

# An unusual presentation of an intra-abdominal rhabdomyosarcoma

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Rhabdomyosarcoma (RMS) is a common neoplasm, representing 5 - 10% of malignant solid tumours in childhood, and is the commonest soft-tissue sarcoma in the paediatric age group. Even though the tumour usually arises from striated muscle, it frequently originates from sites devoid of striated muscle such as the urinary bladder, prostate gland and gallbladder.

We present an unusual case of a large intra-abdominal embryonal RMS which demonstrated a mesenteric cleft inferolaterally with feeding vessels entering the mass. This unique presentation of embryonal RMS proved to be a diagnostic challenge.

## Case report

A 23-month-old boy was brought to casualty with a 2-week history of persistent vomiting associated with an unknown duration of progressive abdominal distension. On physical examination he was pale and dehydrated and a large abdominal mass, which was separable from the liver and spleen, was palpated. An abdominal radiograph demonstrated bulging of the peritoneal lines with a paucity of bowel gas pattern. Abdominal ultrasound (US) demonstrated a large inhomogeneous intra-abdominal mass that extended into the pelvis. Owing to the patient's poor clinical condition but normal renal function tests, computed tomography (CT) was performed urgently. Magnetic resonance imaging (MRI) is a better investigation for the evaluation of soft-tissue masses, but has limited use in an emergency setting because of the need for general anaesthesia.

## CT findings

A CT scan of the abdomen with intravenous and oral contrast demonstrated a well-defined, non-encapsulated intra-abdominal (intra-peritoneal) mass that extended into the pelvis (Figs 1 - 3). The mass enhanced inhomogeneously and predominantly centrally with peripheral hypodensities suggestive of necrosis (Figs 1 and 2). It was poorly separable from the abdominal wall and bladder but displayed no features of bladder invasion on delayed views (Fig. 3). Associated bilateral hydronephrosis and hydro-ureters were demonstrated due to obstruction of the ureters by the mass at the level of the pelvis (Figs 3 and 4). No areas of calcification were demonstrated. A unique feature was a mesenteric cleft with the prominent feeding artery sign, i.e. visualised feeding vessels entering the mass inferolaterally (Fig. 1).

The stomach and surrounding bowel were laterally displaced by the mass. There were no features of ascites or metastases to the liver, lung and visualised bone. The prostate gland, scrotal sac and contents were normal.

## Differential diagnoses

The differential diagnoses for this radiological presentation include rhabdomyosarcoma, mesenchymal tumours, lymphoma and giant teratoma.

Owing to the extensive, predominantly peripheral necrosis of the mass, an US-guided fine-needle aspirate (FNA) was non-diagnostic. Due to worsening bowel obstruction, a decision was made to surgically remove the mass.



Fig. 1. Coronal post-contrast CT image demonstrating a left mesenteric cleft with feeding vessels entering the mass inferolaterally.

## Surgery

The tumour was surgically removed after the vessels in the inferolateral mesenteric cleft were successfully isolated and tied off. The tissue was sent for histological evaluation.

## Pathological findings

**Macroscopic evaluation** of the specimen revealed a non-encapsulated mass weighing 1 266.5 g. It was attached to omentum and adherent to the fundus of the bladder as well as the abdominal wall. No calcifications or bony elements were present.

**Microscopy** confirmed the presence of a high-grade malignant tumour consisting of primitive mesenchymal cells at various stages of myogenesis in the form of rhabdomyoblasts. Positive staining with myogenin, myoD1 and desmin confirmed rhabdomyoblastic/skeletal muscle differentiation. The immunophenotypic features were in



Fig. 2. Sagittal post-contrast CT image illustrating the extent of the mass as well as central contrast enhancement.

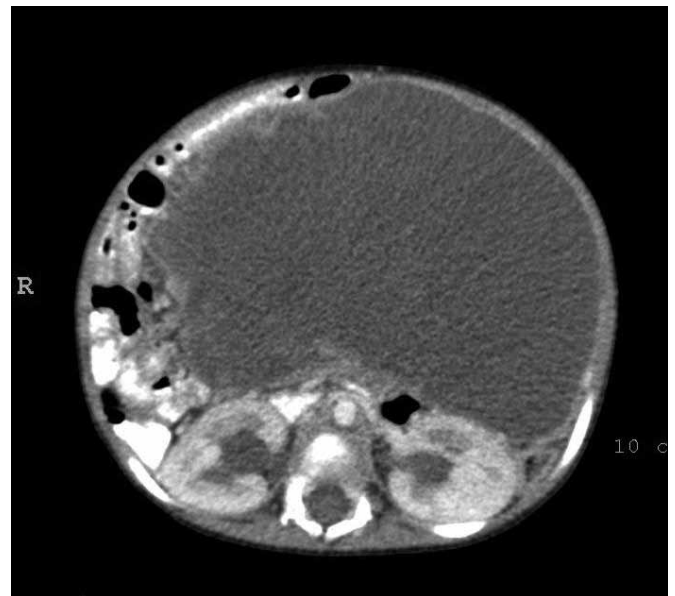


Fig. 4. Axial post-contrast CT illustrating the predominantly hypodense intra-abdominal mass with lateral displacement of bowel and bilateral hydronephrosis.

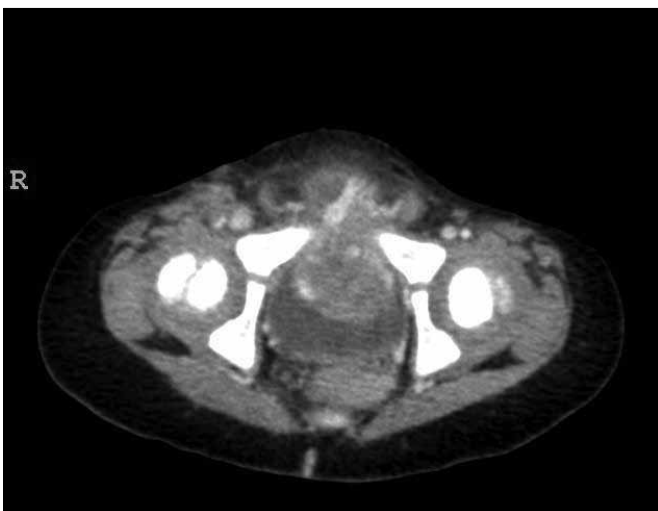


Fig. 3. Axial post-contrast CT at the level of the pelvis demonstrating the mass abutting the bladder anteriorly.

keeping with an embryonal rhabdomyosarcoma (RMS), spindle cell variant (Fig. 5).

The tumour cells showed an enhanced peritheliomatous growth pattern with large areas of coagulative tumour necrosis. There was focal invasion into the muscularis propria of bowel. Microscopic invasion of the bladder and abdominal wall was not demonstrated.

**Diagnosis**

Intra-abdominal embryonal RMS, spindle cell variant.

**Post-surgical treatment**

The patient had postoperative chemotherapy and radiation using IRS (International Rhabdomyosarcoma Study) regimen 36, with abdominal radiation of 1.8 Gy daily to a total of 36 Gy. He did well as a result of having no metastases and a favourable histological sub-type of embryonal RMS (spindle cell variant).

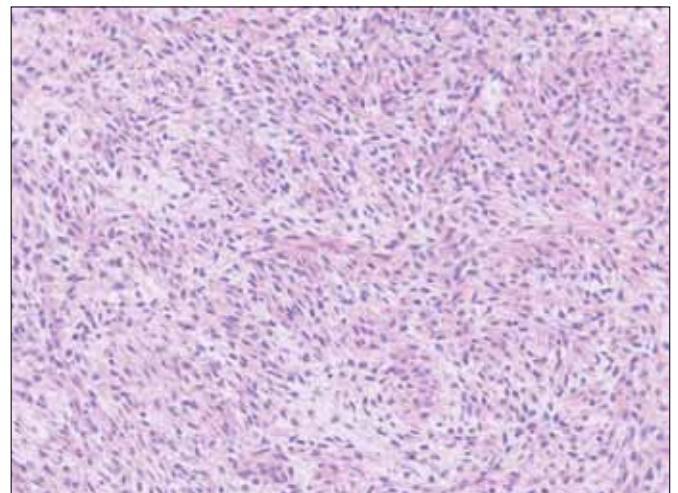


Fig. 5. Photomicrography of the tumour illustrating uniform spindle cells with eosinophilic fibrillar cytoplasm. Immunorexpression of myogenin, myoD1 and desmin confirmed rhabdomyblastic differentiation, in keeping with an embryonal rhabdomyosarcoma, spindle cell variant.

**Discussion**

RMS is a common neoplasm, representing 5 - 10% of malignant solid tumours in childhood, and is the commonest soft-tissue sarcoma in the paediatric age group.<sup>1</sup> Among childhood tumours RMS ranks fourth in frequency after central nervous system tumours, neuroblastomas and nephroblastomas. Although RMS usually arises from striated muscle, it frequently originates from sites devoid of striated muscle such as the urinary bladder, prostate gland and gallbladder.<sup>2-4</sup>

The two main histological subtypes of RMS affecting the paediatric age group are embryonal (with botryoid and spindle cell variants) and alveolar. The remaining cases are of pleomorphic or undifferentiated histology. Embryonal RMS accounts for nearly 70% of all cases and affects children less than 8 years of age.<sup>3,4</sup> It commonly arises in the mucosal-lined structures of the nasopharynx, auditory canal and genito-urinary and gastro-intestinal tracts.<sup>4</sup> Botryoid RMS is a variant of the embryonal type and occurs in hollow structures such as the vagina, bladder and biliary tract. Spindle cell RMS, as described in our patient, is a rare variant of the embryonal type with a favourable



**Rhabdomyosarcoma is the commonest soft-tissue sarcoma in the paediatric age group.**

clinical course. This tumour tends to arise in younger patients, with a predilection for males. Spindle cell RMSs commonly arise from the paratesticular soft tissue (38%), followed by the head and neck region (27%) and rarely the abdomen, retroperitoneum and urinary bladder. There is a paucity of cytogenetic evaluations of spindle cell RMSs.<sup>5</sup> Alveolar RMS accounts for 20% of all cases and has the worst prognosis. It commonly occurs on the trunk and lower extremities.<sup>4</sup>

Approximately 10 - 12% of RMSs are intra-abdominal.<sup>6</sup> Abdominal tumours commonly involve the retroperitoneum, biliary tree and abdominal wall. Owing to their sheltered location these masses are discovered very late, as in our patient, making it difficult to determine the precise organ of origin.<sup>6</sup> The tumour described in our case is not a classic RMS. Although it was poorly separable from the abdominal wall there was no microscopic evidence to suggest its origin. Owing to the size of the intra-abdominal component, the lack of bladder infiltration and the normal prostate gland and scrotum, the mass was considered to be of intra-abdominal origin, organ unknown. The intra-abdominal origin of the tumour was further supported by the prominent feeding artery sign, where the feeding vessels are visualised on CT in the mesenteric cleft<sup>7</sup> (Figs 1 and 2).

The mesenteric cleft is an unusual finding, adding to the unique presentation of this case. To our knowledge an abdominal RMS associated with a mesenteric cleft and prominent feeding artery sign has not been described in the literature. The predominant central enhancement of this mass, with peripheral hypodensities visualised on CT, correlated with the microscopic findings of an enhanced peritheliomatous growth pattern with large areas of coagulative tumour necrosis. This pattern of necrosis in the mass is supported by the vascular supply, i.e. the central mesenteric cleft vessels. Owing to this pattern of necrosis, pre-operative diagnosis by US-guided FNA was non-diagnostic.

## Conclusion

This presentation of an intra-abdominal embryonal RMS, spindle cell variant, of unknown origin is unusual. The mesenteric cleft with the prominent feeding artery sign adds to this unique presentation. Our patient had good prognostic features, which included no metastases and the histological sub-type of spindle cell variant. RMS should always be considered as a differential diagnosis in a paediatric patient presenting with an intra-abdominal mass not clearly arising from a specific organ, as RMS is a heterogeneous disease that may arise in virtually any organ or tissue except bone.

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