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Rhabdoid tumour of the kidney: imaging findings

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Abstract *Background.* Rhabdoid tumour of the kidney (RTK) is a rare tumour, but it is the most aggressive malignant neoplasm of the kidney in children.
Objective. To analyse the radiological findings of RTK in children.
Materials and methods. The clinical and radiological findings in seven children (age range 6 months to 4.7 years; median 18 months) with pathologically proven RTK were retrospectively reviewed. We analysed tumour size, tumour location, tumour margin, subcapsular haematoma, tumour necrosis, haemorrhage, calcification and lymphadenopathy.
Results. Tumour size varied from 5 to 12 cm. Four tumours were located mainly in the central portion of the kidney, while three tumours were mainly sited peripherally. The margins of the tumour were ill-defined in four (57 %) of seven cases, a lobulated tumour surface was depicted in all seven (100 %), subcapsular haematoma was present in four (57 %), tumour necrosis or haemorrhage in seven (100 %), calcifications in three (43 %) and retroperitoneal lymphadenopathy in four (57 %).
Conclusions. Imaging findings of RTK are subcapsular haematoma, a lobulated surface of the tumour, calcification and tumour necrosis or haemorrhage.

Introduction

Rhabdoid tumour of the kidney (RTK) is a highly malignant tumour accounting for 2 % of malignant renal neoplasms [1, 2]. RTK is unique among childhood renal neoplasms in its frequent association with primary or metastatic CNS lesions [3, 4]. It had been regarded as a rhabdomyosarcomatoid variant of Wilms' tumour [5], but it is now thought to be a separate entity [6–8]. We retrospectively analysed the radiological findings of RTK in children.

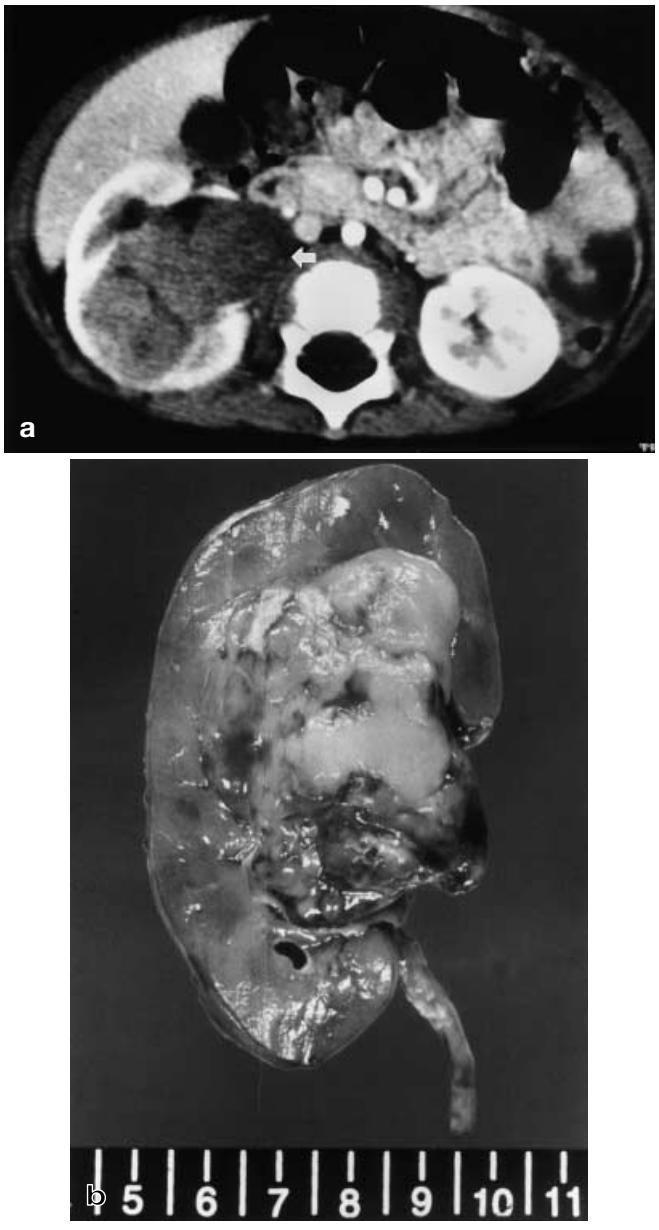

Fig. 1 a, b A centrally located rhabdoid tumour within the right kidney in 12-month-old boy. **a** Contrast-enhanced CT shows a poorly enhancing mass occupying the renal pelvis of the right kidney, which is protruding into the renal hilum (*arrow*). The renal pelvis is expanded by the tumour. There is no subcapsular haematoma. **b** Cut surface of the gross specimen shows the mass confined in the renal sinus. Note involvement of the renal hilum

Materials and methods

We retrospectively reviewed the clinical and imaging features in seven patients with a histological diagnosis of RTK, identified from three participating institutions from January 1992 to December 1997. There were five boys and two girls, aged 6 months to 4.7 years (mean 2 years; median 1.5 years). The major initial symptoms and signs included gross haematuria ($n = 4$) or palpable mass ($n = 3$).

All seven patients had undergone abdominal CT evaluation and two had also undergone abdominal MRI. All CT scans were performed with IV contrast medium and non-contrast CT scans were available in three cases. Abdominal US was available in three cases. Three CT scans and one MRI study of the brain were performed. Imaging studies were performed with a wide variety of equipment and over many years; thus, protocols regarding imaging parameters, contrast material injection and so forth were not standardized.

We analysed the tumour size, tumour margin, tumour location, subcapsular haematoma, tumour necrosis or haemorrhage, calcification, lymphadenopathy and distant metastases. Determination of tumour location within the involved kidney was obtained by a review of all preoperative renal imaging studies. A centrally located tumour was defined as extending centrally into the kidney beyond the renal medulla into the renal sinus and expanding the renal pelvis (Fig. 1). A peripherally located tumour was defined as being peripherally located in the kidney and displacing the renal pelvis (Fig. 2).

Results

Of the seven patients with RTK, four were on the right and three were on the left. Tumour diameter ranged from 5 to 12 cm (mean 9 cm). The margins of the tumour were well-defined from adjacent renal parenchyma in three cases and ill-defined in four cases. The surface of the tumour was lobulated in all cases (Fig. 3). Four tumours were located predominantly in central portion (Fig. 1) in the involved kidney and three tumours were predominantly in a peripheral portion (Figs. 2, 3). Among the centrally located RTKs, one case was confined within the kidney (Fig. 1) and three cases were located mainly in the central portion with peripheral extension (Fig. 4).

In four of the seven cases, the RTK was associated with a subcapsular haematoma, which was large in those three cases in which the tumour had a peripheral location (Figs. 2, 3). The remaining one case with a small subcapsular haematoma had a mainly centrally located RTK with peripheral extension. Areas of low attenuation compatible with haemorrhage or tumour necrosis were detected in all cases. Calcifications were present in three cases (two on non-contrast CT and one on US; Fig. 4 a). Retroperitoneal lymphadenopathy was noted in four cases.

Of three children with a follow-up study, two had local recurrence and one had pulmonary metastasis. Brain tumour was present in one of four cases on brain CT, or MR and the brain tumour (not surgically confirmed) was located in the midline of the posterior fossa (Fig. 3 c).

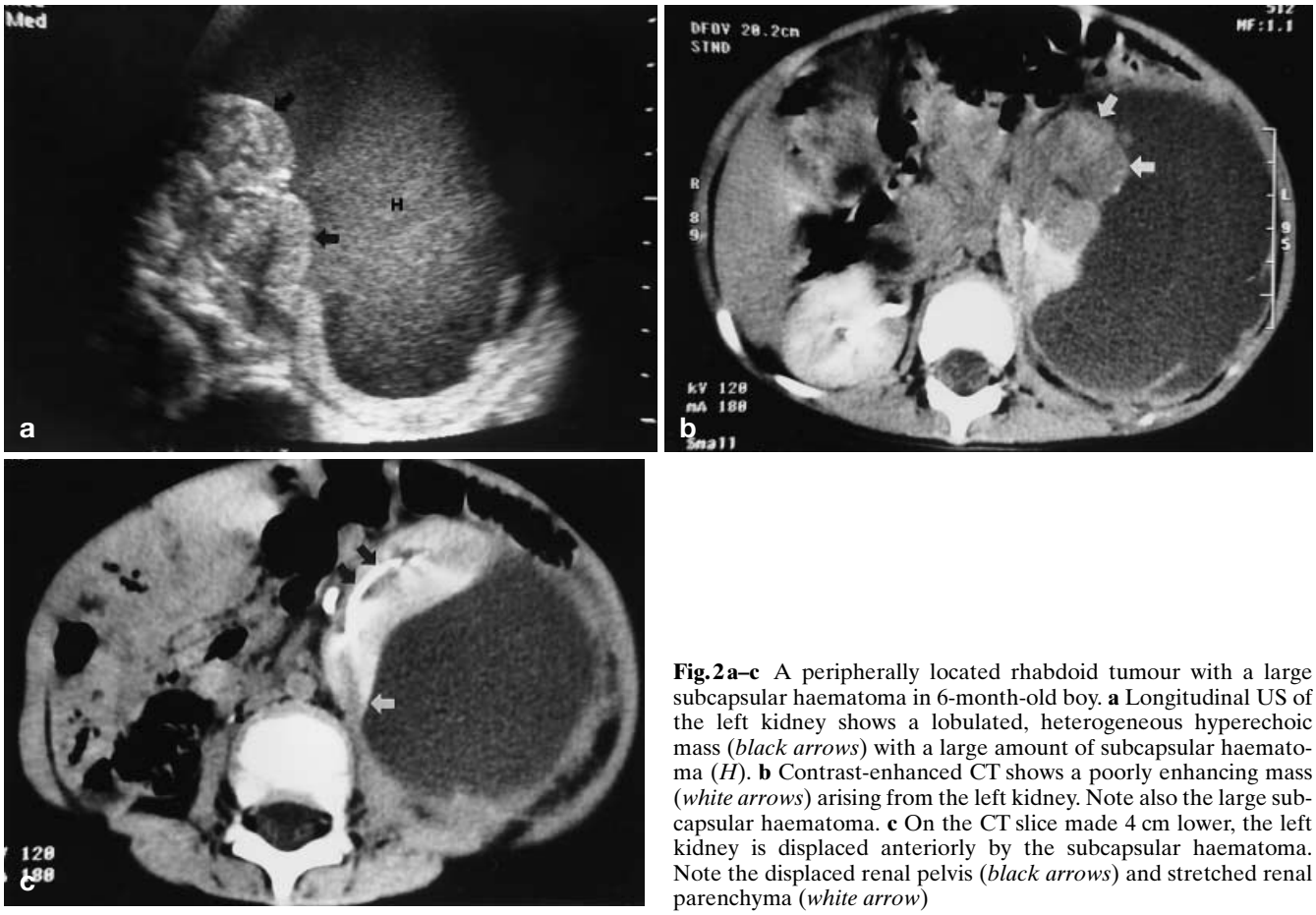

Fig. 2 a–c A peripherally located rhabdoid tumour with a large subcapsular haematoma in 6-month-old boy. **a** Longitudinal US of the left kidney shows a lobulated, heterogeneous hyperechoic mass (*black arrows*) with a large amount of subcapsular haematoma (*H*). **b** Contrast-enhanced CT shows a poorly enhancing mass (*white arrows*) arising from the left kidney. Note also the large subcapsular haematoma. **c** On the CT slice made 4 cm lower, the left kidney is displaced anteriorly by the subcapsular haematoma. Note the displaced renal pelvis (*black arrows*) and stretched renal parenchyma (*white arrow*)

Discussion

RTK is the most aggressive malignant neoplasm of the kidney in children and is a rare tumour, accounting for 2–3 % of all renal neoplasms in the National Wilms' Tumor Study (NWTS) [6]. RTK occurs in infants and young children and is extremely rare over the age of 5 years. The median age at diagnosis is 11 months [1]. Weeks et al. [1] suggested that RTK might originate in primitive cells involved in the formation of the renal medulla. RTK usually arises from the perihilar renal parenchyma, infiltrating the medulla, renal sinus and collecting system [1]. This tumour is often large at presentation (> 9 cm) and spreads throughout the renal parenchyma.

RTK has a high relapse rate and a mortality of over 80 %. Of all primary paediatric renal tumours, the prognosis of children with RTK is the poorest. There is a high rate of local tumour recurrence following surgical resection. Metastatic spread to the lungs, liver and brain is common, and metastases to the brain and skeleton have also been reported [1]. Invasion of the renal vein is common. An association of RTK with a second primary brain tumour or metastasis has been reported, and the primary brain tumours are mostly of posterior fossa origin and include medulloblastoma, primitive neuroectodermal tumour, ependymoma and cerebellar or brainstem astrocytoma [1–4].

Chung et al. [9] noted several CT findings which were suggestive of RTK – calcification, subcapsular haematoma and lobular appearance in a large, centrally located and heterogeneous renal mass in a child. They reported that subcapsular fluid collection was seen in 8 (44 %) of 18 cases of RTK. Agrons et al. [10] also indicated that a peripheral crescent-shaped fluid collection was characteristic of RTK, but other renal neoplasms in children also show this sign. They described a peripheral crescent of fluid attenuation on CT scans in 15 (71 %) of the 21 patients with RTK and 19 (12 %) of 153 patients with other renal neoplasms. Because other renal neoplasms were more frequent than RTK, the overall incidence of a peripheral crescent-shaped fluid collection for RTK was lower than for other renal neoplasms. Thus they reported that this finding was not pathognomonic for the diagnosis of RTK.

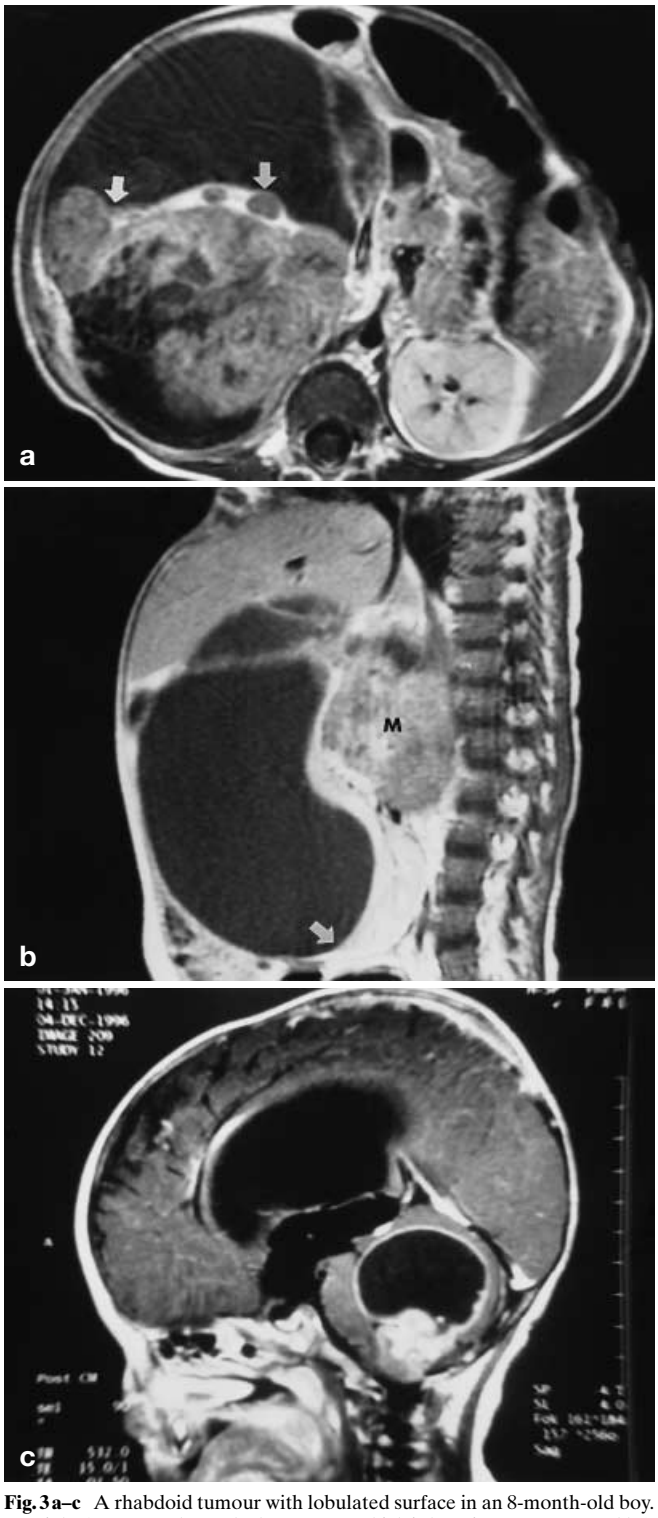

Fig. 3a–c A rhabdoid tumour with lobulated surface in an 8-month-old boy. **a** Axial T1-W MRI shows the large mass, which is hypointense to normal left kidney, with lobulated surfaces (*white arrows*). Note also the large subcapsular haematoma. **b** Sagittal, T1-W gadolinium-enhanced MRI shows a poorly contrast-enhanced mass (*M*) in the upper pole of the right kidney. Note the large subcapsular haematoma and stretched renal parenchyma (*white arrow*). **c** Sagittal, T1-W gadolinium-enhanced MRI of the brain shows a midline, posterior fossa cystic mass with an intensely enhancing solid portion

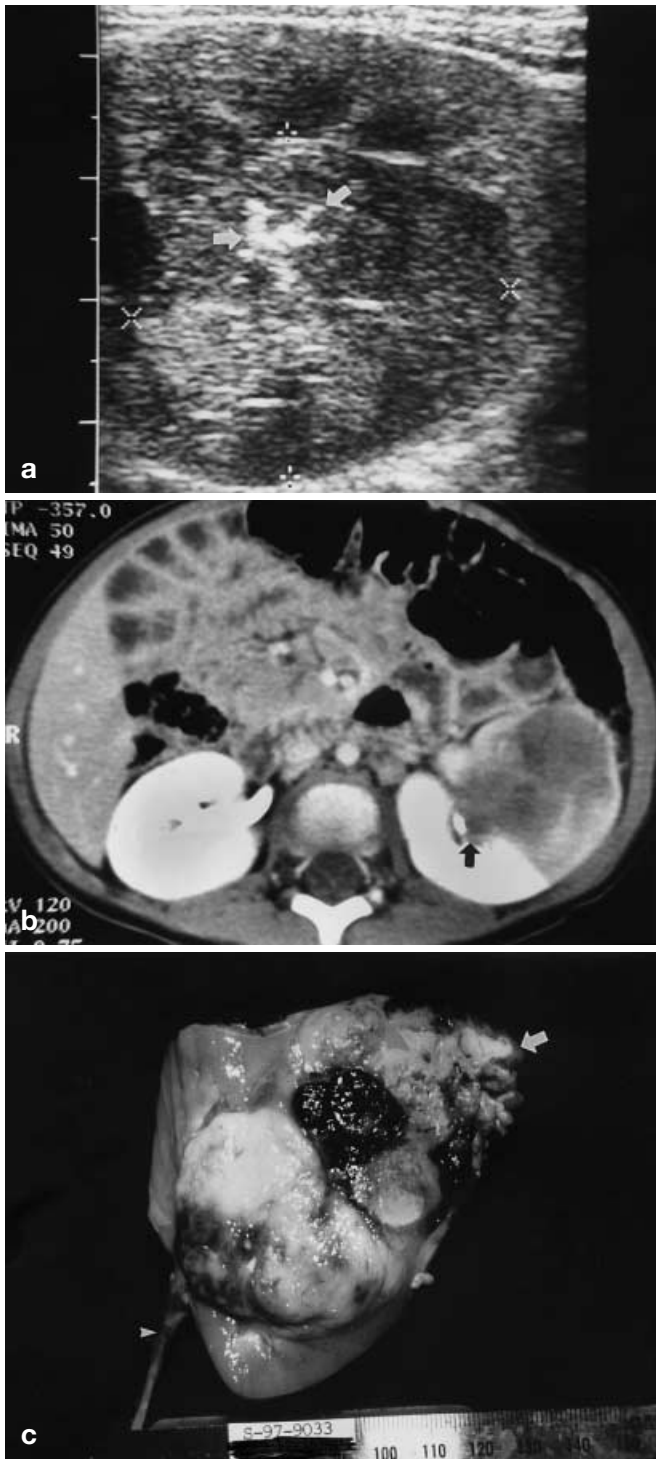

Fig. 4a–c A rhabdoid tumour with involvement of the renal hilum and peripheral extension in an 18-month-old girl. **a** Transverse US demonstrates an inhomogeneous echogenic mass in the left kidney. Note hyperechoic calcifications (*white arrows*) in the kidney. **b** Contrast-enhanced CT shows a mass of lower attenuation than that of the renal parenchyma. Note involvement of renal hilum (*black arrow*) and peripheral extension. **c** The surgical specimen, sectioned longitudinally, confirms the mass arising from the central part of the kidney, invading the sinus and reaching the renal capsule (*white arrow*). Also note invasion of the proximal ureter (*arrowhead*)

In our study, four of seven cases had subcapsular fluid collections. Three cases with a large subcapsular haematoma showed mainly peripherally located RTK, whereas centrally located RTK showed absent or small subcapsular haematoma. Montgomery et al. [11] reported that one case was located centrally without subcapsular haematoma. Eftekhari et al. [12] reported a centrally located tumour without subcapsular haematoma and a large tumour nearly replacing the renal parenchyma associated with a large subcapsular haematoma. We suggest that a centrally located RTK tends to have a small or absent subcapsular haematoma, whereas a mainly peripherally located RTK or a RTK nearly replacing the renal parenchyma tends to have a large subcapsular haematoma.

A lobular appearance of the RTK was clearly present in 8 (44 %) of 18 tumours in the series of Chung et al. [9]. In our study, the lobulated surface was seen in all cases. Although not mentioned in the cases reported except for those of Chung et al. [9], we suggest that this finding may be helpful for the diagnose of RTK.

Calcification was seen on two of four non-contrast CT scans in the series of Chung et al.; these calcifications were described as hyperdense linear areas outlining the tumour lobules. Panuel et al. [13] reported a RTK with stippled calcification on CT. In our cases, calcifications were present in three cases.

In conclusion, the imaging findings of RTK are subcapsular haematoma, the lobulated surface of the tumour, calcification and tumour necrosis. Knowledge of these features may assist in the diagnosis of RTK.

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