Fig. 4- SMA CYTOPLASMIC POSITIVITY

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