Case Report

Inflammatory Myofibroblastic Tumour in Children: A Report of Two Cases and Review of Literature

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Abstract

Inflammatory myofibroblastic tumour in paediatric patients present with a diagnostic dilemma because of its clinical, radiological and histopathological features overlapping with other mesenchymal tumours common in this age. Because of its rarity, the exact features are still unclear. Here, we are reporting clinical, radiological and histopathological appearances of two such cases. In both cases, the exact diagnosis was confirmed only after immunohistochemistry. There is a need for further detailed study to exactly determine the natural course and prognosis of these tumours.

Keywords: Immunohistochemistry, inflammatory myofibroblastic tumour, mesenchymal tumours, paediatric

Introduction

Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm of mesenchymal origin.[1] It is composed of inflammatory cells like lymphocytes and histiocytes; and mesenchymal spindle cells in variable proportions including fibroblasts and myofibroblasts. It can occur in variable anatomic locations. The most common sites are lung, omentum and mesentry.[2] Being rare, the exact features of this tumour are poorly understood. It is difficult to distinguish IMT from other mesenchymal tumours solely on the basis of radiological imaging. Histopathological analysis and immunohistochemistry (IHC) are required to establish the diagnosis.

We are reporting two cases, both female, who presented with complaints of an abdominal mass with associated symptoms like vomiting and pain abdomen. After an initial evaluation with radiological imaging and blood investigations, both were operated. The resected mass was reported as IMT in histopathological analysis and confirmed by IHC. Due to its rarity, the exact clinical, radiological and histopathological features are still confusing. We are adding features of two more cases to augment the limited available literature for this lesion.

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Case Reports

Case 1

A 9-month-old female child presented with swelling in the abdomen for 2 ½ months, loose stools for 2 months and vomiting after meals for 25 days. On general physical examination, patient had pallor, bilateral lower limb oedema with tachypnoea. On abdominal examination, there was a mass palpable just under the skin with fixity to the anterior abdominal wall, with a size of approximately 15 cm × 10 cm in vertical and horizontal dimensions occupying the right hypochondrium, right lumbar, right iliac and umbilical region. The mass was non-tender with mixed consistency and mobile in horizontal dimensions.

Routine investigations were normal except reduced haemoglobin level of 6.1 g/dl. Alpha fetoprotein and lactate dehydrogenase (LDH) were slightly raised and CA 19-9 and carcinoembryonic antigen were within normal limits.

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Contrast-enhanced computed tomography (CECT) abdomen was done in which there was a 11 cm × 10 cm × 7.5cm size heterogenous swelling on the right side epicenterd in the right lumbar region, almost reaching the inferior surface of the liver, displacing bowel loops, with no definite calcifications and fat attenuation, suggestive of neoplastic aetiology with mesenteric mesenchymal solid lesion [Figure 1a]. Ultrasound guide fine-needle aspiration cytology of the mass showed spindle cell lesion. Tru-cut biopsy revealed infantile fibromatosis with moderate to diffuse cytoplasmic positivity of Desmin in tumour cells. SMA, S-100, CD34, EMA and MyoD1 were negative in tumour cells. After initial stabilization, the patient was planned for excision of the mass. Intra-operatively, there was a huge solid multilobulated mass of size 12 cm × 12 cm, densely adherent to the rectus muscle anteriorly [Figure 2a]. After carefully dissecting the mass away from the abdominal wall, the tumour was further inspected. It was found to be adhered to terminal ileum, the tip of the appendix and caecum with adhesion to the surrounding mesentery [Figure 2b]. The tumour along with the involved bowel segments was resected and end to end ileo-colic anastomosis was performed in a single layer interrupted fashion with absorbable suture material. Post-operative period was uneventful and patient was discharged on post-operative day 6 after establishing oral feed.

Histopathological examination showed a large well-circumscribed spindle cell tumour arranged in fascicular pattern. The individual tumour cells were spindle shaped with blunt ends showing moderate cytological atypia, vesicular nulcei, conspicuous to prominent nucleoli and abundant eosinophilic cytoplasm. Tumour cells were mixed with inflammatory infiltrate. There was no focus of necrosis and no mitotic activity. Thus, there was confusion between infantile fibromatosis and IMT. On IHC, it showed anaplastic lymphoma kinase (ALK) positivity, confirming the diagnosis of IMT [Figure 3]. After 8 months of follow up, the patient is clinically asymptomatic and growing well.

Case 2

A 4 ½-year-old female child presented with complaints of a left abdominal lump and occasional fever for 1 year. It was not associated with vomiting or altered bowel and bladder habits. On examination, there was a globular, soft to firm non-tender mass of size 5 cm × cm 6 × 5 cm occupying left hypochondriac and lumbar region. It was mobile in horizontal dimensions. She was evaluated with radiological imaging and blood investigations. CECT abdomen showed a large well defined hyperenhancing intraperitoneal mass of size 6.22 cm × 5.92 cm × 7.05 cm with hypoenhancing areas within located in left lumbar region, probably arising from bowel loops [Figure 1b]. Blood investigations revealed serum ferritin level 393.0 ng/ml and a serum LDH value of 580.20 U/L. Her haemoglobin was 6.3 g/dl at admission for which she was transfused one unit of packed cells. Her 24 h urinary vanillylmandelic acid and homovanillic acid values were 10.20 and 4.43 mg/24 h, which were in the normal ranges. With these above findings, she had excision of mass. Intra-operatively, there was a smooth, almost globular, solid reddish mass of size 7 cm × 6 cm × 6 cm arising from the greater omentum located just over splenic flexure with adhesion to its serosa layer [Figure 2b]. The tumour was meticulously excised leaving behind an intact bowel with few serosal tears which were repaired with absorbable suture. Post-operative period was uneventful. She was discharged on full oral feed. The histopathological report was suggestive of benign spindle cell tumour with fibrocollagenous stroma infiltrated by lymphoplasmacytic infiltrate with differential diagnosis of IMT and gastrointestinal stromal tumour (GIST). IHC was reported as ALK positive, thereby confirming diagnosis of IMT.

Discussion

IMT is a rare lesion of unknown cause. It has been referred to with many synonyms like inflammatory pseudotumour, myxoid hamartoma, lymphoid hamartoma, inflammatory myfibrohistiocytic proliferation, etc., which reflects the variable biological nature of this tumour.[3] Although benign, IMTs are considered as locally aggressive and potentially malignant lesions with a rare risk of local recurrence and distant metastasis.[4] These lesions have been reported in all

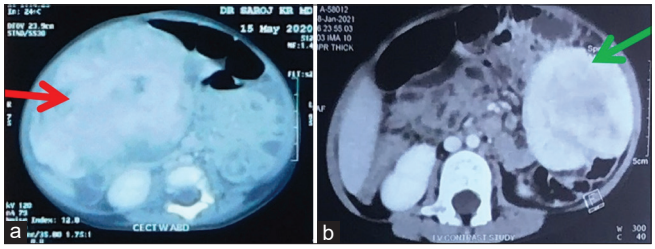


Figure 1: CECT abdomen showing- (a) 11 cm × 10 cm × 7.5 cm size heterogenous swelling on right side (Red arrow) epicentred in the right lumbar region, almost reaching the inferior surface of liver. (b) Solid hyperenhancing mass of size 6.22 cm × 5.92 cm × 7.05 cm (Green arrow) with hypoenhancing areas within located in left lumbar region in close relation to colon. CECT: Contrast enhanced computed tomography

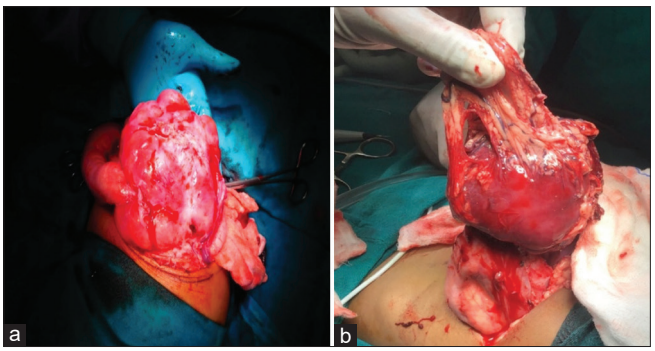


Figure 2: Intra-operative pictures showing - (a) Solid multilobulated mass of size 12 cm × 12 cm, densely adhered to rectus muscle anteriorly and mesentry of ileoceacal junction. (b) Solid, well defined, smooth, reddish mass covered with greater omentum, lying over splenic flexure of colon

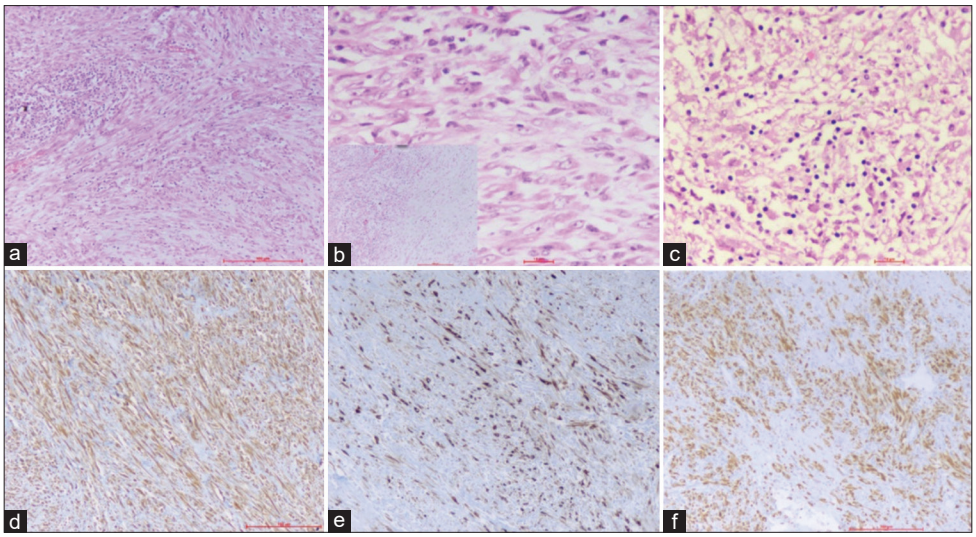


Figure 3: (a) Cellular areas of tumour composed of myofibroblastic spindle cells arranged in fascicles (H and E, ×100). (b) Myxoid hypocellular areas (inset, H and E, ×100) with oval to spindle cells having minimal nuclear atypia, inconspicuous nucleoli and scant cytoplasm (H and E, ×400). (c) Numerous admixed inflammatory cells chiefly lymphocytes (H and E, ×400). (d-f) Vimentin, desmin and anaplastic lymphoma kinase diffusely positive in tumour cells (Immunohistochemistry, ×100)

age groups, however the median age is around 10 years and occurrence in infancy is extremely rare.[5] Dalton *et al*. reported a series of 32 paediatric IMTs, out of which only two were in infants and the abdomen was the most common site (28%).[6] Abdominal IMT presents as abdominal mass with or without associated symptoms. Rarely, it may also present with features of acute abdomen, fever or weight loss.[7] In our series, the age of presentation is 8 months and 4 ½ years. The symptom at presentation was abdominal lump with vomiting in first case and occasional fever in second case.

There is no characteristic radiological appearance distinguishing IMT from other abdominal masses because of its rare occurrence.[8] The differential diagnosis of intra-abdominal solid mass arising from the mesentry include mesenteric fibromatosis, desmoplastic small round cell tumour, rhabdomyosarcoma, Burkitt lymphoma, IMT and Castleman disease. As there is dearth of literature on imaging findings of IMT, radiologists need to have high index of suspicion to recognize this lesion in paediatric age group.

Surgery is the main modality of treatment of IMT. Complete surgical excision is curative in most of the cases. Local recurrence has been reported in around one-fourth of cases and only <5% develop distant metastasis.[9] Surgical removal of lesions is determined by the location of the lesion where there is involvement of internal organs (viscera), symptomatic lesions, or lesions that present an immediate threat due to their location near vital organs and/or if they have shown progression.

These lesions do not have characteristic histological features. They contain a mixture of myofibroblasts and inflammatory cells with fibroblasts arranged in whorls or fascicles and variable proportion of spindle cells. Unfavourable factors include increased mitosis, high cellularity and proliferative histological pattern. Due to overlapping features with other mesenchymal tumours, IHC is mandatory to confirm the diagnosis. ALK gene rearrangements have been reported in majority of these tumours.[10] Tyrosine kinase inhibitors targeting ALK (crizotinib and ceritinib) has changed the treatment paradigm for ALK positive IMT. In our series too, both cases were positive for ALK.

Owing to its rarity and poorly defined histological features, exact diagnosis of IMT is difficult. In these two cases, we had diagnostic dilemma between infantile fibromatosis, IMT and GIST. Diagnosis was confirmed with IHC only, because radiological and histopathological characteristics of the mesenchymal tumours are almost similar. Both of our patients are doing well in follow up, but long-term follow-up to watch for the development of additional complications of these visceral lesions is warranted.

Conclusion

IMT is rare entity with overlapping features with other mesenchymal tumours. The prognosis for patients with IMT is difficult to predict because of the rarity of the disease. There should be high index of suspicion while encountering solid lesions in abdomen of paediatric age group. IHC is helpful tool to confirm the diagnosis and guide further management.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her

consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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