A case report of uncommon paratesticular liposarcoma of spermatic cord presenting as a scrotal mass

Layla Zeitouni a, Shahinur Rahman b, Kimberley Chan b, Khin Thway c, Ajay Arora d, Mohamed Y. Hammadeh b,*

a University of Glasgow, School of Medicine, Dentistry & Nursing, UK
b Urology Department, Lewisham and Greenwich NHS Trust, London, UK
c Sarcoma Unit, The Royal Marsden Hospital, London, UK
d Radiology Department, Princess Royal University Hospital, King’s College Foundation NHS Trust, London, UK

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ABSTRACT

Paratesticular liposarcomas are uncommon malignancies, often misdiagnosed preoperatively, that present as a painless scrotal mass. There is no universal consensus on their management due to scarcity of reported cases. Early detection and prompt surgical intervention provide the best disease outcome. We present the case of a 74-year-old man with a painless left testicular mass. Ultrasound and CT scan showed a paratesticular tumour. He underwent left high inguinal orchidectomy. Histopathological, immunohistochemical and cytogenetics studies confirmed the diagnosis of a Grade 2 well differentiated paratesticular liposarcoma with clear surgical margins. The case was referred to our regional sarcoma centre and currently undergoes regular follow-up.

Introduction

Primary paratesticular tumours are uncommon, accounting for 7% of all intra-scrotal tumours. Liposarcomas account for 5% of these tumours. Little is still known about their exact pathogenetic mechanisms, but they arise de novo in the connective tissue surrounding the testis, epididymis, and spermatic cord, and do not arise from malignant transformation of a pre-existing lipoma. The spermatic cord is the most common site, accounting for 76% of liposarcomas.

We present an uncommon case of a paratesticular liposarcoma of the spermatic cord. We reviewed the English literature on diagnosis and management of paratesticular liposarcoma.

Case description

A 74-year-old patient presented with one-month history of painless left testicular swelling. Clinical examination showed soft tissue mass anterior to the left testis with normal right testis and unremarkable abdominal examination. He had normal α-fetoprotein and β-HCG.

Scrotal ultrasound scan showed a left 5.5cm extra testicular mass with increased heterogeneity and vascularity and normal right testis (Fig. 1).

Chest, abdomen, pelvic and scrotal CT scan confirmed a left mixed density paratesticular mass. (Fig. 2). Chest was normal and there was no pelvic or abdominal lymphadenopathy. There was some abnormality in the small bowel near the ileocecal valve with calcification, nature of which was unclear.

The patient underwent a left high inguinal orchidectomy. Intraoperatively, oedema and inflammation surrounding the mass was seen. The tumour was resected intact along with the left testicle and spermatic cord.

Macroscopically the tumour appeared as a solid, mucoid mass, around 65 × 35 × 40 mm without any infiltration into epididymis or testis. Microscopically, the testis, epididymis, and spermatic cord did not show any significant abnormality. Histologically, the paratesticular mass was a cellular neoplasm, composed of fascicles of oval to spindle-shaped nuclei with mild to moderate pleomorphism and fibrillary cytoplasm in myxocollagenous, stroma with prominent interspersed thin-walled curvilinear vessels. The mitotic index was 2–3/10HPF, and there was no necrosis (Fig. 3). The surrounding adipose tissue contained fibrous septa with plump focally atypical spindle cells, in keeping with a component of well-differentiated liposarcoma. The resection margin of the spermatic cord was free of tumour. Fluorescence in situ hybridization demonstrated co-amplification of the MDM2 gene at 12q15 and of...
Paratesticular mass with mixed density, is separate from the testes, with some fat contents within. No nodal disease seen in the inguinal, iliac, or retroperitoneal region.

Fig. 1. US showing a paratesticular mass in the left hemiscrotum with mixed echogenicity, not arising from the epididymis. Shows vascularity, appearances are non-specific, but suspicious of malignancy.

Fig. 2. Paratesticular mass with mixed density, is separate from the testes, with some fat contents within. No nodal disease seen in the inguinal, iliac, or retroperitoneal region.

Fig. 3. Dedifferentiated liposarcoma: Histologically, the tumour is composed of fascicles of mildly to focally moderately atypical cells with spindle to ovoid nuclei and scanty mitoses with occasional interspersed mature adipocytes on 100X magnification. MRI did not detect any residual disease.

The patient underwent a colonoscopy to investigate the suspicious lesion near terminal ileum; and the biopsy was normal.

He was referred to our regional sarcoma centre and discussed in their multidisciplinary meeting. As the tumour was 5.5cm and grade 2, the consensus was to observe clinically and follow up with clinical examination and abdomen and pelvic CT scan, thus reserving radiotherapy in case of relapse. At 10 months follow up the patient remains tumour free.

Discussion

Fewer than 200 cases of paratesticular liposarcomas have been reported in the literature. Liposarcomas comprise around 20% of all paratesticular tumours, and 5% of all paratesticular sarcomas. They are more frequently seen age 50–60 years.

There are three main subtypes of liposarcomas: well-differentiated and dedifferentiated liposarcoma, which are recognised to constitute a histologic and behavioural spectrum of one disease entity, myxoid liposarcoma now incorporating ‘round cell’ (high-grade myxoid liposarcoma), and pleomorphic liposarcoma. Well-differentiated liposarcoma is common (40–50%) with slow growth and good prognosis; it can recur but is not able to metastasise.

The majority of paratesticular tumours (76%) arise in the spermatic cord, as in our case. Due to variance in clinical presentation, liposarcomas are often misdiagnosed as an inguinal hernia, hydrocele, scrotal lipomas and epididymitis. In this case, the preoperative suspected diagnosis was a malignant scrotal mass.

Ultrasound can identify a solid hypoechoic lesion, however, cannot distinguish between a lipoma and liposarcoma, especially if it is well differentiated. CT scan is helpful in establishing tumour location, staging and follow-up. In our case, CT scan showed a mixed density paratesticular mass, and was informative in staging, allowing us to proceed to radical scrotal surgery. MRI is the gold standard for imaging when suspecting a liposarcoma pre-operatively. In our case, liposarcoma was not suspected, therefore an MRI was not carried out pre-operatively.

Most liposarcomas exhibit a tendency to extend locally and spread through haematogenous or lymphatic route. Retroperitoneal lymph node dissection is usually done in widespread metastatic disease. Kamitani et al. showed that 3-years recurrence-free survival rates were significantly higher for those who underwent high inguinal orchidectomy than for those who underwent tumorectomy (79.8% vs 54.1% respectively). 3-year recurrence-free survival rates for those with positive and negative margins were 54.2% and 88.6% respectively. In our case we treated our patient with high inguinal orchidectomy, and the surgical margins were tumour free.

A combination of surgery and radiotherapy has been used in positive surgical margins, lymphatic invasion, or high-grade tumours cases. The effects of adjuvant postoperative radiotherapy remain unclear and in a recent review of subgroup analysis of patients with positive surgical margins, adjuvant radiotherapy had no effect on recurrence-free survival.

Li et al. reported a 5-year survival rate of 75% and a recurrence rate of around 50–70%. Kamitani et al. showed that tumour size and histological subtype were independent risk factors for recurrence. Multi-disciplinary follow up remains of paramount importance. In our sarcoma MDT, it was agreed that the size and grade of the tumour warranted clinical monitoring with clinical examination and abdomen and pelvic CT scan follow up. Radiotherapy was reserved in case of relapse.

Conclusion

Paratesticular liposarcomas are uncommon tumours, which are often misdiagnosed preoperatively. Whilst ultrasound, CT Scan and MRI contribute to the diagnosis and differential diagnosis, the confirmative diagnosis is based on histological, immunohistochemical and molecular genetic studies. High inguinal orchidectomy with local excision should
be carried out immediately upon clinical suspicion. MDT discussions combined with regular follow ups are key to improving disease outcomes.

Informed consent

Obtained from patient.

Declaration of competing interest

None.

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