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Case Report

Rare and aggressive: imaging insights of intraperitoneal and retroperitoneal rhabdomyosarcoma in children and young adults

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ABSTRACT

This case series presents two cases of rhabdomyosarcoma (RMS), a rare malignancy of mesenchymal origin that accounts for 5% of all paediatric cancers and is extremely rare to present in adults. It can develop in various sites, including the head and neck, thorax, liver, biliary tract, retroperitoneum, urinary bladder, vagina, extremities, and paratesticular locations. Herein we present a case series of intrabdominal RMS, comprising the first case, an intraperitoneal RMS in a 2-year-old male child, and the second case, a retroperitoneal RMS in a 22-year-old male. Intraperitoneal RMS is extremely rare. Intraperitoneal and retroperitoneal RMS are aggressive tumours and have a poor prognosis. Both cases were of the spindle cell type of RMS, which is the most uncommon type.

Keywords: Rhabdomyosarcoma, Intra-peritoneal tumor, Retro-peritoneal tumor, Aggressive, Soft-tissue sarcoma, Paediatric oncology

INTRODUCTION

Rhabdomyosarcoma (RMS) is a rare malignancy of mesenchymal origin that accounts for approximately 5 % of all paediatric solid tumours.[1] It is the third most common extracranial malignancy after neuroblastoma and Wilms' tumour.[2] Soft tissue sarcomas represent <1% of all adult solid tumour malignancies, and RMS accounts for only 3% of all soft tissue sarcomas in adults.[3]

It arises from skeletal muscle progenitor cells and can occur in various locations throughout the body, like the head and neck, thorax, liver, biliary tract, retroperitoneum, urinary bladder, vagina, and extremities, with the most common site being the head and neck.[4]

Intrabdominal RMS (intraperitoneal and retroperitoneal) is a rare entity and has a poor prognosis. The prognosis of RMS largely depends on the tumour's location and stage. Here, we report two cases of intra-abdominal RMS.

CASE REPORTS

Case 1

Intraperitoneal rhabdomyosarcoma in a 2-year-old male

A two-year-old male child presented with abdominal pain and a lump for three months. On physical examination, a palpable non-tender mass was identified in the right umbilical and epigastric region.

We did an ultrasound of the patient, which revealed a large heterogeneously hypoechoic mass within the peritoneal cavity extending from the epigastric region to the hypogastric region. (Figure 1). Bowel loops were seen posterior to it, with no continuity of bowel loops with the mass present. All other solid organs were normal.

Contrast-enhanced computed tomography was performed post-distension of the bowel loop with positive contrast. It revealed a large well-defined hypodense mass of size

8.1×12.1×12.8 cm (AP×Tra×CC) in the peritoneal cavity extending from L1 vertebral level to S2. It was showing feeding vessels from the anterior abdominal wall on the venous phase and homogenous enhancement on the delayed phase with a few non-enhancing necrotic areas within (Figure 2).

Omental caking was seen anterosuperiorly around the mass (Figure 3). There was no haemorrhage or calcification seen within it. There was no significant lymphadenopathy. There was no ascites or peritoneal deposit. The rest of the organs were normal. There was no vascular invasion by the mass.

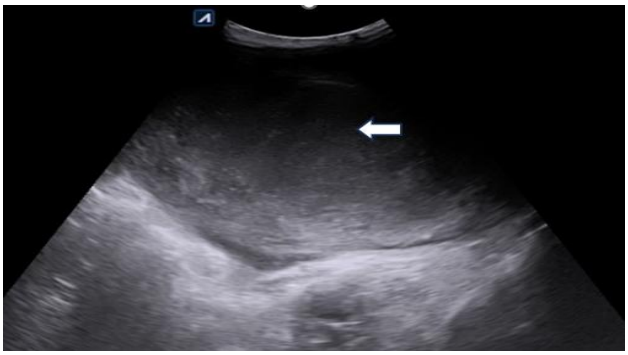

Figure 1: Ultrasound of the abdomen reveals a large heterogeneously hypoechoic mass lesion (white arrow) with bowel loops displaced posteriorly.

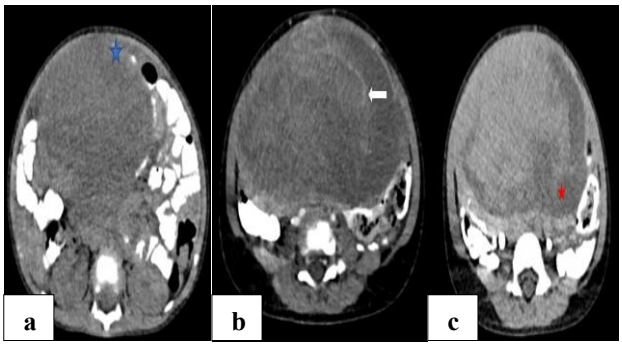

Figure 2: Computed tomography of the abdomen (axial sections) shows (a) hypodense mass in the intraperitoneal cavity on a non-contrast scan (blue asterisk), (b) on the venous phase, feeding vessels are seen (white arrow) arising from the abdominal wall, and (c) on the delayed phase, enhancement is seen with non-enhancing necrotic areas (red asterisk).

The patient underwent surgical resection by exploratory laparotomy via the supraumbilical transverse incision. The mass was seen within the mesentery, adhering to the bowel loops and bladder. It was excised completely after being separated from the bowel wall and bladder.

The Histopathological examination of the surgical specimen after USG-guided biopsy was suggestive of spindle cell rhabdomyosarcoma. On immune-histochemistry, the cells were positive for desmin and CD99. Occasional cell positivity was noted for myogenin, too.

The patient was planned for chemotherapy; however, the child expired within 1 month of surgical resection.

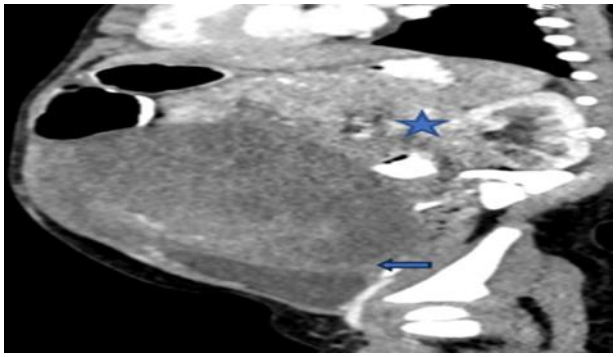

Figure 3: Computed tomography of the abdomen (sagittal section) shows the extensive omental caking (blue asterisk). It is abutting the urinary bladder inferiorly (blue arrow).

Case 2

Retroperitoneal rhabdomyosarcoma in a 22-year-old male

A 22-year-old male presented with a 3-month history of abdominal pain and distension. On physical examination, a non-tender, hard palpable mass was identified predominantly in the left hypochondrium and lumbar region.

Ultrasonography was performed, which showed a large heterogeneously hypoechoic mass in the abdominal cavity anterior to the left psoas muscle (Figure 4).

CEMRI abdomen revealed a well-circumscribed encapsulated retroperitoneal mass of size 10.8×11.5×12.6 cm (AP×Tra×CC) on the left side, crossing the midline, extending from L3 vertebral level to S2 vertebral level. The mass was hypointense on T1WI and heterogeneously hyperintense on T2WI (Figure 5a).

Multiple septations and whorled areas, appearing hypointense on T2WI, were seen within the mass (Figure 5b). On post-contrast T1WI, heterogeneous enhancement of solid areas, septae, and capsule was noted (Figure 5c). Focal areas within the mass showed diffusion restriction on DWI and corresponding ADC maps (Figure 6). The mass displaced the aorta medially. The bowel loops were displaced superiorly and to the right side. It was abutting the anterior abdominal wall anterolaterally and the iliopsoas muscle posteriorly. There was a focal loss of the fat plane between the left psoas and the mass at the L5-S1 intervertebral level (Figure 7).

There was no significant lymphadenopathy, ascites, peritoneal deposit, or omental caking. The rest of the solid

organs were normal. There was no vascular or solid organ invasion seen.

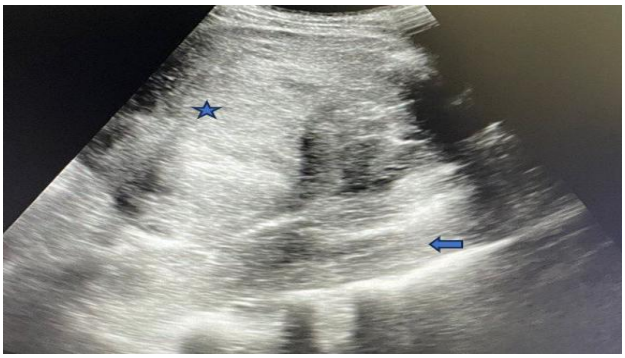

Figure 4: Ultrasound of the abdomen reveals a large heterogeneously hypoechoic mass lesion anterior to the left psoas muscle.

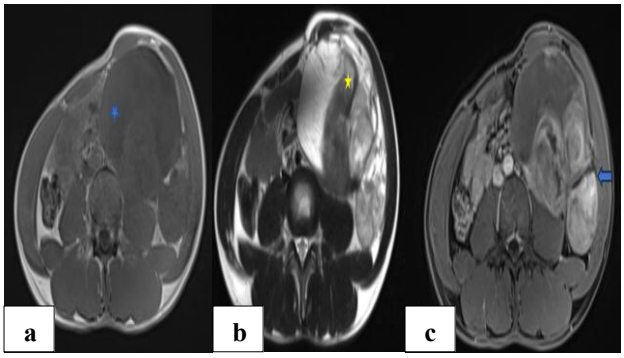

Figure 5: MRI abdomen axial images show (a) T1WI axial image shows a well-defined hypointense mass in the retroperitoneum on the left side crossing the midline (blue asterisk), (b) T2WI axial image shows a heterogeneously hyperintense mass with multiple hypointense septae, giving a whorled appearance. (yellow asterisk), and (c) post-contrast T1WI shows enhancement of solid components (blue arrow).

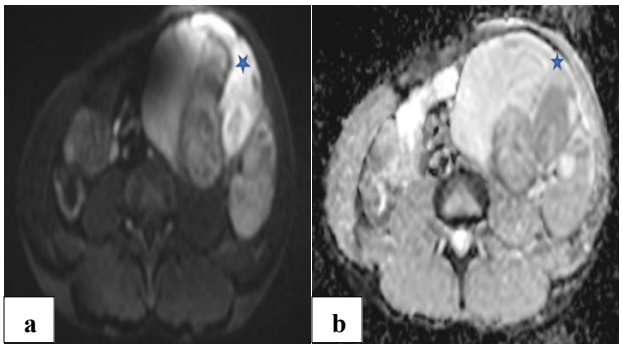

Figure 6: MR axial images (a) DWI, and (b) corresponding ADC map show a focal area (blue asterisk) within the mass appearing bright on DWI and dark on the ADC map.

USG-guided core biopsy was done. The histopathological examination suggested spindle cell RMS. Immunohistochemistry of tumour cells revealed positivity for desmin and myogenin.

The patient underwent surgical resection of the mass and received adjuvant chemotherapy. He was disease-free at the 2-year follow-up.

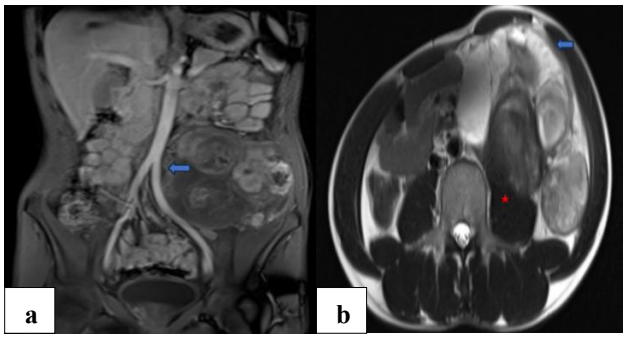

Figure 7: MRI images showing the relationship and extension of the mass (a) coronal T1WI post-contrast image shows the mass abutting the aorta and causing its medial displacement (blue arrow); however, no vascular invasion is seen. It is displacing the bowel loops superiorly and to the right side, and (b) axial T2WI MR image shows focal loss of fat plane with left psoas (red asterisk). It is abutting the anterior abdominal wall anteriorly (blue arrow).

DISCUSSION

Background

RMS is a malignant and aggressive tumour. The intraperitoneal and retroperitoneal sites for RMS are rare presentations with only a few cases of intraperitoneal RMS described in the literature.[5]

RMS usually affects the paediatric population less than 10 years of age, with a male predilection.[6] Most cases are sporadic, but an association with various syndromes like Li-Fraumeni syndrome, neurofibromatosis type 1, Rubinstein–Taybi syndrome, Beckwith–Wiedemann syndrome, Costello syndrome, Noonan syndrome, hereditary retinoblastoma, and Gorlin basal cell carcinoma nevus syndrome is seen.[7]

Pathogenesis

The exact pathogenesis of RMS is not fully understood. However, it is believed to arise from the proliferation of undifferentiated mesenchymal cells that have the potential to differentiate into skeletal muscle cells and can develop in areas where skeletal muscle is not normally found.[8,9]

Based on histology, RMS is of four types: Embryonal, pleomorphic spindle cell/sclerosing, and alveolar. Embryonal RMS is the most common subtype. Embryonal RMS can be of three types: myxoid or botryoid, spreading, and epithelioid. Embryonal and alveolar are seen in the

paediatric and adolescent populations. Pleomorphic variety is seen in adults. The spindle cell type is the most uncommon.[8-10]

Clinical presentation

The clinical presentation of RMS depends on the location and size of the tumour. Intraperitoneal and retroperitoneal RMS can present with abdominal pain, distension, and a palpable mass. Other symptoms may include nausea, vomiting, and weight loss. Complications like small bowel obstruction, neurovascular compression, and hydroureteronephrosis can occur due to mass effects. In some cases, RMS can metastasize to other organs, such as the lungs and bones.

Imaging modalities

Imaging modalities, such as ultrasound, CT, and MRI, are essential for the diagnosis and staging of RMS.

Ultrasound can provide initial information about the location and size of the tumour.

Cross-sectional studies, such as CT and MRI, can reveal the extent of the tumour and its relationship with surrounding structures. They demonstrate the nodal and distant spread of the tumour, aiding in staging and surgical planning. The location in the retroperitoneum or intraperitoneal area can be distinguished based on the displacement of retroperitoneal structures.

Retroperitoneal RMS usually displaces the sigmoid colon supero-anteriorly, the urinary bladder inferiorly, and abuts the iliopsoas posteriorly. Intraperitoneal RMS appears as a mesenteric mass and is associated with omental caking, ascites, mesenteric nodules, pseudomyxoma peritonei, and peritoneal deposits. RMS exhibits calcification, necrosis, post-contrast heterogeneous enhancement, and feeding vessels within.[10-12] Hematogenous spread is observed with metastasis to the lung, liver, bone, lymph nodes, and brain.

The differentials for retroperitoneal RMS are neurogenic tumours, nerve sheath tumours, neuroblastoma, ganglioneuroma, leiomyosarcoma, fibrosarcoma, foetal rhabdomyoma, triton tumor, lymphoma, and undifferentiated pleomorphic sarcoma. Inflammatory myofibroblastic tumour, mesenteric fibromatosis, mesothelioma, desmoplastic small round blue tumours, immature teratoma, yolk sac tumour, malignant fibrous histiocytoma, and lymphoma are mimickers of intraperitoneal RMS.[11,13,14]

Diagnosis is confirmed with histopathology and immunohistochemistry. Microscopy shows malignant spindle cells with atypical mitosis arranged in a herringbone pattern. Undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma) can mimic RMS on microscopy, and thus, IHC helps in confirming RMS, as cells in RMS show positivity for specific markers of skeletal muscle differentiation like myogenic, muscle-specific action, desmin, and MyoD1.[15]

The PET scan is useful for demonstrating metastasis, staging, and is also used in follow-up to monitor recurrence or residual tumour.

Treatment and outcome

The treatment of intrabdominal RMS follows the IRSG (Intergroup Rhabdomyosarcoma studies group) protocol in both adult and paediatric cases, which involves a multimodal approach that includes surgery, chemotherapy, and radiation therapy.[16]

Intraperitoneal and retroperitoneal RMS have a poor prognosis due to the difficulty in achieving complete surgical resection and the high risk of recurrence. However, with aggressive multimodal therapy and regular follow-up, some patients can achieve long-term survival and a disease-free period.

CONCLUSION

RMS is a rare malignancy of mesenchymal origin that can occur in various locations throughout the body. Intraperitoneal and retroperitoneal RMS are uncommon and associated with a poor prognosis. Histopathology and immunohistochemistry are important to differentiate it from other soft tissue tumours, mesenteric masses, and retroperitoneal tumours. Timely diagnosis, appropriate staging, and multimodal treatment can improve the chances of cure and long-term survival.

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