Role of magnetic resonance imaging in the early diagnosis of paratesticular rhabdomyosarcoma

T Shah, O Abu-Sanad, H Marsh

Medway Maritime Hospital, Gillingham, UK

ABSTRACT

INTRODUCTION Paratesticular lesions are common, and one subgroup is paratesticular rhabdomyosarcoma. The latter is a relatively uncommon (but aggressive) tumour that affects children and adolescents predominantly. Ultrasound is the first-line investigation, but can be inconclusive. Magnetic resonance imaging (MRI) can provide useful information, but its role in the diagnosis of rhabdomyosarcoma is not clear.

CASE HISTORY We report a 17-year-old male who presented with a one-month history of a rapidly enlarging, non-tender, lump in the right testicle. Urgent ultrasound of the scrotum revealed a heterogeneous paratesticular mass that was hypervascular and showed calcification in the right inguinal area. MRI of the pelvis showed a solid, enhancing lesion of dimension located superior to the upper pole of the right testes and a slightly heterogeneous T2 signal, but was homogenous post-contrast. The patient underwent right radical orchidectomy, and histology results were assessed. He received chemotherapy and is being followed up.

CONCLUSIONS Improvements in imaging in addition to early surgical intervention and chemotherapy treatment are crucial to improve survival chances against rhabdomyosarcoma. Ultrasound findings for benign diseases may mimic those seen in rhabdomyosarcoma. In such cases of diagnostic uncertainty, our surgical team suggest MRI to reduce the risk of a delayed diagnosis and time to treatment.

KEYWORDS

Paratesticular lesions – Rhabdomyosarcoma – Magnetic resonance imaging – Ultrasound – Chemotherapy – Histology

Accepted 25 October 2015; published online XXX

CORRESPONDENCE TO

Talib Shah, E: talibshah@doctors.org.uk

Paratesticular lesions are common, and are usually benign, cystic or inflammatory. Primary tumours of paratesticular tissue are rare, but a prevalence of 3–16% has been reported for patients referred for ultrasound of the scrotum.¹

One subgroup is paratesticular rhabdomyosarcoma, which is a relatively uncommon but aggressive tumour that affects children and adolescents predominantly. Ultrasound is the first-line investigation, but can be inconclusive. In these cases, magnetic resonance imaging (MRI) can provide useful information, but its role in the diagnosis of rhabdomyosarcoma is not clear.

Case History

We report a 17-year-old male who presented with a one-month history of a rapidly enlarging, non-tender, lump in the right testicle. There were neither important risk factors such as an undescended testis or family history nor an important history of medical/surgical treatment. A thorough clinical examination revealed a firm, non-tender, mobile lump with a lobular surface adjacent to the right testis.

Urgent ultrasound of the scrotum revealed a heterogeneous paratesticular mass that was hypervascular and showed calcification in the right inguinal area. MRI of the pelvis showed a solid, enhancing lesion of dimension 37 × 30 × 28mm located superior to the upper pole of the right testes. It demonstrated a slightly heterogeneous T2 signal (Fig 1), but was homogenous post-contrast (Fig 2). Testes and inguinal lymph nodes appeared unremarkable. There was no appreciable abnormality within the thorax, abdomen or pelvis on computed tomography (CT). Blood testing showed normal levels of lactate dehydrogenase, alpha-feto-protein, and β-human chorionic gonadotropin.

The patient underwent an uneventful right radical orchidectomy. This case was discussed in a multidisciplinary team meeting in the context of histology results.

Macroscopic and Microscopic Findings

Macroscopic examination revealed a firm, tan-coloured nodule of dimension 35 × 30 × 30mm. A cut section showed a uniformly grey mass (Fig 3).

Figure 1 Magnetic resonance imaging of the testes shows an enhanced lesion of dimension $37 \times 30 \times 28$ mm located superior to the upper pole of the right testis (arrow) and demonstrates a slightly heterogeneous T2 signal

Figure 2 Magnetic resonance imaging of the testes after contrast administration reveals a homogenous lesion superior to the upper pole of the right testis (arrow)

Figure 3 Cut section of a firm, tan-coloured nodule of dimension $35 \times 30 \times 30$ mm reveals a uniformly grey mass

Figure 4 Histological staining of a nodule (haematoxylin and eosin; $\times 200$ magnification) shows some cells with more eosinophilic cytoplasm, thereby suggesting rhabdomyoblastic differentiation

Histology revealed varying myxoid and cellular zones composed of primitive ovoid and spindle-shaped cells that occasionally contained eosinophilic cytoplasm and immature cartilage (Fig 4). Immunohistochemistry showed the tumour to be strongly positive for desmin and vimentin.

The testis and epididymis were unremarkable with regard to histology. The resection margin appeared to be clear of tumour cells. Based on these findings, a diagnosis of embryonal rhabdomyosarcoma was confirmed.

Our patient received chemotherapy in a tertiary cancer centre according to the protocol for non-metastatic rhabdomyosarcoma set by the European Paediatric Soft Tissue Sarcoma Study Group. This protocol comprised four cycles of vincristine, actinomycin B and isophosphamide followed by five cycles of vincristine and actinomycin B at 3-week intervals. His follow-up will comprise ultrasound of the abdomen,

chest radiography, and clinic appointments at 3-month intervals for the first 2 years, then at six-month intervals until 5 years after surgery.

Discussion

Paratesticular rhabdomyosarcoma is a rare tumour comprising only 7% of all rhabdomyosarcoma cases. It arises from the mesenchymal tissue of the epididymis, spermatic cord and testes.² There is a bimodal distribution with peak incidences at 5 years and 16 years, and the median age at the diagnosis is 7 years.¹ Rhabdomyosarcoma is regarded to be a highly aggressive and recurring tumour, with ~26–71% of cases presenting with metastases to regional lymph nodes (usually to para-aortic and iliac nodes). Haematogenous spread to the lungs, liver and cortical bone can also occur. Typical presentation is a short history of a painless, unilateral lump in the scrotum.^{1,2}

Ultrasound is the first-line investigation for scrotal masses due to its high accuracy, low cost and wide availability. Ultrasound commonly shows a large, ill-defined, heterogeneous vascular mass. However, these findings can be inconclusive due to the similar appearance of other scrotal masses (eg fibromas, leiomyomas, adenomatoid tumours).^{1,5} In this instance, MRI can be useful because it provides better characterisation of tissue, and can be used to differentiate between benign and metastatic lesions with high accuracy. Such characteristics have led to the suggestion that MRI findings may be closely correlated with the histological type of testicular tumours. If such a correlation exists, diagnostic uncertainty can be reduced and, in cases of benign disease, surgical intervention may be avoided, resulting in improved management of disease and reduced healthcare costs.⁵ In addition, MRI allows for precise localisation of tumours and assessment of involvement of surrounding structures. Given the aggressive nature of rhabdomyosarcoma, CT of the chest, abdomen and pelvis is essential for investigation of lymph-node involvement and disease staging.

There are three main histological subtypes of rhabdomyosarcoma: embryonal, alveolar and pleomorphic. Embryonal RMS represents 90% of all cases of paratesticular rhabdomyosarcoma.² Embryonal rhabdomyosarcoma usually appears as an encapsulated grey-white mass with haemorrhagic and cystic areas, and microscopic examination reveals small, round cells with myoblastic differentiation.¹

Multimodal management of paratesticular rhabdomyosarcoma by resection, chemotherapy and radiotherapy has

become standard practice worldwide. Orchiectomy with high resection of the spermatic cord through an inguinal approach is favoured over a transscrotal approach (which can result in contamination and an increased risk of recurrence due to microscopic residual disease in the tumour bed).⁴ Previously, chemotherapy using vincristine, actinomycin D and cyclophosphamide has been employed, but more recently cyclophosphamide has been replaced by ifosfamide.⁴ Radiotherapy is recommended alongside chemotherapy for patients with nodal or microscopic residual disease, but patients should be selected carefully due to an increased risk of complications if these treatments are combined.⁵ The prognosis of paratesticular rhabdomyosarcoma is considered to be poor but, with changes in the management approach, overall survival can be ≤94%.⁴

Conclusions

Rhabdomyosarcoma is a rare condition with considerable morbidity for children and adolescents, particularly in advanced cases, due to a late diagnosis. Improvements in imaging in addition to early surgical intervention and chemotherapy treatment are crucial to improve survival chances. Ultrasound findings for benign diseases such as adenomatoid tumours, leiomyomas and fibromas may mimic those seen in rhabdomyosarcoma. In such cases of diagnostic uncertainty, our surgical team suggest MRI to reduce the risk of a delayed diagnosis and time to treatment. This report is a reminder of the importance of initial clinical evaluation and rapid imaging for management of this unusual tumour in a urology unit in a general district hospital.

References

1. Akbar SA, Sayyed TA, Jafri SZH *et al.* Multimodality imaging of paratesticular neoplasms and their rare mimics. *Radiographics* 2003; **23**: 1,461–1,476.
2. Kumar R, Kapoor R, Khosla D *et al.* Paratesticular rhabdomyosarcoma in young adults: a tertiary care institute experience. *Indian J Urol* 2013; **29**: 110–113.
3. Cassidy FH, Ishioka KM, McMahon CJ *et al.* MR imaging of scrotal tumours and pseudotumours. *Radiographics* 2010; **30**: 665–683.
4. Dall'Igna P, Bisogno G, Ferrari A *et al.* Primary transscrotal excision of paratesticular rhabdomyosarcoma. Is hemisectomy really mandatory? *Cancer* 2003; **97**: 1,981–1,984.
5. Hughes LL, Baruzzi MJ, Ribeiro RC *et al.* Paratesticular rhabdomyosarcoma: delayed effects of multimodal therapy and implications for current management. *Cancer* 1994; **73**: 476–482.