The testis and epididymis were separate from the lesion. The lump was dissected from the cord structure. The cut surface was white, solid and hard to the touch. The testis was repositioned in the scrotum and the inguinal incision was closed in layers. The patient's postoperative recovery was uneventful.

Microscopic examination revealed cells arranged in well defined fascicles with central hyperchromatic nuclei and a moderate amount of eosinophilic cytoplasm (Fig. 2). There were 4 mitoses per 10 high power fields. Small foci of necrosis were noted with the appearances of a spindle cell lesion of probable smooth muscle origin. The high cellularity, pleomorphism, necrosis and mitotic activity suggested this was a malignant tumour. The tumour was classified as a grade 2 (differentiation = 2, mitoses = 1, necrosis = 1) on the Fédération Nationale des Centres de Lutte Contre le Cancer system. These histological features were suggestive of a leiomyosarcoma. The immunohistochemistry revealed interlacing fascicles of smooth muscle (positive for desmin, smooth actin and negative for S 100) (Fig. 3A and Fig. 3B). The histology showed infiltration at edges with margins positive for tumour.

The diagnosis of leiomyosarcoma and positive margin on histology prompted us to refer the patient to the regional sarcoma clinic for further treatment. His staging CT scan of the chest, abdomen and pelvis did not reveal any local or distant tumour. Subsequently, he underwent a left radical orchidectomy with histology showing fibrosis of the spermatic cord.
cord and negative margin. Clinical and radiologic follow-up with CT of the chest, abdomen and pelvis at 12 months showed no recurrence.

Discussion

Malignant neoplasms of non-testicular origin are uncommon and are generally sarcomas. In a series of 1583 adult soft tissue sarcomas at the Memorial Sloan-Kettering Cancer Center, 43 were urological and 14 (0.8%) were paratesticular (5 rhabdomyosarcomas, 4 leiomyosarcoma, 3 liposarcomas, 1 malignant fibrous histiocytoma and 1 undifferentiated sarcoma).1

Leiomyosarcoma accounts for 5%-10% of soft tissue sarcoma.2 Though leiomyosarcoma of the spermatic cord is rare, about 110 cases have been reported in the literature.2 A review of 10 series of paratesticular sarcomas in adults showed that leiomyosarcoma is the most commonly reported histological variety, with a peak incidence in the sixth and seventh decade.2 Leiomyosarcoma originates from the spermatic cord, the scrotum or the epididymis. The most common spermatic cord type arises from undifferentiated mesenchymal cells of the cremasteric muscle and vas deferens. The less frequent epididymal form originates from the smooth muscle surrounding the basement membrane of the epididymis canal. The scrotal types are described as arising from the dartous layer. The spermatic and epididymal types drain to the retroperitoneal lymph nodes in contrast with the scrotal form that drains to the inguinal, internal and external iliac nodes.

Due to the rare occurrence of the disease, it is important to understand the modes of spread. The most common means of spread is lymphatic, then hematogenous and last, by local extension. The route of lymphatic dissemination may involve the external iliac, hypogastric, common iliac and para aortic nodes. The lung is the primary site for blood-borne metastases. Local spread to the scrotum, inguinal canal or pelvis is possible along the pathway of the vas deferens.

The natural course of leiomyosarcoma depends on site, size, grade and evidence of nodal or distant metastasis. Anatomically, leiomyosarcoma is divided into 3 subgroups: deep soft tissue, cutaneous–subcutaneous and vascular origin. According to the American Joint Committee on Cancer, staging system spermatic cord leiomyosarcoma is a deeper variety of subgroup.1

Preoperative diagnosis of spermatic cord leiomyosarcoma is difficult and usually made by histological examination. Clinically leiomyosarcoma presents as a

Fig. 1. Fig. 1A is a scrotal ultrasound showing a well-circumscribed lesion of mixed heterogeneous echogenicity on the left side. Fig. 1B is a Doppler scan demonstrating increased vascularity.
painless, firm, para-testicular intra-scrotal mass and therefore diagnostic evaluation should be similar too that of any testicular tumour. Scrotal ultrasound is a useful primary way to assess the mass and its relation to the testis. Once the diagnosis of leiomyosarcoma has been established by surgery, clinical staging is necessary. Magnetic resonance imaging can be a useful problem-solving tool and is particularly helpful in better characterizing extra testicular solid masses, as well as delineating the anatomic extent of the tumour.

Radical orchidectomy is the standard primary surgical procedure followed by adjuvant radiotherapy to reduce the local recurrence. The primary treatment with radical orchidectomy is an essential approach, but a single case report with local excision and surveillance has been reported. A reported survival rate is 50%–80% and it has been observed that simple excision is suboptimal as repeat wide excision has demonstrated microscopic residual disease in 27% of cases and therefore warrants an additional adjuvant treatment. A wide excision is almost mandatory if negative histological margins are not achieved during primary surgery. Even if extensive resection is required to clear the margins, advances in micorsurgical techniques have made it possible to reconstruct large anatomic defects in this area by local flaps. Due to the high incidence of loco-regional recurrence in the lymph nodes, 2 different treatment alternatives, prophylactic retroperitoneal lymph node dissection (RPLND) and radiotherapy, have been tried. The proponents of RPLND indicate that there is a 29% risk of metastatic potential to regional lymph nodes. A review of 101 patients by Banowsky and Schultz described 29 cases of RPLND with lymph node involvement. Of these, 17 patients had isolated lymphatic dissemination. Despite such a high incidence of lymphatic spread, no report has yet shown a significant survival benefit from the addition of RPLND to radical orchidectomy.

A study from Massachusetts involved 18 patients with spermatic cord sarcoma who were subdivided into 2 groups of 9 patients. One group had surgery and the other group had surgery plus radiation. Of the 9 patients treated with radical orchidectomy alone, 5 developed loco-regional failure, 2 of which were limited to lymph nodes. In contrast, there were no loco-regional recurrences among the 9 patients who received adjuvant radiation to the regional lymph nodes. These findings, which are consistent with those of Catton and colleagues, suggest that adjuvant radiation may effectively control loco-regional microscopic disease. In our case, histology from the orchidectomy (the specimen) showed clear margins, therefore, radiotherapy was not recommended.

In conclusion, spermatic cord leiomyosarcoma, although rare, should be one of the first differential diagnoses for a firm-to-hard lump in the cord. Apart from radical orchidectomy, there has been added benefit of adjuvant radiotherapy to prevent any loco-regional lymph node recurrence.
References


Correspondence: Mr. Chandra Shekhar Biyani, Department of Urology, Pinderfields General Hospital, Mid Yorkshire Hospitals NHS Trust, Aberford Road, Wakefield, West Yorkshire, UK WF1 4DG; shekharbiyani@hotmail.com