Inguino-scrotal sarcomas

BY PAUL CLEAVELAND AND VIJAY SANGAR

A sarcoma is a malignant tumour that originates from mesenchymal cells such as adipose tissue, bone, cartilage and smooth muscle. Although these tumours histologically do not originate from the urogenital tract, urologists are often involved in their diagnosis and management when they arise in the genitals or in the retroperitoneum. They most commonly arise from the spermatic cord, epididymis, testis (paratesticular structures) and can extend into the inguinal canal or retroperitoneum. Inguino-scrotal sarcomas are very rare despite the fact that they comprise the most common urological soft tissue malignancy in adults [1].

Incidence

Soft tissue sarcomas constitute 1% of all adult neoplasms. Genitourinary sarcomas make up about 2% of soft tissue sarcomas [2]. Sarcomas of the spermatic cord, epididymis and testis are the most common genitourinary sarcomas in adults. Approximately 3% of malignant lesions of the spermatic cord are liposacromas [3]. Most patients present in their 50s to 60s with a slow growing mass.

Pathology

Mesenchymal tumours can be classified according to the tissue of origin. The most common subtype in adults is liposarcoma followed by leiomyosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma and fibrosarcoma. Embryonal rhabdomyosarcoma is the most common subtype in men under the age of 30.

Inguino-scrotal sarcomas can present as paratesticular, spermatic cord tumours or retroperitoneal tumours extending into the inguinal canal and scrotum. These tumours can be either benign or malignant.

Benign tumours

Benign lipomas are the most common paratesticular or spermatic cord tumour. They almost never transform into malignant liposarcoma which is the case for other benign mesenchymal tumours. Therefore, it is more likely that malignant tumours arise de novo as opposed to undergoing malignant transformation from their benign counterparts. Benign paratesicular tumours include lipoma, leiomyoma, adenomatoid tumours, fibroma or haemangioma.

Malignant tumours

Malignant paratesticular tumours include leiomyosarcoma (32%), rhabdomyosarcoma (24%), liposarcoma (20%) and malignant fibrous histiocytoma (13%) [4].

Liposarcomas can be categorised into five categories based on the World Health Organization:
- Well differentiated
- Myxoid
- Round cell
- Dedifferentiated
- Pleomorphic

The first two are considered low grade and the last three high grade.

Myxoid liposarcomas are composed of primitive lipoblasts without the typical fat rich cytoplasm. They resemble primitive mesenchymal cells. Well-differentiated liposarcoma closely resembles a lipoma making their distinction challenging. Round cell liposarcoma is composed of small round cells equal in size and packed closely together. De-differentiated liposarcoma has areas of well and poorly differentiated tumour. At the time of recurrence other phenotypes may be present that were not present in the initial tumour. Pleomorphic liposarcoma has a disorganised growth pattern with cellular pleomorphism, giant cells and anaplastic pyknotic nuclei.

Sarcomas have a low tendency to metastasise however, they are locally aggressive and invasive. High grade round cell and pleomorphic subtypes have a more frequent tendency to metastasise to the regional lymph nodes.

Diagnosis / investigations

Inguino-scrotal sarcomas will manifest as painless palpable lumps which tend to be large in size once noticeable. They are often mistaken for inguinal hernias, lipoma of the cord or testicular tumours. They are often asymptomatic apart from the presence of a mass. The diagnosis is usually made perioperatively or histologically after they have been locally excised [5].

Ultrasonography will demonstrate a solid mass but it may not distinguish between benign and malignant pathology. It may also be difficult to differentiate between a sarcoma and germ cell tumour. Any solid mass in the scrotum external to the tunica albuginea should be suspected and explored through the inguinal canal. Ultrasound has been found to have a sensitivity of 95-100% in distinguishing between extratesticular and intratesticular tumours. Paratesticular sarcomas are seen as heterogenous hypervascular tumours on ultrasound [6]. Liposarcomas are heterogenous with varying amounts of fat and hyperechoeic areas [7].

CT may help distinguish hernias or lipomas in the inguinal region from liposarcomas when there is doubt. They will also help assess the extent of local spread from the tumour when the inguinal canal is involved. Imaging can also establish whether the tumour is arising from the retroperitoneum and invading into the inguino-scrotal area or if it is a primary spermatic cord tumour. This can help plan the extent of the resection and whether any other involved organs may need to be removed [6].

MRI scan can give better soft tissue differentiation and determine whether the

Table 1: TNM staging of paratesticular sarcoma.	
T stage	
T1a	<5cm superficial
T1b	<5cm deep
T2a	>5cm superficial
T2b	>5cm deep
N1	Regional lymph nodes
M1	Distant metastasis

Table 2: The TNM and American Joint Committee on Cancer sarcoma staging system.				
Stage	**Grade**	**Tumour**	**Lymph Nodes**	**Metastases**
IA	G1-2	T1a-b	N0	M0
IB	G1-2	T2a	N0	M0
IIA	G1-2	T2b	N0	M0
IIB	G3-4	T1a-b	N0	M0
IIC	G3-4	T2a	N0	M0
III	G3-4	T2b	N0	M0
IV	Any G	T1a-2b	N0-1	M0-1

mass is solid or cystic [6].

Staging of paratesticular tumours is based on histological grading and the presence of metastasis. Tumours are staged with CT chest, abdomen, pelvis and MRI. The staging classification is based on TNM and the American Joint Committee on Cancer [8,9] (Table 1 and 2). Grading is based on the TNM system (G1 – well differentiated, G2 – moderately differentiated, G3 – poorly differentiated or G4 – undifferentiated). Another system used is the French Federation of Cancer Centres Sarcoma Group where tumour differentiation, mitotic activity and the amount of tumour necrosis are scored individually [10].

Management

The rarity of inguino-scrotal sarcomas means that the optimal treatment is based on previous experience from various groups. Most patients will have localised disease when they present and they have a tendency for local recurrence if inadequately excised.

Surgery

The mainstay of treatment is surgery. This should involve a high inguinal approach with wide excision of the spermatic cord and testis. This should be done with high ligation as close to the deep ring as possible and a wide margin in order to avoid local recurrence. A wide margin is achieved by performing a hemi-scrotectomy. The outcome is fairly good when there are clear margins indicating complete excision [11-13]. The main risk factors for local recurrence include large tumour size, inguinal tumour location, positive margins and previous intra-lesional surgery [12,14].

When there is an incomplete resection as is often the case (i.e. the patient has had a local excision through the scrotum or part of the cord has been left behind) there is an increased risk of local recurrence and this constitutes an incomplete resection. The local recurrence rates after orchidectomy have been reported between 25-37% [14]. This can be avoided by having a high level of suspicion prior to treatment in order to ensure a wide margin. An incomplete resection should be managed with a repeat wide excision which would incorporate en bloc excision of the ipsilateral scrotum (hemi-scrotectomy), groin wound, adipose tissue and inguinal canal if necessary.

Adjuvant and systemic treatment

Adjuvant radiotherapy can be used postoperatively when there is doubt about the adequacy of the resection. This is especially the case when the inguinal canal is involved meaning that complete excision can be difficult to achieve. The efficacy of radiotherapy however is debated. One study found no local recurrence in patients treated with adjuvant radiotherapy compared to 37% in those treated with orchidectomy alone [15]. Another study reported 14% local failure rates in patients who had adjuvant treatment with either wide local excision or radiotherapy [16]. Furthermore, microscopic disease has been found in 27% of patients undergoing wide local excision after apparent complete tumour excision with orchidectomy [12-15].

Local and regional lymph nodes are rarely involved therefore, prophylactic lymph node dissection is generally not indicated unless there is evidence of tumour involvement. Retroperitoneal lymph node dissection has been suggested in high risk pathology (rhabdomyosarcoma, malignant fibrous histiocytoma) or in radiologically evident disease [17-18].

The role of adjuvant chemotherapy in adults is controversial and not established. Chemotherapy is reserved for those patients with evidence of retroperitoneal lymph node involvement or distant metastasis.

Follow-up

Late recurrences have been reported and so follow-up should be done for at least 10 years [11]. This involves clinical examination and CT imaging at intervals [19].

Prognosis

The long-term survival for paratesticular sarcoma is approximately 50%. Liposarcoma has a better prognosis while malignant fibrous histiocytoma and leiomyosarcoma have the worst prognosis. Ten and 15-year outcome data have been quoted. Actuarial local control rates have been quoted as 72% and 61%, respectively. The distant metastasis-free rates were 85% and 85%, respectively. The overall survival rates were 63% and 52%, respectively [20].

Inguino-scrotal sarcomas are rare and tend to be diagnosed histologically following local excision. Their aggressive nature and high risk of local recurrence should be recognised so that they can be managed in specialist tertiary centres where wide local excision can be carried out.

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AUTHORS

Paul Cleaveland,
ST7 Urology, Christie NHS Foundation Trust, Manchester.

Professor Vijay Sangar,
Consultant Urological Surgeon and Director of Surgery, Christie NHS Foundation Trust, Manchester.

Declaration of competing interests: None declared.

SECTION EDITOR

Arie Parnham MBChB, FRCS(Urol)
Consultant Urologist, The Christie NHS Foundation Trust, Manchester, UK.