Sclerosing liposarcoma of the Spermatic Cord – Case Report and Literature Review

Abstract
Liposarcoma are a group of malignant tumours which are derived from mesodermal tissue. The spermatic cord is a rare site for liposarcoma; around 200 cases of liposarcoma of the spermatic cord have been reported in the English literature to date. They typically present as a non-tender, slow growing masses of variable size in the scrotum or inguinal region. They are frequently misdiagnosed as other common pathologies, such as an inguinal hernia. We present a case of liposarcoma of the spermatic cord, which histopathology confirmed to be of sclerosing subtype.

Keywords: Liposarcoma; Spermatic cord; Orchidectomy; Rare

Introduction
Liposarcoma are a group of malignant tumours which are derived from mesodermal tissue [1]. These tumours can occur anywhere in the body where fatty tissue is present [1]. Almost 70% of cases are located in the extremities and the retroperitoneum [2]. The spermatic cord is a rare site for liposarcoma; around 200 cases of liposarcoma of the spermatic cord (LSC) have been reported in the English literature to date [3,4]. LSC usually presents as a non-tender, slow growing mass of variable size in the scrotum or inguinal region [1,5]. Occasionally, their growth is rapid [2]. Few cases present with pain [4]. The mean age of presentation is 61 [3]. However, cases in ages ranging from 16 to 88 have been noted [5]. Though cases have been reported worldwide, almost a quarter have been described in Japanese men [3]. Clinically, LSC is often mistaken for more common pathologies, such as an inguinal hernia, hydrocele, spermatocele, or tumours of the testes and epididymis [2]. Below, we present a textbook case of liposarcoma of the spermatic cord.

Case Presentation
A 77 years old gentleman presented to his GP with a non-tender long-standing scrotal swelling for the last 10 years. He had a previous anterior resection for bowel cancer. On examination, in the outpatient’s clinic, the right hemiscrotum contained a small hydrocele, but also a firm mass which prompted urgent ultrasound scan. The scan showed a soft tissue mass in close relationship to but sparing the testicle, measuring 8x4 cm (Figure 1). Given the patient’s age and the nature of the mass suggested by imaging, he was advised on surgical exploration and consented for possible orchidectomy, pending on intraoperative findings, and direct inspection of the mass. Surgical exploration of the scrotum was performed. The testicle and epididymis looked normal. A lump of fatty tissue mimicking an inguinal hernia was noted protruding down from the inguinal canal. After careful dissection, a hard mass was found adjacent to and engulfing the spermatic cord. Orchidectomy was performed, ensuring wide excision of the mass and its surroundings.

Post excisional inspection confirmed a hard yellowish-white mass, mixed with fatty tissue, enclosed in a pseudo-capsule. This was arising from the spermatic cord. The testicle looked entirely normal, but was impossible to be spared due to oncological principles (Figures 2-4). The pathology report confirmed a 90x60x53 mm sclerosing liposarcoma of the spermatic cord pT2bNxMx, grade 1, with clear surgical margins. The sections show a relatively well circumscribed tumour composed predominantly of numerous various sized adipocytes with occasional lipoblasts and scattered large atypical cells. There is also a prominent hyalinised and sclerotic fibrous tissue component which also contains more spindled and atypical nuclei. No necrosis or dedifferentiated areas were seen. The patient was followed up by the sarcoma specialist team, and had recovered very well after surgery.

Discussion
LSC can be classified histologically into five subtypes. From highest to lowest incidence, these are: well-differentiated, dedifferentiated, myxoid, round cell, and pleomorphic [6]. The well-differentiated group can be further divided into adipocytic (lipoma-like) and sclerosing liposarcoma, as well as two rare types; inflammatory and spindle cell liposarcoma [7]. The majority of LSC are locally aggressive, and tend to spread primarily by local extension [7-9]. Well-differentiated and myxoid liposarcoma are considered to be low grade, and have a low potential for metastases, and favourable prognosis [1,10]. Round cell, pleomorphic, and de-differentiated liposarcoma are considered to be high grade, and have been known to metastasise via the blood and lymphatics [1,7,9,11].
Generally, the scrotum and its contents can be reliably imaged using ultrasonography, contrast enhanced CT and MRI [3]. In this case, ultrasound scan reliably distinguished the suspect mass as being separate from the testicle (Figure 1). However, diagnosing LSC via imaging studies is difficult. For example, ultrasound cannot reliably distinguish between well differentiated liposarcoma and benign lipoma [11]. Further, CT and MRI imaging commonly misdiagnose LSC as a hernia, due to its fatty composition [6]. Hence, LSC is usually diagnosed intra- or post-operatively [1].
The treatment of choice for LSC is radical inguinal orchidectomy, with wide local excision, and high ligation of the spermatic cord [4,9]. To prevent local recurrence, scrotectomy may be considered in patients with high grade tumours [4]. Achieving disease-free margins is important; margins of 10mm or more are associated with a risk of recurrence below 10% [9]. Sometimes a second resection is advised if margins are positive [10]. Lymph node dissections are not routinely performed, as lymph nodes are rarely involved [3,12]. The risk of local recurrence in LSC is well documented; 55 to 70% of cases have recurrence of disease [4,9]. Cases of recurrence have been reported even after 20 years from diagnosis [12]. Hence, long term follow up of these patients is vital [12].

Risk factors for local recurrence include the presence of a high grade tumour, tumour size of greater than 5cm, and positive margins [9]. Recurrences are best treated with repeat wide local excision of the lesion [9]. Radiotherapy reduces the local recurrence rate, though it does not affect overall survival [6]. It is recommended in addition to surgery for high grade tumours, lymphatic invasion, inadequate margins or relapses [2]. Chemotherapy is not recommended as liposarcoma tend to have relative resistant toward chemotherapeutic agents [9,10].

References