

### THE RAREST FORM OF PARATESTICULAR LEIOMYOSARCOMA

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Paratesticular tumours make up less than 5% of intra-scrotal tumours and of these, approximately 30% are malignant tumours with sarcomas accounting for the majority.

Leiomyosarcoma is a malignant tumour arising from soft tissues containing smooth muscle. They are reported as comprising between 5-30% of paratesticular sarcomas.<sup>1,2</sup> It is extremely rare that they arise from tissue other than the spermatic cord or epididymis.

The authors describe such a case of paratesticular leiomyosarcoma in a 54-year-old man who presented with a six-month history of a painful testis and who subsequently underwent radical inguinal orchidectomy and high ligation.

#### CASE REPORT

A 54-year-old man presented to his doctor with a 6-month history of a painful left testis and was referred to urology. On examination the left testis was palpable anterior to the scrotum with a hard, craggy mass felt posteriorly.

Scrotal ultrasound described a 5.9 mm left upper pole mass of mixed echogenicity and increased blood flow from the left testis. Tumour markers were normal.

Following multidisciplinary team discussion, a repeat ultrasound was performed by a consultant radiologist with urology expertise. The results indicated

a large extratesticular mass likely arising from scrotal layers of uncertain nature but soft tissue sarcoma not ruled out.

Computed tomography of the thorax, abdomen, and pelvis showed no evidence of metastasis or lymphadenopathy.

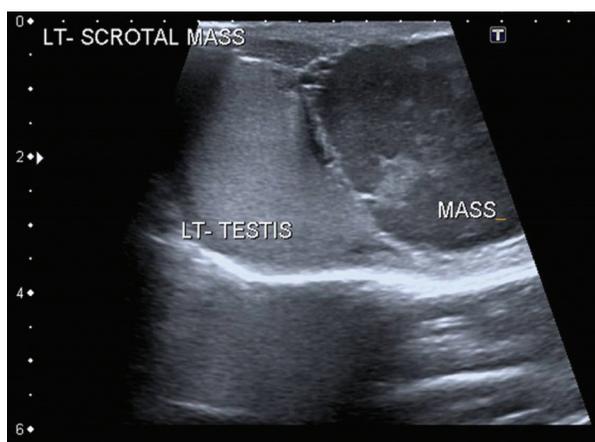
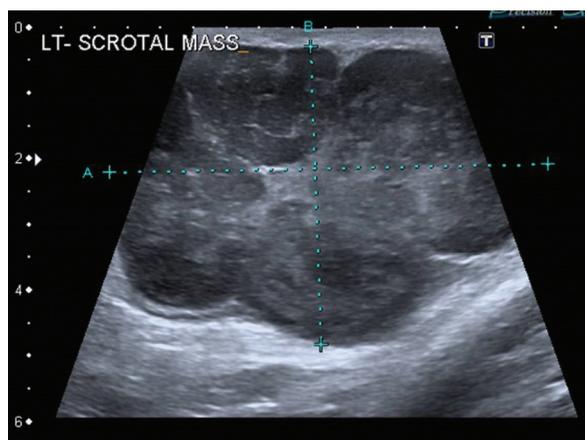
After a multidisciplinary team assessment, the patient was counselled for a left radical inguinal orchidectomy. Intra-operatively the tumour was intra-tunical and appeared closely adherent to the testis and spermatic cord which made for a more challenging orchidectomy but the patient made an uneventful recovery and was discharged on post-operative day 1.

Macroscopically, there was a firm, white lobulated tumour measuring 72 × 50 × 50 mm, compressing the testis which measured 40 × 10 mm.

Histopathological assessment identified a malignant tumour of the paratesticular region with no connection seen to the testis, rete testis, epididymis, or spermatic cord.

Morphological features included plump, spindle-shaped cells arranged in fascicular, storiform, and whorled architectural patterns. The nuclei showed coarse chromatin with moderate eosinophilic cytoplasm and exhibited moderate to marked atypia with foci of moderate to severe pleomorphism with multinucleate and large pleomorphic tumour cells. Numerous

**FIG. 1** and **FIG. 2** Scrotal ultrasound demonstrating left paratesticular mass.



mitoses were noted with up to 30 in 10 high-power field (HPF) including several abnormal forms with no evidence of necrosis. Immunohistochemistry was positive for moderate expression of smooth muscle actin and desmin. The features were suggestive of sarcoma and review by a sarcoma specialist confirmed that the findings were consistent with a leiomyosarcoma, grade 3 (Table 1).

### DISCUSSION

Paratesticular tumours arise from a complex anatomical region including the spermatic cord, testicular tunics, epididymis and vestigial remnants. Paratesticular tissues are derived from a combination of epithelial, mesothelial and mesenchymal cells and as a result the tumours form a heterogenous group which exhibit a broad range of behaviours and histological appearances.<sup>1,4</sup> They can present a diagnostic challenge as they are often clinically and radiologically indistinguishable from a testicular mass.

Around 30% of paratesticular tumours are malignant with sarcomas accounting for approximately 90%, however still comprising less than 5% of all sarcomas and 2% of urological malignancies.<sup>5</sup>

Leiomyosarcomas are malignant tumours which develop from undifferentiated smooth muscle cells of mesenchymal origin anywhere in the body, rarely presenting as primary paratesticular sarcomas. When tumours are paratesticular, they most commonly arise from the spermatic cord where they originate

**TABLE 1** French Federation of Cancer Centres Sarcoma Grading System

Score	Differentiation	Necrosis	Mitoses per 10 HPF
0	—	Absent	-
1	Closely resembles normal adult mesenchymal tissue	<50%	0–9
2	Histological typing is certain	<50%	10–19
3	Embryonal and undifferentiated sarcomas, sarcomas of doubtful type	-	≥20

Grade 1 = total score 2–3; Grade 2 = score 4–5; Grade 3 = score 6–8; HPF = high-power field. Adapted from Galosi AB et al.<sup>3</sup>

from cremasteric muscle and the vas deferens. Less frequently, the epididymis is the origin, arising from the smooth muscle surrounding the basement membrane of the epididymis canal. The scrotal form occurs least frequently, from the dartos layer. This case demonstrates the *most rare* leiomyosarcoma subtype arising separately from the tissues highlighted above.

Clinical presentation is commonly as a painless, slowly enlarging mass with a discrete, nodular feel and may be associated with a hydrocele.<sup>6</sup> Age at presentation is most commonly between 50 and 70 years.<sup>7</sup> Etiology of paratesticular LMS remains unclear though some authors have suggested that exposure to childhood radiation is a risk factor.

Ultrasound is the initial investigation of choice<sup>8</sup> and usually reveals a solid, heterogenous mass that may demonstrate increased vascularity on colour Doppler<sup>9</sup>; however, the accuracy of the scan is operator-dependent; in this case the patient required a repeat scan to confirm that the mass was paratesticular. Computed tomography can be used to assess the mass but is of most value of assessing for metastatic disease.

The diagnosis is made based on typical histological features of leiomyosarcoma, including spindle cells arranged in fascicular fashion, eosinophilic cytoplasm, blunt ended nuclei, cellular atypia and variable mitotic activity.<sup>10</sup> Immunohistochemical staining is frequently positive for smooth muscle actin and desmin and there may be expression of CD-34 and cytokeratin.

Radical inguinal orchidectomy is the widely accepted standard treatment, though due to the rarity of such tumours the natural history is not clear. There is no consensus on the role of lymph node dissection, adjuvant radiotherapy or chemotherapy. Local recurrence of sarcomas is common<sup>1</sup> and patients require regular follow-up to monitor for recurrence.

### CONCLUSION

This patient had a high-grade leiomyosarcoma originating from an extremely rare location within paratesticular tissue – separate to the spermatic cord, epididymis and dartos. The tumour exhibited typical clinical, radiological, and histopathological features for this type of sarcoma and subsequently underwent complete resection.

The patient was referred to a specialist Abdominal and Retroperitoneal Sarcoma Unit for further follow-up; however, there is currently no standardized protocol for duration of surveillance imaging to identify recurrence due to the rarity of the disease.

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