Recurrent Leiomyosarcoma Scrotum of Dartos Muscle - A Nonplussed Differential in Scrotal Swelling

Surya Ram G. K.,1 Emil Phinehas2, Authy K.3

1, 2, 3 Department of General Surgery, Pondicherry Institute of Medical Sciences, Kalapet, Pondicherry, India.

PRESENTATION OF CASE

A 63-year-old gentleman presented with 2 months history of painless, progressively increasing lump on the anterior surface of the left side of the scrotum. He had similar swelling 4 years back at the same site, following which he underwent surgical resection elsewhere and was told to have sarcoma. He could not produce health records of his previous treatment. The lump was a 2 × 2 cm in size subcutaneous in location, on the left hemiscrotum. It was non-tender, firm, and was not fixed to the testis or underlying structures. There was no inguinal lymphadenopathy. Ultrasound of the scrotum revealed a well defined heteroechoic lesion 2 x 2 cm in the left hemiscrotum. A contrast-enhanced computed tomography scan from neck to pelvis showed no distant metastases.
Clinical Diagnosis
Scrotal sarcoma

Differential Discussion
Liposarcoma, rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma, malignant fibrous histiocytoma, desmoplastic round cell tumour. Rare tumours like dermatofibrosarcoma protuberans, benign leiomyoma, fibrous mesothelioma, various benign fibrous tumours and pseudotumors, and fibromatosis.

Pathological Discussion
He underwent wide local excision of scrotal swelling with a 1.5 cm margin. The histology report showed a grey-white nodular lesion measuring 0.6 × 0.8 cm., arising from the dartos layer of the scrotum. The tumour was composed of interlacing fascicles and bundles of spindle-shaped cells. The cells had enlarged elongated blunt-ended nuclei showing moderate atypia with vesicular to hyperchromatic nuclei, few showing prominent nucleoli, a few large bizarre cells, and multinucleated cells were noted with focal myxoid change, mitoses were 4-5/10 hpf, suggestive of leiomyosarcoma (LMS) arising from dartos muscle, stage IA. He was advised for regular follow-up and currently has been disease-free for 4 months following surgery.

Microscopy
The tumour was composed of interlacing fascicles and bundles of spindle-shaped cells.

Figure 1.

Figure 2.

Discussion of Management
Genitourinary leiomyosarcomas constitute about 2.1 % of soft tissue sarcomas and usually arise from the urinary bladder, kidney, or prostate. Paratesticular LMSs originate from testicular tunica (48 %), spermatic cord (48 %), epididymis (2 %).[1] Topographically LMSs are divided into three groups: LMS of the deep soft tissue, LMS of the cutaneous and subcutaneous tissue, and LMS of vascular origin.

Leiomyosarcomas constitute 10 %–20 % of soft tissue sarcomas and most often develop in the uterus, gastrointestinal tract, or retroperitoneum. Leiomyosarcomas are malignant mesenchymal neoplasms that arise from smooth muscle, and their aetiology is unknown. Scrotal leiomyosarcomas are very rare. They arise between the fourth and eighth decades of life as painless, slowly growing skin lesions of 2-9 cm in diameter. They are described as smooth muscle tumours of the skin that are found in the muscle linings of arterioles and veins in the subcutaneous layer. It is extremely rare for them to arise in the scrotal skin or subcutaneous layer, and only 38 cases of leiomyosarcoma of the scrotum have been reported.

LMSs of subcutaneous tissue or dartos muscle of the scrotum are extremely rare and constitute only 2 % of paratesticular LMSs. The literature on the management and prognosis of LMS arising from dartos muscle is limited to a few case reports.[1–3] They are mostly diagnosed between the 4th and 8th decades of life, presenting as painless lumps.[2] and slowly growing skin lesions.[4] The differential diagnoses of scrotal sarcoma include liposarcoma, rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma, malignant fibrous histiocytoma, desmoplastic round cell tumour. Rare tumours like dermatofibrosarcoma protuberans, benign leiomyoma, fibrous mesothelioma, various benign fibrous tumours and pseudotumors, and fibromatosis are benign lesions which could present as paratesticular lesions.[3] Histological examination clinches the diagnosis. Immunohistochemistry reveals positivity for actin and desmin. CD34 and cytokeratin expression have also been reported in some cases. It has been observed that 30 % of the patients had a recurrence, 30 % had metastases (lymph nodes, lungs, liver), and 30 % died after a 4-year follow-up. A surgically positive margin detected at the first excision dramatically increases the risk of local recurrence.

Over 95 % of paratesticular leiomyosarcomas originate from the spermatic cord or the epididymis, dartos muscle, or subcutaneous scrotal muscle. Superficial leiomyosarcomas are rare malignant smooth muscle tumours. They account for 4 – 6.5 % of all soft tissue sarcomas, less than 2–3 % of cutaneous soft tissue neoplasms, and 0.04 % of all cancers.[5] Leiomyosarcoma of the scrotum, which arises in the

High Power View
The cells had enlarged elongated blunt-ended nuclei showing moderate atypia with vesicular to hyperchromatic nuclei, few showing prominent nucleoli, a few large bizarre cells, and multinucleated cells were noted with focal myxoid change, mitoses were 4-5/10 hpf, suggestive of leiomyosarcoma (LMS) arising from dartos muscle, stage IA.
subcutaneous scrotal layer, is a rare type of tumour. Subcutaneous scrotal tumours have been reported to be associated with an increased risk of local recurrence and distant metastasis.[6] To the best of our knowledge, less than 38 cases of leiomyosarcoma of the scrotum have been reported in the literature.

Local infiltration to surrounding tissues results in the radial expansion of the tumour. Owing to the small number of patients in the literature, definitive data regarding the role of adjuvant therapy are limited. They may play a role in abrogating the tumour’s hematogenous metastatic potential. Inguinal lymph node dissection is not advocated unless a high degree of suspicion of metastasis is present. Complete surgical resection with a negative margin is the standard of care. The data for adjuvant therapy is limited, though adjuvant radiotherapy has been reported to reduce recurrences in some reports.[6][7] Our patient had a sub centimetric tumour. After discussion, we opted to follow up after surgery with adjuvant therapy. Prophylactic lymph node dissections were not advocated in these tumours. Long term follow up is required because of the reported risk of late local recurrence.[8]

Leiomyosarcomas can rarely metastasize to the lungs, liver, and bone hematogenously, and the prognosis of leiomyosarcoma depends on the size, depth, and grade of the tumour and the presence/absence of distant metastasis at diagnosis. Wide excision is required for the treatment of localized leiomyosarcoma of the scrotum because of the risk of recurrence/distance metastasis and long-term follow-up is required because of the reported risk of late local recurrence.[9]

To conclude, LMS of dartos muscle is an extremely rare tumour presenting as a scrotal lump. This report aims to brief the existing literature on this rare tumour. Surgical excision is the treatment of choice, the role of adjuvant therapy is controversial.

**Final Diagnosis**

Recurrent leiomyosarcoma scrotum of dartos muscle.

**REFERENCES**


