Inflammatory myofibroblastic sarcoma of the spermatic cord

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Abstract Inflammatory myofibroblastic tumours are rare clinical presentations and are often mistaken for a malignant neoplasm. They can arise from various organs, mainly in children and young adults and most commonly from the urinary bladder. Paratesticular tissues including spermatic cord are uncommon sites for these tumours. In those arising from the paratestis, wide excision usually including an orchidectomy is the treatment of choice. A rare such tumour in a 64-year-old patient is reported here.

Keywords Inflammatory myofibroblastic tumour · Paratestis · Radical excision

Introduction

While testicular tumours are well known, paratesticular neoplasms are uncommon tumours occurring in the paratesticular tissues such as the epididymis and vas deferens. They are slow growing and usually occur in the younger age group. Sometimes confused with malignancy because of being hard and their rapid growth, they are benign on histology with immunohistochemistry being required for exact diagnosis. Complete local excision is the treatment of choice and recurrences are rare.

Case report

A 64-year-old male patient presented to the surgical OPD with a slowly growing painless swelling in the upper part of the right side of the scrotum of 6 months duration. However, over the last 1 month it had rapidly increased in size. The swelling was not reducible, non-inflammatory and was not associated with weight loss or anorexia.

Examination revealed a discrete, non-tender swelling of approximately 8 × 5 cm in the upper part of the right side of the scrotum, separate from the testis and stony hard in consistency. There was no inguinal or cervical lymphadenopathy and abdominal examination was normal (Figs. 1, 2).

Haematological investigations including α -fetoprotein and β -hCG levels were normal. Ultrasonography with colour Doppler studies showed a heterogenous texture and moderate vascularity with a clear demarcation from the testis. Abdominal sonography was normal. Fine-needle aspiration cytology revealed spindle cells with buckled nuclei suggesting a mesenchymal neoplasm with the possibility of a nerve sheath tumour, though a sarcoma could not be ruled out conclusively.

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Fig. 1 The swelling seen in the right side of the scrotum superior to the testis

Fig. 2 The swelling is completely separate from the testis

An inguinal exploration was undertaken with consent taken for a right orchidectomy. It revealed a lobulated tumour with a thin capsule arising from the spermatic cord, adjacent to, but separate from the testis. A high ligation of the cord was performed and the swelling including the entire spermatic cord and right testis was excised, in effect a right radical orchidectomy.

Histopathology showed an intermediate grade spindle cell sarcoma (Fig. 3) with penetration through the pseudocapsule while immunohistochemistry confirmed an inflammatory myofibroblastic tumour (IMT) of the spermatic cord. Postoperative recovery was uneventful except for a small organised haematoma in the right scrotum 3 weeks following surgery which needed evacuation through a small scrotal incision.

Fig. 3 Histopathology showing intermediate grade spindle cell sarcoma, (confirmed on immunohistochemistry as inflammatory myofibroblastic sarcoma of the spermatic cord)

Discussion

Mesenchymal tumours of the spermatic cord are rare and include liposarcoma, leiomyosarcoma and rhabdomyosarcoma of which IMT is the rarest. It has been described by a variety of names such as inflammatory pseudotumour, pseudosarcoma or even as proliferative funiculitis [1]. It mainly occurs in children and young adults, the commonest site being the urinary bladder [1]. Very few cases have been reported in the paratestis, a term comprising the spermatic cord, epididymis and coverings of the testis [2]. Our patient was 64-year-old and with the lesion in the spermatic cord was therefore a rare presentation.

Exact aetiology of these uncommon tumours is unknown and ischaemia, infection or chronic irritation have all been postulated as possible incriminating factors [3]. Association with a hydrocele or haematocele is not unknown. They are mostly ill defined, firm grey or white tumours, generally <3 cm in diameter though in our patient it was larger, 8 × 5 cm in size [2]. It was stony hard prompting us to strongly consider a malignant neoplasm, specially in view of its reported rapid increase in size over the last 1 month. In fact Yamashina M, et al. [4] had reported a series of three cases of IMT mimicking an epididymal sarcoma. The tumours are considered locally invasive with penetration through the pseudocapsule or into surrounding fat. Though considered benign, lesions occurring in the retroperitoneum, mesentery or mediastinum are considered potentially malignant and could recur after treatment [5].

There are three microscopic variants of these paratesticular neoplasms which could either be of the nodular fasciitis, fibrous histiocytoma or the scar-like tissue type. Basically the tumour consists of myofibroblasts with spindle cell proliferation of variable cellularity against a background of mixed inflammatory infiltrate and loose collagenous

stroma. However cellular atypia and mitotic activity is not seen. Immunohistochemistry is of vital importance in the diagnosis of these tumours. Immunoreactivity is seen for vimentin, muscle specific actin and desmin but not for myoglobin and S100 protein [6].

Differential diagnosis includes benign conditions such as varicocele, spermatocele or tuberculosis and malignancies such as rhabdomyosarcoma, inflammatory fibrosarcoma or liposarcoma. Extensive immunohistochemical analysis using a large number of immunostains is required for this purpose. This was true for our patient too where histopathology had shown an intermediate grade spindle cell sarcoma and immunohistochemistry was more specific in classifying it as an inflammatory myofibroblastic sarcoma of the spermatic cord.

The treatment of choice for paratesticular IMT is wide surgical excision. In our patient this was achieved by excising the tumour along with an orchidectomy and high ligation of the cord at the internal ring. Recurrences are uncommon in the encapsulated lesions and are a possibility if the excision has been incomplete. Other treatment options include radio-therapy and chemotherapy with methotrexate or steroids but these are generally used for patients with recurrence [5]. Overall prognosis after complete surgical excision is excellent. Our patient is symptom free for close to 3 years now.

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