Case Report

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Mesenteric Tumor of an Infant: A Clinical and Pathological Surprise

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a tumefactive proliferation of spindled myofibroblastic cells admixed with inflammatory infiltrate. This tumor has a predilection for pulmonary location and also involves visceral soft tissues in children and adults. Tumors in extrapulmonary locations like abdomen, mesentery, bowel, head & neck, genitourinary and musculoskeletal are more aggressive locally and mimic malignancy. IMTs from colon are very rare and are treated by surgical excision. We report a case of a 9-month old female with IMT of sigmoid colon with no bowel complaints, treated by complete surgical excision with no further treatment needed.

Keywords: *Anaplastic Lymphoma Kinase-1 (ALK-1), Colon, Inflammatory Myofibroblastic Tumor, Mesentery*

Introduction

Inflammatory myofibroblastic tumor (IMT) is an infrequent solid, benign neoplasm of myofibroblastic origin and is grouped under fibro-inflammatory disorders which are composed of predominantly spindle cells along with a variable inflammatory component of lymphocytes and plasma cells. Although, individuals of any age may be affected, it has a preponderance for children and young adults [1]. The most common location is thoracic cavity [2], but there are rare reports of occurrence in extrapulmonary sites like mesentery of the small intestine, omentum, central nervous system, salivary glands, larynx, breasts, pancreas, spleen, liver and skin [1]. Clinical presentation of these masses vary according to the site of the tumor. Those arising from gastrointestinal tract may present with intestinal obstruction, anemia, blood in stools, vomiting. In around 15- 30% of cases, IMTs may be associated with systemic symptoms which include fever, weight loss, pain, anemia [3]. Pre-operative diagnosis is taxing as the clinical and radiological features impersonate a malignant neoplasm. These tumors have an intermediate malignant potential and have a tendency for local recurrence. Here, we present a rare case of sigmoid colon mesentery IMT presenting in infancy with a large abdominal mass and no other signs and symptoms of bowel pathology, managed by complete surgical excision.

Case History

A 9-month old female infant presented with abdominal distension since last 2 months. The mass gradually progressed in size. However, there was no history of abdominal pain, fever, vomiting, loss of appetite, weight loss, bowel bladder symptoms, or any signs of obstruction. On examination there was a large 10x10 cm mass occupying the central abdomen. The mass was non-tender, freely mobile with a smooth surface, firm consistency, regular margins and no free fluid. Laboratory studies showed hemoglobin 12.53 gm/dl, total leukocyte count 10,510 cells/mm^3, platelet count 470 x 10^9/L, AFP 5.36 ng/ml, HCG 0.17 mIU/ml.

Contrast-enhanced computerized tomography (CECT) abdomen showed a large well circumscribed tumor measuring 10.5 x 10 x 7 cm, arising in the mesentery with non-enhancing fluid attenuation and few enhancing areas and septations within it (Fig 1a). The lesion was noted in the central abdomen with internal characteristics, extensions, and mass effect and no invasion into the surrounding organs; possibility of lymphoma was suggested. Intraoperatively, a large lobulated mass was seen arising from the mesentery of sigmoid colon (Fig 1b). The tumor was excised along with a 5 cm segment of sigmoid colon adherent to it. There were multiple large lymphnodes in the mesentery of colon which were removed along with the tumor which were reactive on subsequent histopathological examination. On gross examination, the tumor was seen in colonic mesentery measuring 10.5 x 10 x 7.5 cm (Fig 1c). Mucosa overlying the mass was flattened out. Cut surface of the tumor was grey white firm with focal areas of haemorrhage.

Microscopically, the tumor was largely present in the mesenteric adipose tissue with focal infiltration into

the serosa of the colon (Fig 1d). Tumor was composed of mixed inflammatory cell infiltrate comprising of neutrophils, foamy histiocytes, lymphocytes, plasma cells, few eosinophils along with few scattered fibroblastic/ myofibroblastic spindle cells showing mild to moderate nuclear pleomorphism (Fig 2a). The stroma was loose oedematous and myxoid in areas (Fig 2b). No increase in mitosis (0-2/10HPF) or necrosis was seen. The neoplastic cells showed positivity for Vimentin and Desmin, confirming myofibroblastic phenotype (Fig 2c). ALK-1 positivity was observed in the tumor cells (Fig 2d). They were immunologically negative for CD117, CD34, S-100 and LCA. Ki67 proliferative index was low. A diagnosis of IMT was rendered. The patient at her most recent follow up (of one and half months) showed no signs of recurrence.

Discussion

IMT is also called as inflammatory pseudotumour, plasma cell granuloma, myofibroblastoma, and inflammatory myofibroblastic sarcoma. There is lack of agreement among experts about nature of IMTs, whether they are reactive or neoplastic. Comprehensive studies on this tumor have proven it to be a neoplasm as the majority of them showed genetic fusion of ALK gene proposing its role in the pathogenesis of IMT [4,5]. IMTs are categorized into intermediate grade neoplasms according to World Health Organization (WHO) classification [4]. Three histological patterns have been described by Coffin et al in their study of 59 cases, which includes myxoid/ vascular, compact spindle cell and fibromatosis-like patterns [6]. The myxoid/ vascular pattern has an appearance similar to fasciitis and is composed of loosely arranged plump spindle cells in an oedematous/ myxoid stroma with prominent blood vessels. The inflammatory infiltrate is richer in neutrophils and eosinophils with relatively sparse plasma cells than the other two patterns. Second pattern showed compact spindle cells with storiform architecture in a collagenous stromal background. These are typically rich in plasma cells and lymphocytes admixed with the spindle cells. The fibromatosis-like pattern is hypocellular, with elongated spindle cells admixed with scattered lymphocytes, plasma cells and eosinophils in a dense collagenous stroma. According to a study by Coffin et al, a large number (72.9%) of IMTs display atypical histologic features including increased cellularity, cellular atypia and presence of multinucleated/ anaplastic giant cells, atypical mitosis and necrosis. The remaining tumors show bland appearance on histology, hence, called classical IMT [6]. The present case had histological features similar to first type and had loose oedematous stroma, rich inflammatory infiltrate and sparse fibroblasts. Based on histology and/or IHC results, other solid mesenteric neoplasms like mesenteric fibromatosis,

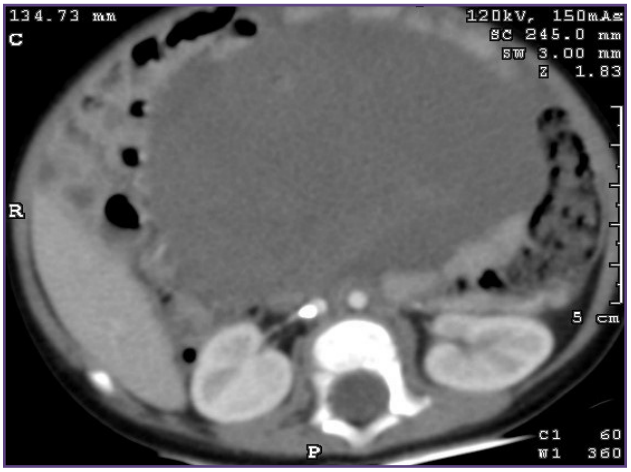

Fig. 1a: CECT image (Axial view) showing a large well circumscribed tumor arising in the mesentery with non-enhancing fluid attenuation and few enhancing areas and septations within it.

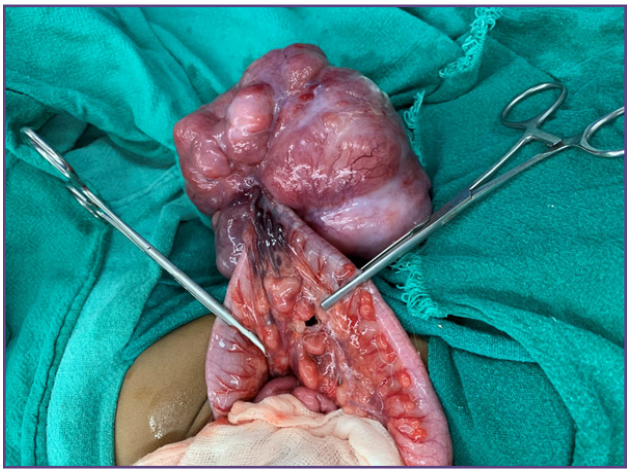

Fig. 1b: Intraoperative image showing a large lobulated mass arising from the mesentery of sigmoid colon.

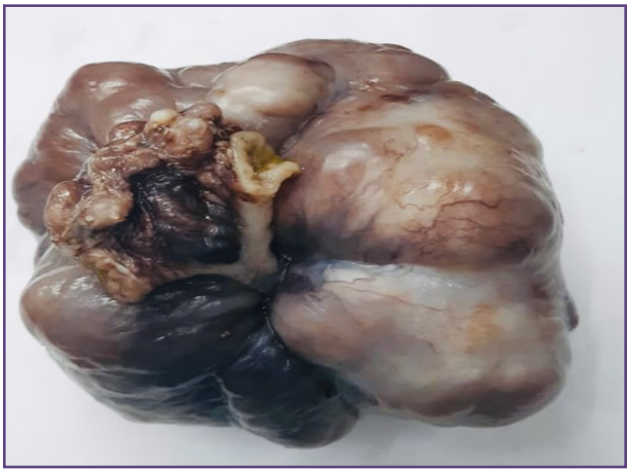

Fig 1c: Gross image showing a lobulated tumor with an adherent segment of sigmoid colon.

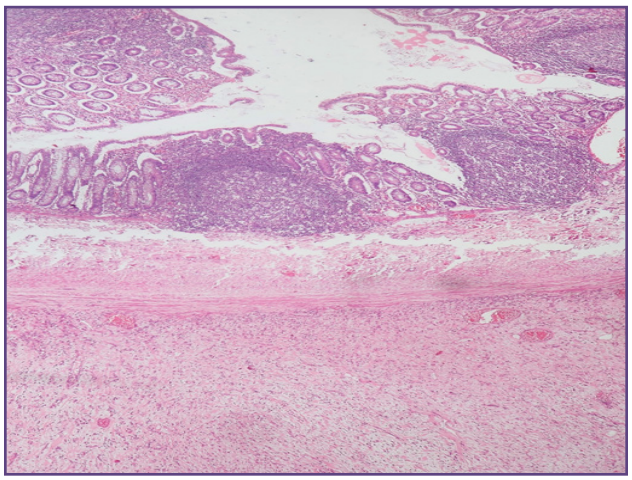

Fig. 1d: Tumor largely present in the mesenteric adipose tissue with focal infiltration into the serosa of the colon (H&E 40X).

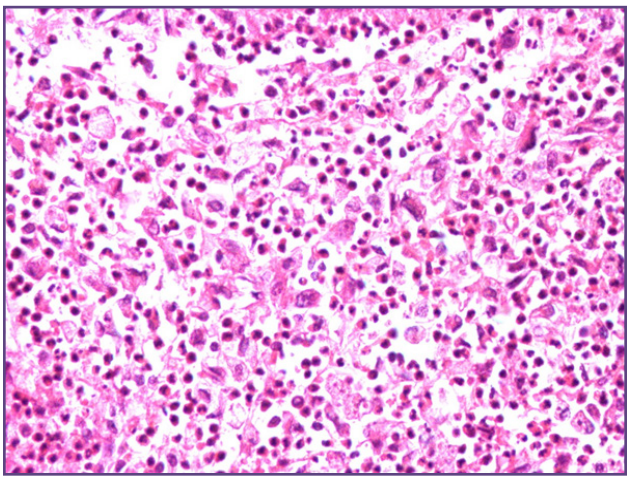

Fig. 2a: Tumor composed of neutrophils, foamy histiocytes, lymphocytes, plasma cells, few eosinophils along with few scattered fibroblastic/ myofibroblastic spindle cells (H&E 100X)

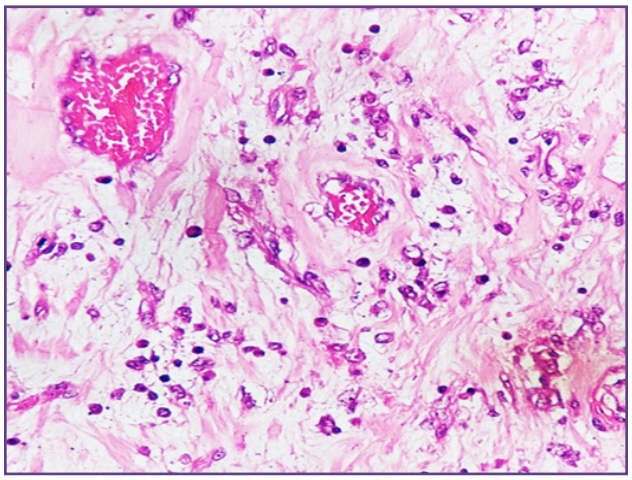

Fig. 2b: Tumor stroma loose oedematous and myxoid in areas (H&E 200X).

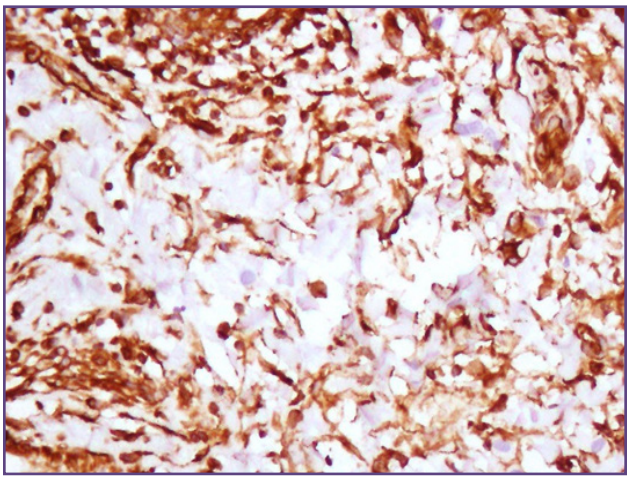

Fig. 2c: IHC staining- Vimentin positivity (200x).

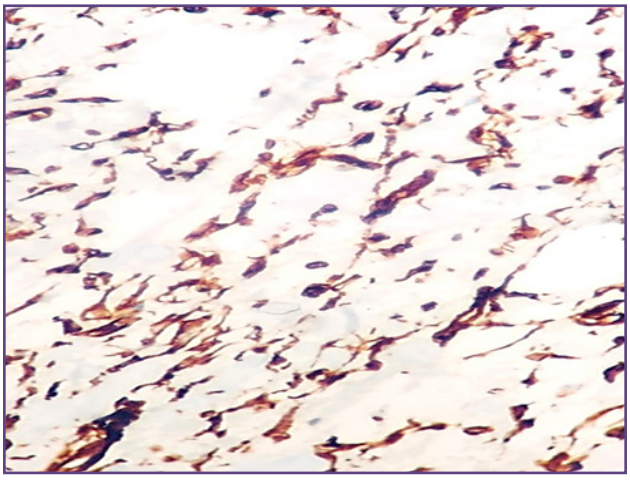

Fig. 2d: IHC staining- ALK-1 positivity (200x)

desmoplastic small round cell tumor, rhabdomyosarcoma or extraskeletal Ewing sarcoma, Burkitt lymphoma were ruled out.

This case was unique because of multiple unusual clinical presentations; one was age of the patient which was nine months as compared to median age of 10 years in the available literature [7]. Secondly, despite the colonic involvement, the patient had no symptoms related to bowel obstruction. Children with colonic IMTs, mostly have associated anemia but the present case had normal hemoglobin levels. These atypical clinical presentations pose a challenge in the diagnosis and management of these tumors.

At present, there are no definite histopathologic, molecular, or cytogenetic features to predict the risk of malignant transformation, recurrence, or metastasis [6]. The mainstay of management of IMT is surgical excision. Incomplete tumour removal frequently results in local recurrence. These tumors have shown regression in response to corticosteroids and nonsteroidal anti-inflammatory drugs [8]. The patient did not receive any other therapy post-surgery, and she is doing well after one and half months of follow-up. No evidence of recurrence or metastasis has been found yet. Alaggio et al in their case series showed a local recurrence rate of 23%, with a 5-year survival rates of 87.4% [9]. A novel ALK targeted inhibitor, crizotinib has been found to be effective in treatment of unresectable and metastatic ALK-expressing IMT [10].

Conclusion

Despite the presence of advanced imaging procedures and laboratory techniques, these lesions are often confused with other neoplastic entities, because of their nonspecific clinical and morphological findings, hence making the diagnosis difficult. The pathological examination remains the essential tool for accurate diagnosis of IMT.

Funding

None

Competing Interests

None

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