INTRODUCTION

Paratesticular leiomyosarcoma (LMS) not involving epididymis or spermatic cord is considered as subcutaneous or cutaneous superficial LMS. Over 95% of all paratesticular LMSs are located in the spermatic cord or epididymis. Johnson, reported the first known case of LMS of the scrotum in 1987[1] and since then, <40 instances of LMS of the scrotum have been reported.[2] Cutaneous or subcutaneous LMS is usually a solitary, robust, skin-colored, or reddish-brown nodule. Its size varies between 2 cm and 9 cm.[3] The prognosis depends on the depth of involvement and mitotic activity. Immunohistochemistry differentiates LMS from other smooth muscle tumors. The panel of antibodies differentiates LMS from other spindle cell tumors. The main differential diagnosis of LMS is from monophasic synovial sarcoma, malignant peripheral nerve sheath tumor, solitary fibrous tumor, spindle cell rhabdomyosarcoma, and spindle cell melanoma.

The following panel of antibodies is used to differentiate them [Table 1]. Many new immunohistochemical markers have been developed over the years and have improved with time, but no one marker is 100% sensitive or specific for a given diagnosis. Clinical and radiographic correlation is still required to make a correct diagnosis. Wide local excision (WLE) with tumor-free margin is the treatment of choice. We report a case of LMS scrotum in a 45-year-old patient presenting as a painless scrotal swelling on the left hemiscrotum.

CASE REPORT

A 45-year-old male presented with a painless external scrotal swelling on the left hemiscrotum [Figure 1], which has been steadily increasing in size for the past 6 months. He was a chronic smoker with a history of 25 cigarettes/day and had no significant medical or surgical history. There was no history of previous irradiation to the region or

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abuse of anabolic steroids. His family history was also not significant. His vitals were within the normal limits.

On physical examination, the swelling was a nontender, firm subcutaneous mass with ulceration of overlying skin [Figure 1]. It was mobile and not adherent to the underlying structures. The testes, epididymis, and the spermatic cords were clinically normal, and there was no inguinal lymphadenopathy. Hematological and biochemical investigations were within the normal range. Contrast-enhanced computed tomography (CT) scans of the abdomen and chest were normal. In view of the malignant potential of the mass, it was surgically removed with extensive local excision, including a 2 cm margin. No adhesion between the tumor and the neighboring structures was observed during the excision. Laxity of the scrotal skin allowed primary closure. Histopathological examination of hematoxylin and cosin-stained smears revealed a spindle cell tumor. The tumor cells were arranged in intersecting fascicles containing eosinophilic cytoplasm and oval to elongated nuclei revealing nuclear pleomorphism, hyperchromasia, prominent nucleoli, and 2–4 mitotic figures/10 high-power fields [Figures 2 and 3]. A few bizarre pleomorphic giant tumor cells were also seen [Figure 4]. The margins were negative, and there was no residual tumor. Immunohistochemistry was advised and turned out to be positive for h-caldesmon [Figure 5] and negative for myogenin suggesting the diagnosis of LMS. The patient is on regular follow-up with biannual clinical examination, ultrasound of abdomen and CT scan of the chest for the past 4 years without the recurrence of the lesion.

DISCUSSION

Sarcoma of the scrotum, excluding extension from the spermatic cord, is extremely rare. Pure scrotal cutaneous LMS is rare. Less than 40 cases have been reported in the literature to date. Nearly 95% of LMSs of the scrotum are located in the epididymis or spermatic cord. Moreover, a cutaneous or subcutaneous location is infrequent. It arises from the dartos muscle. The age of presentation reported in the literature ranges from 35 to 89 years. Cutaneous LMS presents as a firm, single, skin-colored, or reddish-brown nodule of size between 2 cm and 9 cm.[3] The most common location for LMS is the lower extremities. The cause remains unclear, and few authors have suggested local irradiation during childhood as a potential cause.[4] LMS comprised of spindle cells on

![Figure 1: Cutaneous leiomyosarcoma of the scrotum with ulceration of the overlying skin](image1)

![Figure 2: Spindle cell tumor showing intersecting fascicles](image2)

| Table 1: Differential diagnosis of spindle cell tumors |
|-----------------------------------------------|
| **IHC** | LMS | Monophasic synovial sarcoma | MPNST | Spindle cell rhabdomyosarcoma | Spindle cell melanoma |
| h-caldesmon | Positive | | | | |
| TLE-1 | Positive | | | | |
| SOX10 | | Positive | | | |
| SS18 | | Positive | | | |
| desmin | Positive | | | | |
| S-100 | Positive | | | | |
| MYOD1 | Positive | | | | |

MPNST: Malignant peripheral nerve sheath tumor, TLE-1: Transducin-like enhancer of split 1, SS18: Synovial sarcoma 18, Myo D1: Myogenin D1, LMS: Leiomyosarcoma, IHC: Immunohistochemistry
histopathological examination. Immunohistochemically, the tumor cells are positive for smooth muscle actin, desmin, and h-caldesmon.\(^5\) Other immunohistochemical markers in addition to these are Vimentin and muscle-specific actin, and LMS is usually negative for S-100 protein and cytokeratin. Prognosis depends on the depth of involvement and mitotic activity. Recurrence is generally associated with inadequate excision. The risk of metastasis is related to size, and a tumor under 5 cm is less likely to metastasize. LMS may metastasize through either hematogenous (lung) or lymphatic route (regional lymph node). Recurrence is more challenging to treat and tends to metastasize early. WLE ensuring a tumor-free margin is recommended.\(^6\) Some authors have used Mohs micrographic surgery (MMS) for margin control successfully, as it appears to be the strongest predictor of clinical recurrence.\(^7\) Conventionally, WLE has empirically and variably maintained margins of 2–5 cm, and their meta-analysis shows that the precision of MMS did produce markedly lower recurrence in the 14 reports comprising 48 cases that they studied.\(^7\) The patients treated with MMS showed a mean recurrence rate of 2.08%–6.25% with a mean follow-up period of 1570.9 days.\(^7\) The reported recurrence rate with WLE is approximately 30%–50%.\(^7\) There were no reports of distant metastases in patients who were treated with MMS.\(^7\) Long-term follow-up is recommended for cases treated either by WLE or by MMS because of the reported risk of late local recurrence.\(^7\)

**CONCLUSION**

Pure scrotal cutaneous LMS are exceptionally rare, and a high index of suspicion is required for diagnosis. Immunohistochemistry is very useful for diagnosis and to differentiate it from other spindle cell tumors. Wide excision is to be performed on clinical suspicion to avoid resurgery. MMS is associated with markedly lower rates of recurrence compared to reported rates of recurrence after WLE.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that their name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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