SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Pediatric General Surgery

Mesenteric Desmoid-Type Fibromatosis Presenting as Obstructing Mesenteric Bands: A Rare Case

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DOI: https://doi.org/10.36347/sasjs.2024.v10i08.007 | **Received:** 30.06.2024 | **Accepted:** 05.08.2024 | **Published:** 09.08.2024

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Abstract **Case Report**

Mesenteric Fibromatosis (MF) is a proliferative fibroblastic lesion of the small intestinal mesentery. It is a rare type of desmoid tumour. It can occur sporadically or in association with familial adenomatous polyposis (FAP). It is generally seen in females in reproductive age group and is a locally aggressive benign tumour. Patients present with abdominal mass or other abdominal symptoms such intestinal obstruction, intestinal perforations, ureteral obstruction, etc. We describe a case of a 7 year old girl who presented with intestinal obstruction due to distal ileal mesenteric bands. Exploratory laparotomy and adhesiolysis of mesenteric bands was done and the excised band was sent for histopathology, which suggested mesenteric desmoid-type fibromatosis. As there is a chance of local recurrence, patient has been kept under regular follow up with ultrasound of abdomen and for any abdominal symptoms. According to our literature search, this is the only reported case of such a presentation of mesenteric fibromatosis presenting as localised mesenteric bands without any mesenteric or retroperitoneal abdominal mass in a child.

Keywords: Mesenteric Fibromatosis, Mesenteric Bands, Intestinal Obstruction, Exploratory Laparotomy.

BACKGROUND

Mesenteric fibromatosis is a proliferative fibroblastic lesion involving the small intestinal mesentery. It is a type of desmoid tumour and constitutes 8% of all desmoid tumours. Desmoid tumours themselves are a rare entity, forming 0.03% of all neoplasms [1]. Most of the patients present with symptoms such as abdominal pain or mass, intestinal obstruction or ureteral obstruction, intestinal perforation or fistulas. Diagnosis is done on histopathology and treatment involves wide local excision of mass. Even with radical complete excision, there is a 25-50% chance of recurrence [2].

CASE REPORT

A 7 year old girl was admitted to our hospital with multiple episodes of non-bilious vomiting. A radiograph of the abdomen showed multiple air-fluid levels with bowel gas in distal colon. Patient had passed stools the same day. On examination, there was mild abdominal distension, and mild tenderness. A Ryle's tube was inserted which drained minimal quantity of light green aspirates. It was decided to manage patient conservatively with enemas initially and try to relieve the obstruction. Twenty four hours after admission, patient stopped passing stools and her abdominal distension increased. The Ryle's tube also drained 100 ml dark green bile. It was decided to take up the patient for exploratory laparotomy.

On exploration, patient had dilated ileal loops upto distal ileum and collapsed terminal ileal loops and colon. There were adhesive bands present in the distal ileal mesentery compressing the lumen of the ileum. There was another adhesive band 5cm proximal partially compressing the ileal lumen. The adhesive bands were excised and the obstruction was relieved. The excised band was sent for histopathology.

The histopathology report revealed a non-encapsulated lesion, irregular lesion composed of bland, wavy spindle cells with pale cytoplasm arranged in fascicles. Thin walled blood vessels with perivascular stromal edema and areas of myxoid change were also noted. These changes are suggestive of "mesenteric desmoid-type fibromatosis".

Post-operative period was uneventful with patient started on liquids on post-operative day 3 and gradually progressing to full diet on day 5.

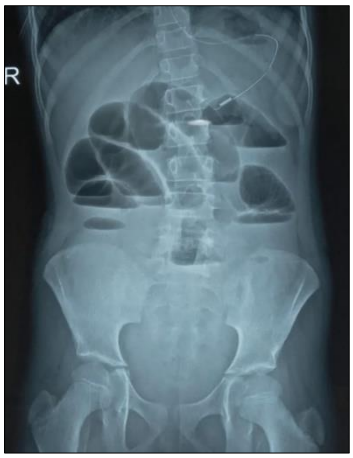

Fig. 1: Plain radiograph of abdomen showing air-fluid levels

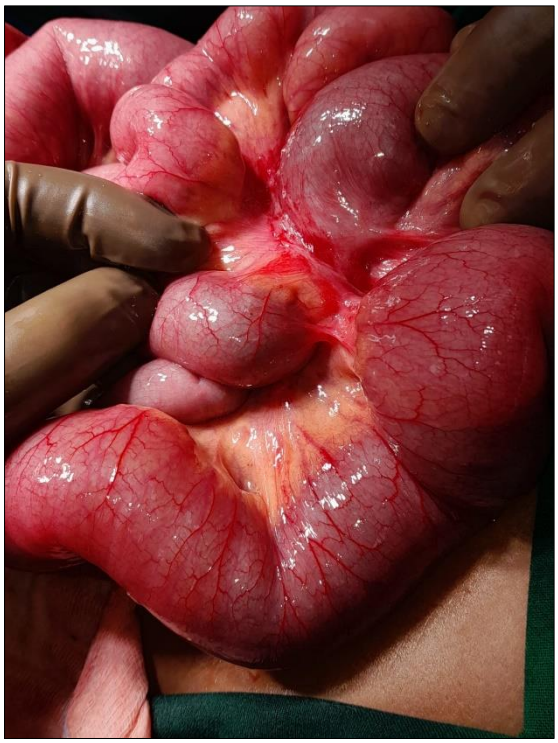

Fig. 2: Obstructing mesenteric bands

DISCUSSION

Desmoid fibromatosis or desmoid tumour (DT), also known as deep fibromatosis or aggressive fibromatosis, is a rare disease. It has two relative incidence peaks reported in literature, one at 6 to 15 years of age and the other between puberty and age of 40 years [3].

Desmoid tumours are classified into 3 types: abdominal, extra-abdominal and intra-abdominal [2]. Intra-abdominal desmoid tumour can occur either in the mesentery or in the retroperitoneum [4]. It can occur either sporadically or in association with familial adenomatous polyposis (FAP) (Gardner's syndrome) with an association of 10-18% [1-4]. Mesenteric fibromatosis (MF) represents 8% of all desmoid tumours with the commonest location being ileal mesentery, followed by mesocolon and gstro-hepatic ligament [5].

Aetiology can include trisomy of chromosome 8 or 20, hormonal stimulation, trauma or previous abdominal surgery[6]. Presentation may vary from asymptomatic abdominal mass to intestinal obstruction or perforation to impairment of functioning of ileo-anal anastomosis in operated cases of FAP. Diagnosis can only be confirmed on histopathology.

Our patient presented with intestinal obstruction without any palpable mass. On exploratory laparotomy she was found to have obstructing ileal mesenteric bands which were excised and sent for histopathology. The diagnosis of mesenteric fibromatosis was confirmed on histopathology.

Treatment remains complete wide excision of lesion to prevent recurrence. Efficacy of radiotherapy and chemotherapy is unpredictable. Recurrence rates are higher for mesenteric or retroperitoneal DT than for abdominal wall DT [7]. Even with complete local excision, recurrence rates can be high in some cases.

Our patient is currently asymptomatic 6 months after surgery. We are clinically following up the patient for any abdominal symptoms or abdominal mass. We have planned a magnetic resonance imaging (MRI) study of abdomen at 1 year follow up.

Conflict of Interest: None to declare

Sources of Funding: None

Acknowledgements

Dr. Tejas Kulkarni, Consultant Pathologist, MRR Children's Hospital

Dr. Praveen Gokhale, Director of Critical Care & Consultant Pediatrician, MRR Children's Hospital

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