Case report

Ultrasound diagnosis of paratesticular rhabdomyosarcoma

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Abstract. Rhabdomyosarcoma is the most common tumour of the lower genitourinary tract in children in the first two decades. Paratesticular rhabdomyosarcoma is associated with a significantly better outcome than lesions elsewhere in the genitourinary tract. Although ultrasound is considered the imaging modality of choice for evaluating intrascrotal pathology, the ultrasound appearance of paratesticular rhabdomyosarcoma has rarely been reported and may be confused with other disease entities such as epididymitis, adenomatoid tumour and leiomyoma. We present the ultrasound features of a paratesticular rhabdomyosarcoma, discussing the clinical features and differential diagnosis.

Although rhabdomyosarcoma has been recognised as one of the most common forms of childhood solid tumour, occurring with a frequency similar to that of Wilms' tumour and neuroblastoma [1], paratesticular rhabdomyosarcomas are extremely rare. The majority of these tumours occur in the first two decades of life and belong to the embryonal histopathologic subtype [2].

Although ultrasound has been considered the image modality of choice in differentiating intratesticular from extratesticular pathology, reports on the ultrasound features of paratesticular rhabdomyosarcoma is limited and preoperative diagnosis may be difficult. Because of the highly aggressive nature of paratesticular rhabdomyosarcomas, early diagnosis is important, as patients with disease localized to scrotum have a good prognosis [3].

Case report

A 14-year-old boy visited the outpatient department with a 1 month history of left scrotal swelling. Initially, the scrotal swelling had been ignored but as the scrotum progressively enlarged, medical help was sought. There was no relevant past medical history, and the patient denied a history of scrotal trauma or infection.

Physical examination revealed a tender mass occupying the whole of the left hemiscrotum. Transillumination test was positive. The right testis was normal. There were no palpable inguinal lymph nodes.

A clinical diagnosis of hydrocele with epididymitis was made and an ultrasound examination was arranged. High resolution ultrasound using a 10 MHz linear transducer demonstrated a 6 cm × 5 cm extratesticular hypoechoic mass in the left scrotum (Figure 1), the mass was found to be hypervascular when colour Doppler was applied (Figure 2), no central hypoechoic lesion indicative of tumour necrosis was seen within the mass. The left testis was intact and could be well delineated from the extratesticular mass. A non-septated hydrocele was found in the left scrotum (Figure 1). The left epididymal tail was

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Figure 1. Longitudinal section (composite image) of high resolution ultrasound shows a well defined hypoechoic extratesticular mass (arrows) was found in the left scrotum, hydrocele was also present.

Figure 2. Colour Doppler ultrasound shows that the mass was hypervascular (arrow).
not seen. The right testis and epididymis were unremarkable. An abdominal CT examination showed no evidence of retroperitoneal lymphadenopathy. Laboratory test results, including a chemistry panel of serum alpha feto protein, and beta-human chorionic gonadotropin were normal. Orchidectomy followed by adjuvant chemotherapy were suggested but rejected by the patient’s parent, hence epididymectomy and orchipexy were performed. On operation a 10 cm x 5 cm x 5 cm brownish colour firm mass arising from the left spermatic cord was found, en-bloc excision was done with preservation of the left testis. Neither haemorrhage nor necrosis were identified within the mass. Pathology showed that this mass was a myxoid tumour composed of pleomorphic cells (Figure 3a). Immunohistochemically, these cells were positive for desmin, but negative for smooth muscle actin, and showed cytoplasmic striation (Figure 3b) suggestive of rhabdomyosarcoma. After the operation, chemotherapy was recommended but was again rejected. Tumour recurrence was seen in the left scrotum 2 months after the operation, consisting of a hypoechoic vascular mass located cephalad to the left testicle, calcifications were seen within the mass (Figure 4). Finally, the patient’s parent agreed to accept radical orchectomy. A 12 cm x 6 cm x 5 cm well demarcated tumour was found attached to the spermatic cord during the operation. Pathology confirmed that this was recurrent rhabdomyosarcoma. The patient was subsequently treated with chemotherapy using endoxan and phamarubicin.

**Discussion**

Ultrasound is the imaging modality of choice for patients with suspected scrotal abnormalities, and is accurate in distinguishing intratesticular from extratesticular lesions. Although most of the extratesticular lesions are benign, malignancy does occur; the most common malignant tumours in infants and children are rhabdomyosarcomas [4]. Rhabdomyosarcomas may develop anywhere in the body, and 4% occur in the paratesticular region [3].

Paratesticular rhabdomyosarcoma is one of the most common non-germinal neoplasms affecting the scrotal contents in children and adolescents. Clinically, the patient usually presents with non-specific complaints of a unilateral, painless intrascrotal swelling not associated with fever. Transillumination test is positive when a hydrocele is present, often resulting in a misdiagnosis of epididymitis, which is more commonly associated with hydrocele.

Paratesticular rhabdomyosarcomas usually spread to lymph nodes such as the paraaortic, para caval nodal region but rarely to the inguinal lymph nodes, unless tumour invasion of the scrotal skin has occurred [1]. The most common distant sites of metastasis are cortical bone, bone marrow and lungs through haematogenous spread [5].

Descriptions of the ultrasound findings of paratesticular rhabdomyosarcoma are limited and variable. In 1989, Solvietti and Tannous [6, 7] described this tumour as an echo-poor mass with or without hydrocele. However, other reports [8, 9] suggested high reflectivity in the epididymis in association with reduced reflectivity of the testicle when the tumour has invaded into the testis.

In 1995, Wood and Dewbury [9] first described the Doppler ultrasound features in a 17-year-old boy as a highly reflective soft tissue mass located in the epididymis, with increased vascularity on colour Doppler, which they initially considered to be epididymitis. Progressive swelling of the scrotum occurred and a scrotal malignancy was suspected 6 months later, when ultrasound disclosed a swollen testis with decreased echogenicity and no discernable plane detected between the testis and the epididymal mass, illustrating that differentiating rhabdomyosarcoma from epididymitis may be difficult.

In the present case, epididymitis was suspected initially as the patient presented with tender mass in the scrotum and transillumination test was positive. At ultrasound, the region of the epididymis was found to be occupied by a hypervascular mass with mixed echogenicity, indistinguishable from epididymitis. However, correlation of the ultrasound features with the clinical findings of a growing scrotal mass in a young children led us to suspect preoperatively the diagnosis of a paratesticular rhabdomyosarcoma. In our opinion when encountering an enlarged intrascrotal extratesticular mass with hypervascularity, the following points may be helpful in differentiating a rhabdomyosarcoma from epididymitis:

![Figure 3.](image1)

**Figure 3.** (a) Pathologically, this tumour was a myxoid tumour, and composed of pleomorphic cells (haematoxylin & eosin stain x 400). (b) Immunohistochemical stain using desmin (x 400) showed that these cells were positive for desmin, and showed cytoplasmic striation (arrowhead).

![Figure 4.](image2)

**Figure 4.** Ultrasound (transverse section) shows recurrent hypoechoic tumour located cephalad to the left testicle, calcifications are seen within the recurrent tumour (arrows).
(1) young age
(2) gradual enlargement of testis, not acute onset of symptoms

Other solid extratesticular masses which should be considered in the differential diagnosis include adenomatoid tumours and leiomyomas. The ultrasound features of adenomatoid tumour are of a well defined round shaped mass having variable echogenicity [lo], usually occurring in the third and fourth decades. Ultrasound reports of scrotal leiomyoma have described this tumour as a solid hypoechoic or heterogeneous mass [11], which may or may not contain calcification, sharp shadows not induced by calcification corresponding to transition zones between the various tissue components of the mass have also reported as a specific feature of leiomyoma [12].

Our case also illustrates the aggressive nature of paratesticular rhabdomyosarcoma. Previous reports suggest that this tumour is associated with a good prognosis when the tumour is localized within the scrotum, and orchiectomy should be followed by chemotherapy [3]. However, in our case this standard treatment protocol was initially rejected with consequent tumour recurrence in a very short term, just 2 months after epididymectomy. Again this emphasises the importance of early diagnosis and proper treatment before metastasis to the other organs occurred.

In conclusion, when encountering a solid extratesticular mass in a young patient, rhabdomyosarcoma should be suspected. If the clinical onset is acute and associated with reactive hydrocele, epididymitis should be considered, if not rhabdomyosarcoma is more likely.

References