Case Report

Intraperitoneal Inflammatory Myofibroblastic Tumour: A Rare Intermediate Tumour of Paediatric Age Group

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Abstract

This case report highlighted a 9 month old child with failure to thrive. Incidental finding of left sided abdominal mass during elective admission for magnetic resonance imaging brain. Radiological investigation followed by histopathological examination revealed the presence of an inflammatory myofibroblastic tumour in abdomen. These were rarely reported intermediate tumours with a strong tendency for recurrence.

Keywords: Inflammatory; intraperitoneal; myofibroblastic; paediatric; pseudotumour

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Date of submission: 12 Dec, 2023 Date of acceptance: 28 Jun, 2024

Introduction

An inflammatory myofibroblastic tumour (IMT), also termed inflammatory pseudotumour, is a rare lesion of unclear pathogenesis. It can occur in intra-abdominal sites in children and can be confused with malignancy because of its large size and location. It is usually an 'intermediate' tumour, with a strong tendency for recurrence. Main modality of treatment is surgical excision. As these tumours are rare, correct diagnosis is important in order to avoid unnecessary wide surgical removal and aggressive chemotherapeutic regimens. Here, we presented a case report of a 9months old infant who presented with failure to thrive.

Case Report

A 9 month old boy was electively admitted to Hospital Tunku Mizan, 2 years ago for magnetic resonance imaging (MRI) brain, as part of investigation of macrocephaly. Incidentally noted to have left sided painless abdominal mass during that admission. An ultrasound abdomen done revealed a left lumbar mass, and child was referred to Hospital Cancelor Tunku Muhriz, Universiti Kebangsaan Malaysia, Cheras, Kuala Lumpur for further investigation. He was admitted under Paediatric Surgical team and noted to have failure to thrive. He was otherwise asymptomatic. There was no history of trauma. He was otherwise active at home. Full blood count revealed leukocytosis. Other blood investigations consisting of liver and renal function tests, peripheral blood film, alpha feto protein (AFP) and human chorionic gonadotropin (bHCG) was otherwise within normal range for age.

An abdominal x-ray showed displaced bowel loops to the right side indicating mass effect (Fig. 1). Another formal ultrasound was repeated which revealed a lobulated hypoechoic solid mass seen at left lumbar region with no vascularity seen within (Fig. 2). A computed tomography (CT) thorax, abdomen and pelvis (Fig. 3) was performed showing a large well defined multilobulated enhancing mass at the left side of the abdomen measuring 5.5 x 5.6 x 6.6cm (AP x W CC) with some areas of hypodensities within in keeping with necrotic component. Some areas of speckled calcification was seen within the mass.It appeared to arise intraperitoneally as it displaced the transverse and descending colon posteriorly. It

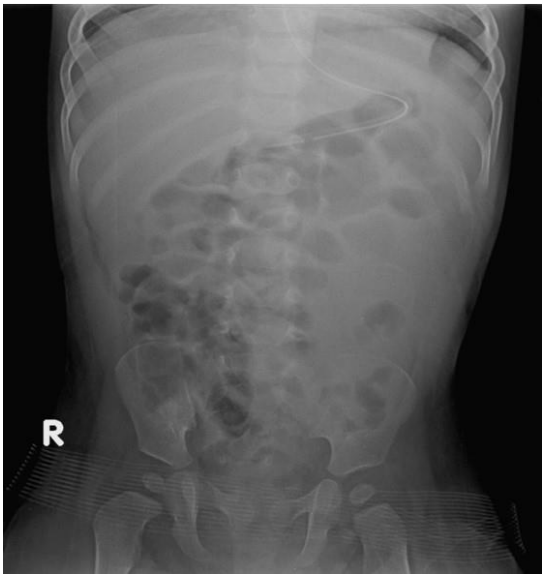

FIGURE 1: Abdominal x-ray showed small bowels displaced to the right side. No bowel dilatation

displaced most of the small bowel loops to the right side, with no bowel dilatation. Posteriorly, the mass compressed onto the anterior aspect of left kidney, with effacement of the anterior perinephric fat.

However, no claw sign to suggest renal in origin, and the left kidney showed normal enhancement with no evidence of obstructive uropathy. No renal vein thrombosis or enlarge lymph nodes. Left adrenal gland was normal. Ultrasound guided biopsy of left intraabdominal mass was performed which revealed a low to intermediate grade spindle cell neoplasm, in keeping with inflammatory myofibroblastic tumour.

The child underwent laparotomy with tumour and large bowel excision with double barrel colostomy. Intraoperative findings revealed an intraperitoneal tumour at the left hypochondrium extending to the left lumbar region. It was adhered to greater omentum and splenic flexure of colon. He was on Total Parenteral Nutrition (TPN) feeding after the surgery, until he underwent reversal of stoma 3 weeks later. Child was later discharged home well on day 3 after the second surgery.

Histopathology examination (Fig. 4) resulted in a lesion consisting of diffuse sheets of neoplastic spindle-shaped cells arranged fascicules embedded within collagenous stroma. The stroma was moderately infiltrated by lymphocytes, plasma cells and occasional eosinophils. A few small fragment of tumour tissue was seen adhered to the serosal layer but not infiltrating into the muscularis propria. A fragment of colonic tissue with relatively preserved colonic mucosa architecture colonic mucosa showed no evidence of dysplasia or malignancy. Immunohistochemical study carried out there is a positivity of smooth muscle actine (SMA), anaplastic lymphoma kinase (ALK) as well as vimentin (Figure 4) and negativity for S100. As a conclusion the histological aspect was in favour of an IMT (inflammatory pseudotumour).

Child was last seen by Peadiatric Surgical team early January 2022 .Clinical assessment showed that child had returned back to normal growth centile. A formal ultrasound was also performed which showed no recurrence or distant metastasis.

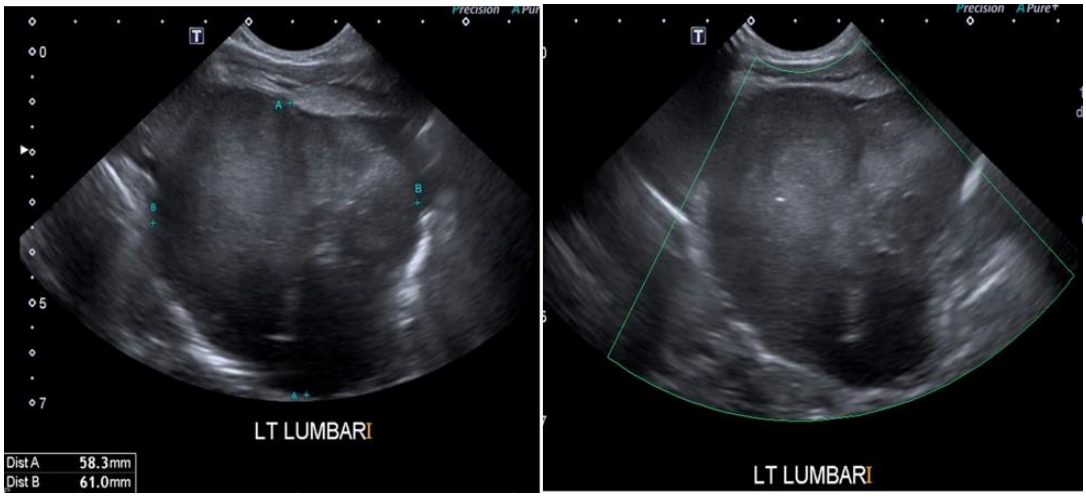

FIGURE 2: Ultrasound abdomen showed a lobulated hypoechoic solid mass seen at left lumbar region with no vascularity seen within. Some areas of calcification seen within the mass

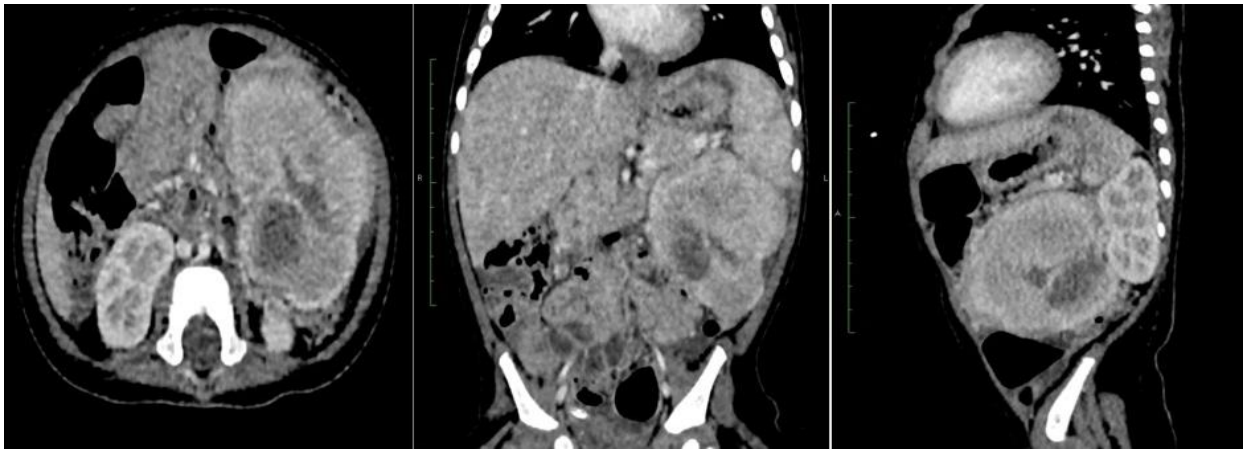

FIGURE 3: CECT abdomen images (in axial, sagittal and coronal slices) showed a large well circumscribed multilobulated enhancing mass at the left side of abdomen, with areas of hypodensities in keeping with necrotic component. Small bowels were displaced to the right side. It was intraperitoneal in location, as it displaced part of the descending colon posteriorly. No claw sign to suggest renal in origin. No enlarged lymph nodes.

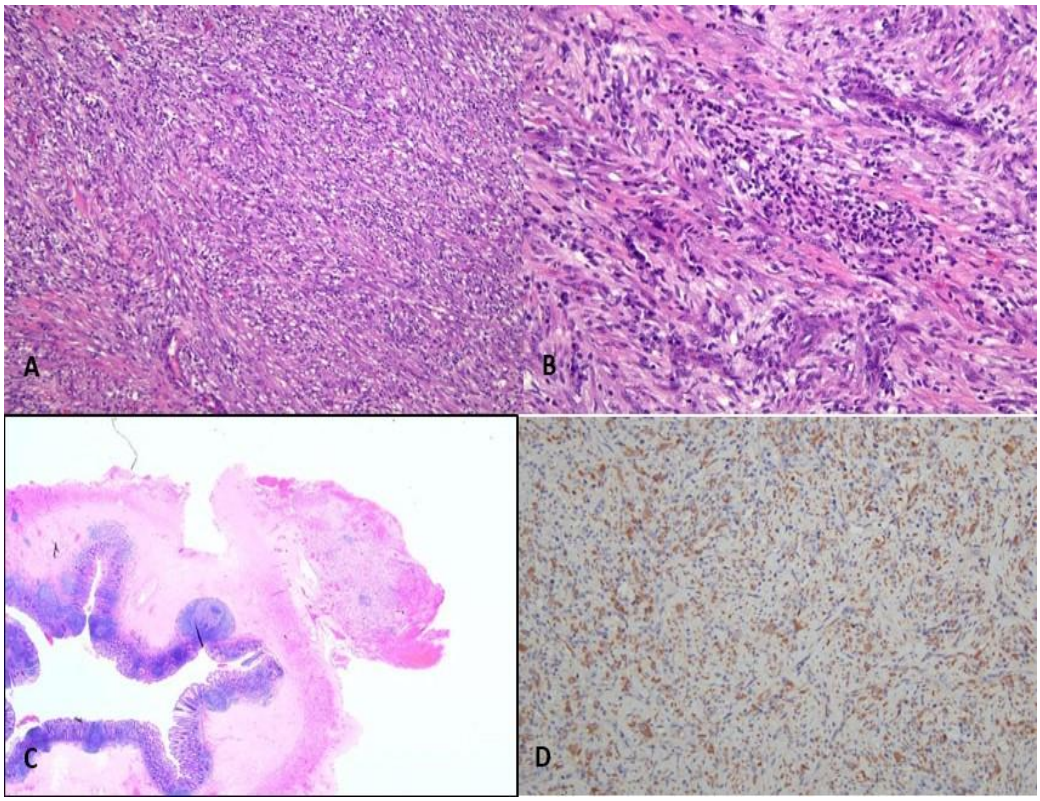

FIGURE 4: (A) Diffuse sheets of neoplastic spindle-shaped cells embedded within collagenous stroma; (B) The stroma was moderately infiltrated by lymphocytes, plasma cells and occasional eosinophils; (C) A few small fragment of tumour tissue was seen adhered to the serosal layer but not infiltrating into the muscularis propria of the adjacent colon; (D) Tumour cells showed positive staining for Anaplastic Lymphoma Kinsase (ALK) by immunohistochemistry

Discussion

IMT is otherwise known as plasma cell granuloma, atypical myofibroblastic tumour, pseudosarcoma, atypical or atypical fibromyxoid tumour. It is a rare solid mesenchymal tumour, simulating a malignant neoplasms. This tumour was first described by Brunn in the year 1939. It was initially detected in the lungs. This neoplasem was classified by the World Health Organisation (WHO) as an 'intermediate' tumour between benign and malignant. However most tumours have a strong tendency for recurrence (1). Exact mechanism remains unknown, however there are a few triggering factors including infection, smoking, minor trauma or after minor surgery, leading to release of inflammatory mediators. Histologically, it is characterised by spindle cells, made up of myofibroblasts, embedded in myxoid stroma, with presence of other inflammatory cells including lymphocytes, plasma cells and histiocytes.

These tumours have a very low prevalence, ranging between 0.04 to 0.1% of the population. It can affect any age group, but mainly affects children and young adults. It is non familial and has no gender predilection. It can also affect any organs. The commonest organ involved would be the lungs (2,3) and intraabdominal especially mesentery and omentum (3). Rarely, it affects the liver, spleen, bladder and retroperitoneum. Amongst the paediatric age group, extrapulmonary IMTs show preference, with the mean age of ~10 years (3).

Clinical symptoms of patients with intraabdominal IMTs includes abdominal mass, associated with fever and abdominal pain (4). Fever is a resultant of inflammatory response. Other symptoms may include poor feeding, altered bowel habits due to bowel obstruction or intussuseption, or failure to thrive in children (1). Laboratory investigation is non-specific such as leukocytosis, anaemia, thrombocytosis or elevated erythrocytes sedimentation rate, which resolve soon after surgical resection (3). Specific tumour markers such as bHCG and AFP may be helpful to exclude other tumours and narrow down differential diagnosis.

Radiological features of IMTs are nonspecific, possibly because of the amount of fibrosis and cellular infiltration related to an inflammatory process. Hence, there is no definite radiological findings to suggest IMT. On ultrasound (5), these lesions have either an ill-defined or well-circumscribed borders and may be hypoechoic or hyperechoic in. Echogenic calcifications or fatty component have been sonographically observed. Some tumours also may show hypoechoic regions correlating with necrosis if large (6,7). On CT scan, these inflammatory tumours usually typically manifests as well defined bulky mass and usually appears hypodense to isodense with skeletal muscle, depending on the amount of collagenous stroma or myxoid content. Some cases have central calcification. On MRI images, these tumour appear as a hypointense signal intensity mass on both T1- and T2-weighted images with progressive enhancement on delayed post contrast imaging, which again reflects its fibrotic nature or component. Contrast enhanced CT and MRI may reveal a homogenous or heterogenous lesion. Rarely, some tumours may show circumferential growth around vessels and extension into the bowel with mural infiltration and ulceration suggesting aggressive growth patterns (6). Some of the possible differential diagnosis for IMT would include soft tissue sarcoma, lymphoma, metastatic disease or even a benign fibrous mesenteric tumour (7).

Histopathological examination (HPE) is usually gold standard in making a diagnosis. Proliferating spindle cells with presence of chronic inflammatory-cells like lymphocytes, neutrophils, eosinophils and macrophages were often described. Detection of anaplastic lymphoma kinase-1 (ALK-1) on immunohistochemistry staining is a useful indicator of chromosomal abnormality seen in IMT, which was detected in our case study. ALK immunoreactivity has been reported only in 36 to 60% of cases (2). Although the literature is mixed, some studies show ALK-positive cases have a better outcome than ALK-negative cases (1). This neoplasm also shows positive immunohistochemistry for SMA, vimentin, and calponin (2).

Studies have shown that the mainstay of treatment is usually complete resection, although surgery could be destructive to adjacent structures and increase morbidity (8,9,10). Other possible treatment options are adjuvant chemotherapy as well as radiation therapy which are usually options in cases of extensive local invasion, positive margins, or incomplete resection (6). Few studies have reported benefits of corticosteroid and non-steroidal inflammatory agents in treating invasive or incompletely resected tumours (9). Targeted therapy at the ALK oncogene has shown promising results (6). 15% to 37% of patients with mesenteric or retroperitoneal IMTs have been reported to develop recurrence. Hence, close follow-up, especially in the 1st year post-surgical resection, is recommended (5).

Conclusion

Even though IMT unusual, should be taken into consideration as one of the possible differentials if a

child presents with intraperitoneal mass. This tumour is often a challenge to diagnose. There is no definite imaging differentiation of an abdominal IMT from other malignancies due to the nature of the mass itself, hence HPE is still the gold standard for diagnosis. Familiarity with the imaging features of IMT and other solid tumours can help to narrow down differential diagnosis and avoid unnecessary radical surgery.

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