Subcutaneous leiomyosarcoma of scrotum presenting as an exophytic mass: An unusual presentation

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ABSTRACT

Paratesticular leiomyosarcoma originates from testicular tunica (48%), spermatic cord (48%), epididymis (2%) and dartos muscle, as well as subcutaneous tissue of the scrotum (2%). Leiomyosarcomas of the scrotum, not involving the testis, epididymis or spermatic cord, are rare, and belong to the group of subcutaneous superficial leiomyosarcomas. To the knowledge of the authors, less than 10 cases of leiomyosarcoma of the scrotum have so far been reported from India. The tumor usually presents as a painless, slow-growing scrotal mass in middle-aged or elderly men. The current approach is wide local excision, often with adjuvant therapy. The prognosis is usually good following complete excision, though a local recurrence rate of 40% has been reported. Long term follow-up is, therefore, necessary to monitor for recurrence. Herein we present the case of 35-year-old male who presented with an exophytic scrotal mass. Histopathological and immunohistochemical findings of the mass were consistent with leiomyosarcoma.

Key words: Leiomyosarcoma, scrotum, subcutaneous

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INTRODUCTION

Over 95% of all paratesticular leiomyosarcomas are located in the spermatic cord or epididymis, and their location in the scrotal skin or subcutaneous tissue is exceptionally rare. It has been suggested that the rarity of leiomyosarcomas of the scrotum is such that the general practitioner would usually see such a tumor once every 20 years.[1] These tumors usually present as firm, rubbery, nontender, irregular masses. Scrotal leimyosarcomas are slow-growing tumors that tend to be present for years.[2] The diagnosis is always based on histological examination.[3] It is best treated by wide local excision.[4] Radiotherapy/chemotherapy should supplement and should not substitute complete excision of the lesion.[1] Chemotherapy may be effective in a selective group of patients who refuse surgery. Long term follow-up is essential, because of the risk of delayed local recurrence and distant metastasis.[2]

CASE REPORT

A 35-year-old male presented with left-sided exophytic scrotal mass [Figure 1] that had been steadily enlarging over last 6 months. It was nontender, firm, nodular and subcutaneous in location, with ulceration of overlying skin. The mass was mobile, and not adherent to the underlying testis, epididymis and spermatic cord. There was no inguinal or abdominal lymphadenopathy. The testis and epididymis on both sides were normal. There were no local or systemic signs and symptoms suggestive of any sexually transmitted disease. There were no other complaints suggestive of any other local or systemic illness. Past medical history of the patient was unremarkable. There was no history of previous local irradiation or long term anabolic steroid abuse. There was no history of high-risk sexual behavior. General and systemic examination was within normal limits. The clinical differentials considered included warty carcinoma/sarcoma, nodular stage of Kaposi sarcoma, leiomyoma with secondary ulceration of the overlying skin and unusually large mass formed by coalesced benign warts.

Hematological examination and routine biochemical investigations were within normal range. He was seronegative for human immunodeficiency virus and venereal disease

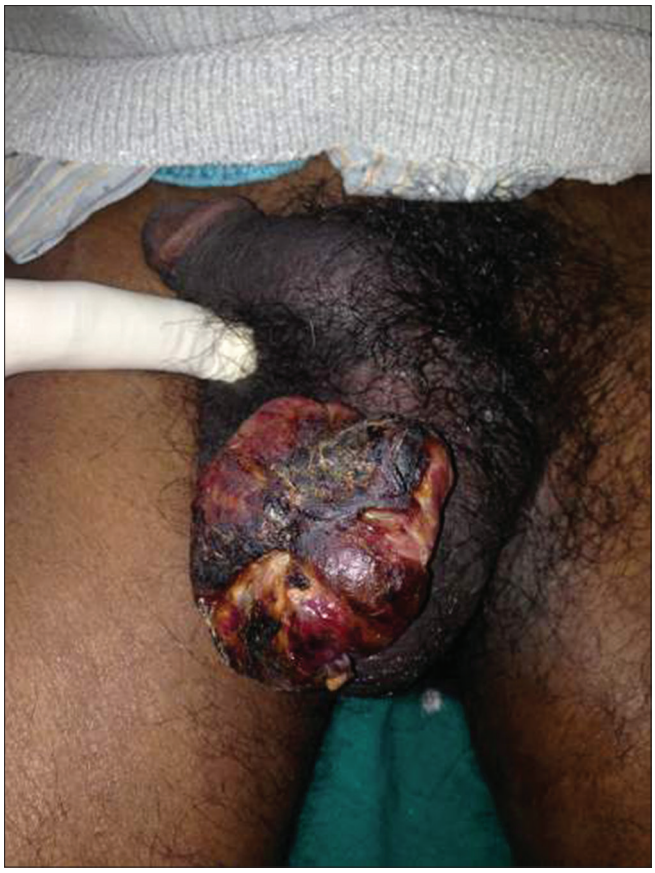

Figure 1: Left-sided exophytic scrotal mass with ulceration of overlying skin

research laboratory test. Culture of urine and urethral swab was negative. Ultrasound (USG) of the mass revealed a well-circumscribed hypoechoic lesion 7 cm × 6 cm × 3 cm in the subcutaneous plane without any calcification. The underlying and contralateral testes were normal.

Wide excision of mass was done under general anesthesia, and the specimen was subjected to histopathological examination. On gross examination, the specimen revealed an ulcerated exophytic growth measuring 7 cm × 6.5 cm × 3 cm [Figure 2a]. Cut section of the mass was grey white [Figure 2b]. Microscopic examination revealed a spindle cell tumor in the subcutaneous location [Figure 3a]. The tumor cells were arranged in fascicles containing eosinophilic cytoplasm and cigar-shaped nuclei revealing nuclear pleomorphism, prominent nucleoli and 3–5 mitotic figures per high power field. A few bizarre pleomorphic giant tumor cells were also present [Figure 3b]. Immunostaining showed the presence of vimentin, desmin and smooth muscle actin [Figure 3c]. Calretinin, CD34, S 100 and desmin [Figure 3d] were negative excluding the diagnosis of malignant mesothelioma, fibromatosis, liposarcoma and rhabdomyosarcoma respectively. A diagnosis of leiomyosarcoma was made. The macroscopic margins taken were free from tumor infiltration.

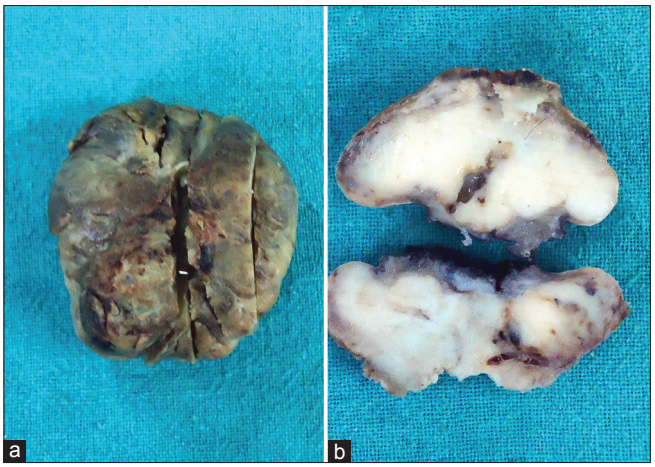

Figure 2: Gross specimen of the tumor revealing an ulcerated exophytic growth measuring 7 cm × 6.5 cm × 3 cm (a) with grey white cut surface (b)

His postoperative period was unremarkable. He was started on six cycles of vincristine, doxorubicin and cyclophosphamide, given three weekly. Computerized tomography (CT scan) of chest and abdomen was performed to rule out lung and liver involvement, which did not reveal any evidence of metastasis. Whole body bone scan was also within normal limits. Radiological investigations (local USG, CT chest, CT abdomen and bone scan) were repeated at an interval of six months. There were no signs of local recurrence or distant metastasis up to one year of follow up.

DISCUSSION

Soft tissue sarcomas account for 1% of all malignancies.[2] Leiomyosarcomas constitute 10–20% of soft tissue sarcomas.[5] Subcutaneous leiomyosarcomas account for 1–2% of all superficial soft tissue malignancies. They arise from smooth muscle in the walls of arterioles and veins.[2] Majority of the paratesticular leiomyosarcomas are located in the spermatic cord or epididymis and their location in the subcutaneous tissue of the scrotum is exceptionally rare with approximately 40 reported cases in the literature.[1]

Leiomyosarcoma of the scrotum is a rare tumor.[4] It is mostly diagnosed in the sixth decade, and more than 80% of patients are over 40 years old. Like other mesenchymal tumors of this region, leiomyosarcoma manifests as a painless mass without hydrocele and the disease may be symptomatic in <1 year.[6] They usually present as firm, rubbery, nontender, irregular masses that tend to be slow-growing tumors and evolve over years. These grow by radial expansion, infiltrating the local tissues as they proliferate. They lack a surrounding capsule so that an excisional biopsy with a macroscopically clear margin will often leave microscopic tumor behind.[1] The size of tumor is usually between 2 and 9 cm with a mean of 5 cm.[6] The etiology

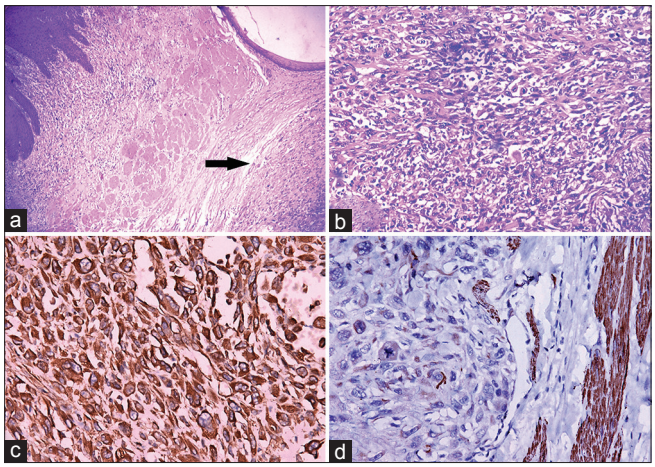

Figure 3: Microphotograph revealing scrotal skin with a tumor mass in the subcutaneous location as highlighted by arrow (a; H and E, ×40) The spindle shaped tumor cells arranged in fascicles, revealed eosinophilic cytoplasm and cigar-shaped nuclei showing nuclear pleomorphism, prominent nucleoli and 3–5 mitotic figures per high power field along with a few bizarre pleomorphic giant tumor cells (b; H and E, ×100) The tumor cells were positive for smooth muscle actin and negative for desmin (c and d respectively; IHC, ×200)

of leiomyosarcomas remains unclear, though some authors suggest local irradiation during childhood as a potential cause.[3]

Confirmation of the diagnosis of leiomyosarcoma is based upon histological examination of biopsy specimen, which reveals spindle cells with cigar-shaped nuclei arranged in interweaving fascicles. The diagnosis of malignancy in leiomyosarcoma is based on the mitotic rate of 2–10 mitoses/HPF, although the presence of nuclear pleomorphism, vascular invasion, tumor depth, and infiltration and the percentage of tissue necrosis are also considered. On immunohistochemistry, leiomyosarcomas are positive for actin and desmin.[1] Other rare tumors, including benign leiomyoma, fibrous mesothelioma, various benign fibrous tumors and pseudotumors, and fibromatosis, should be considered in the differential diagnosis of paratesticular leiomyosarcomas.[7]

These grow by radial expansion, infiltrating the local tissues as they proliferate.[4] The mode of spread of leiomyosarcoma is primarily hematogenous to lung, liver, and bone. The prognosis of leiomyosarcoma depends upon the size, depth and grade of the tumor and presence or absence of distant metastases.[8] There is limited data on behavior of leiomyosarcoma presenting in the paratesticular region due to relatively small number of cases reported in the literature. Fisher *et al*. observed that 30% of the patients had recurrence, 30% had metastases (lymph nodes, lungs, liver) and 30% died (all grade 3 tumor patients) after 4 year follow-up.[7]

The tumor should be treated by wide excision with a clear margin of at least 10 mm. There is a place for adjuvant therapy (radiotherapy/chemotherapy) in some cases.[1] One of the controversial issues regarding the surgical management of scrotal leiomyosarcomas has been the definition of an adequate resection margin.[4] Wide excision of scrotal leiomyosarcomas with a clear margin of at least 10 mm was associated with a better outcome in comparison to tumors with involved margin or <10 mm clear margin.[1] Tumor recurrence or failure of margin clearance necessitate further excision, which is fraught with problems.[4]

Owing to the small number of patients in the literature, definitive data regarding the role of adjuvant therapy is limited.[2] The role of chemotherapy is not well established. However, it may be effective in the selected group of patients who refuse surgery. It has been further suggested that it might have a role in abrogating the tumor's hematogenous metastatic potential.[1] Chemotherapy with gemcitabine, paclitaxel, vincristine, doxorubicin and actinomycin-D has been used with limited success.[2] It may be noted that chemotherapy or radiotherapy can only be used as adjuvant therapy, and it should not be substituted for radical surgical excision.[1] Inguinal lymph node dissection is not advocated, unless a high degree of suspicion is present for lymph node metastasis.[2] Since the tendency of hematogenous metastasis is high, the effect of retroperitoneal lymphadenectomy is unclear and generally not suggested.[6] Late local recurrence and distant metastases occurs in some cases, therefore long term follow-up is recommended.[1]

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