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Case Report

Mesenteric fibromatosis involving intestine mimicking a gastrointestinal stromal tumor

Sakshi J. Parmar*, Manmohan M. Kamat, Avani Mishra, Saisree Setipalli

Department of General Surgery, Nanavati Max Super Speciality Hospital, Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Sakshi J. Parmar,
E-mail: sakshi.parmar0706@gmail.com

ABSTRACT

Mesenteric fibromatosis (MF) or intra-abdominal desmoid tumour is a rare proliferative disease affecting the mesentery. MF is a locally aggressive tumour that lacks metastatic potential, but the local recurrence is common. We report a case of 33-year-old female with chief complaints of mass in abdomen which was hard on palpation arising from the pelvis and extending up to the umbilicus. Magnetic resonance imaging (MRI) suggestive of soft tissue mass and exploration was planned. Intra-operatively a mass of 16×16×8 cm with attached ileum and caecum which was excised completely and sent for histopathological examination. Histopathology was suggestive of Mesenteric fibromatosis spindle cell type. Considering the rarity of this tumour and difficulties in diagnostic and therapeutic ambit, we believe it is justified to publish this case which came to our observation.

Keywords: Mesenteric fibromatosis, Intestinal involvement, GIST, Desmoid tumour

INTRODUCTION

Mesenteric fibromatosis (MF) is a cytologically bland, at most moderately cellular, deep infiltrative fibro-proliferative process primarily involving mesentery or other intra-abdominal sites.[1] This tumour may remain asymptomatic in the early period and usually manifest as a painless mass. Because of the tumour's characteristic of local infiltration of adjacent organs, symptoms caused by compression can occur as the tumour grows. At present, the precise aetiology of this tumour is still undefined, and surgical resection remains the appropriate treatment for intra-abdominal fibromatosis in most cases. A giant MF with ileocecal involvement has been rarely reported in the literature.

CASE REPORT

A case of 33-year-old female homemaker by profession came walking in outpatient department with chief complaints of Mass in abdomen since six months which was gradually increasing in size associated with feeling of heaviness in abdomen since one month. Menstrual history/past history/personal history and family history was not contributory. On local examination hard globular mass was felt per abdomen arising from the pelvis extending up to umbilicus. Magnetic resonance imaging (MRI) s/o well circumscribed soft tissue mass arising from pelvic region approximately 16×16×7 cm (Figure 1).

Differential diagnosis of Uterine fibroid and Gastrointestinal stromal tumour were considered and patient was taken up for exploratory laparotomy. Intra-operatively a globular intra-peritoneal mass measuring 16×16×8 cm arising from ileo-caecal junction was noticed with attached distal ileum and caecum while uterus appeared to be normal (Figure 2).

The mass was resected completely with 5 cm margin on either end and ileo-colic anastomosis was done in 2 layers. The postoperative course was uneventful and the patient was discharged in a good health condition on 5th post-operative day.

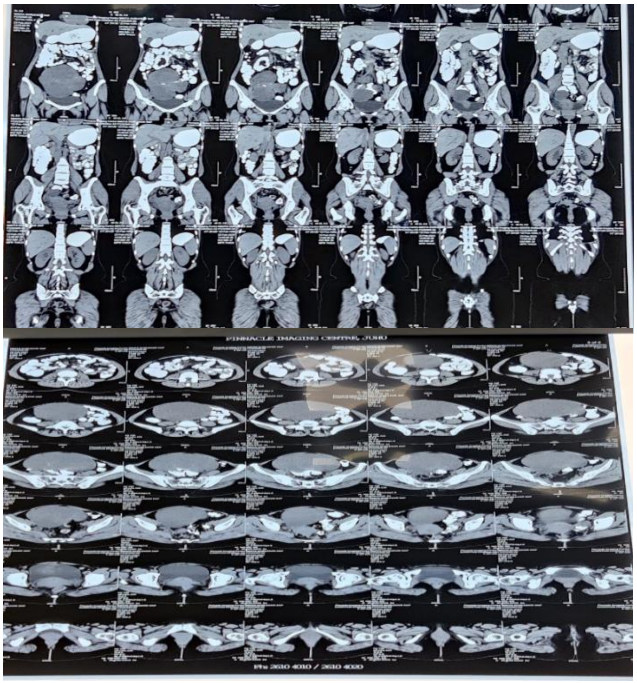

Figure 1: MRI showing soft tissue mass arising from the pelvic cavity.

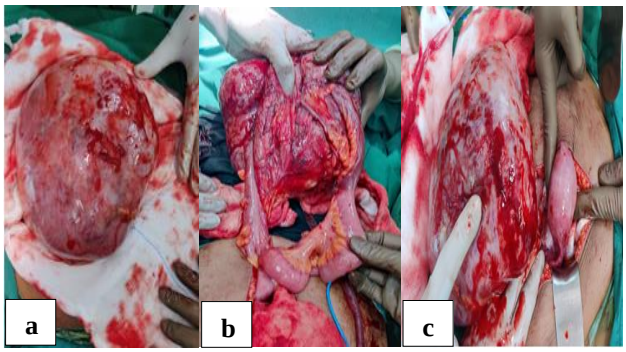

Figure 2: Intra-operative images (a) a mass of 16×16×8 cm, (b) with attached distal ileum and cecum, and (c) while uterus appears to be normal.

Histopathology

Gross examination

A capsulated mass measuring 17×17×8.5 cm with attached segment of small intestine and cecum measuring 18 cm in length and appendix stretched Over the mass, measuring 10cm in length. Cut surface of the mass shows homogenous greyish white soft fibrous areas with an area adherent to part of small intestine. No cystic areas noted. No evidence of necrosis grossly (Figure 3).

Microscopic examination

Sections show a circumscribed tumour composed of elongated spindle wavy cells in a collagenous to myxoid stroma. The cells show minimal atypia, pale eosinophilic and mitotic activity. Tumour is seen infiltrating muscularis layer of small intestine and infiltrating adjacent adipose tissue. The cells are positive for desmin, vimentin, calretinin and weakly for PR. Catenin shows focal nuclear positivity suggestive of mesentric fibromatosis – spindle cell type (Figure 4).

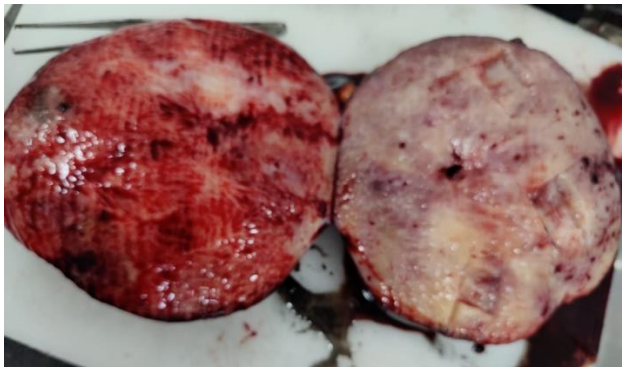

Figure 3: Cut section.

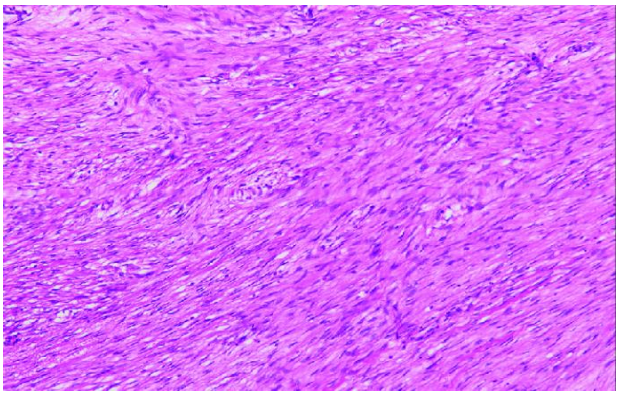

Figure 4: Microscopic appearance showing spindle cell type mesenteric fibromatosis.[5]

A medical oncology opinion was taken in view of starting chemotherapy/radiotherapy to avoid recurrence. She was advised to follow up 3 monthly with a computed tomography (CT) scan.

DISCUSSION

MF or intra-abdominal desmoid tumour is a rare proliferative disease affecting the mesentery. MF is a locally aggressive tumour that lacks metastatic potential, but the local recurrence is common. It resembles gastrointestinal stromal tumours (GIST) that are mesenchymal neoplasms of the digestive tract and show a varied malignant potential. Although GISTs and mesenteric fibromatosis are distinct entities, they are often confused clinically, radiologically and not uncommonly pathologically as well. Differential diagnosis of mesenteric desmoid-type fibromatosis mainly includes GIST, leiomyoma, leiomyosarcoma, solitary fibrous tumour, and neurofibroma. All these tumours may variably exhibit myxoid stromal changes. GIST differs from desmoid-type fibromatosis in that it is stained for CD34

and CD117 stains, not usually expressed in desmoid-type fibromatosis. Misdiagnosis might result in inappropriate therapeutic decisions and worse prognosis. Differentiating features of mesenteric fibromatosis and gastrointestinal stromal tumour are tabulated in Table 1.[1]

Table 1: Differentiating features of mesenteric fibromatosis and gastrointestinal stromal tumour.[1]

Mesenteric fibromatosis	Gastrointestinal stromal tumour
CD34 negative	CD34 60-70% positive
CD117 frequently negative, variable reports of focal/weak staining	CD117 74-95% positive
DOG1 negative	DOG1 87-94%
Beta-catenin positive 90% (nuclear)	Beta-catenin negative
Low to moderate cellularity	Moderate to high cellularity
Cytologically bland	May be cytologically atypical
Prominent thin-walled dilated veins	Lacks prominent veins
Infiltrative margin	Usually circumscribed, pushing margin
No cystic degeneration or necrosis	May have cystic degeneration or necrosis

Its pathogenesis is not completely understood, even if some cases have been associated with antecedent abdominal trauma, including previous surgery (25%), genetic disorders like Gardner's syndrome and familial adenomatous polyposis, infectious aetiology (human herpes virus), and autoimmune diseases (Crohn's diseases) or hyper-estrogenic states.[3] Hormonal estrogenic disorders may be involved in the pathogenesis, and this could explain the high incidence of this disease in females (female-male ratio 3: 1), frequently in young pregnant or postpartum women with an age ranging from 25 to 35 years.[2]

Some researchers have suggested that the risk of local recurrence still remained, and ranged from 25 to 50% in most studies of complete surgical resections. The surgical operation itself is a kind of trauma that may lead to further recurrence. In addition, the unpredictable nature of these tumours and the factors associated with recurrence are inconsistent. Dalén et al reviewed 5 of 8 patients with intro-abdominal desmoid tumours who received no treatment, and they concluded that some types of desmoid tumours with few or no symptoms may spontaneously decrease or disappear, which has led the researchers to assess the role of a wait-and-see policy.[4]

Radiotherapy, systemic therapy (including tamoxifen, non-steroidal anti-inflammatory drugs, and chemotherapy), molecular-targeted therapy, ultrasound-guided high-intensity focused ultrasound (HIFU) ablation may be valuable options in treating MF for patients who are unable to undergo surgery, positive histological margins after surgical resection, postoperative recurrence, and surgery may lead to unacceptable damage of the function of vital organs.[4] In our case, we performed complete surgical resection of the tumour, and we did not prescribe further adjuvant treatment, except routine follow-up.

CONCLUSION

The diagnosis of mesenteric fibromatosis should always be considered in the case of mesenchymal tumours apparently originating from the bowel wall that diffusely infiltrate the mesentery. The final diagnosis depends on the pathology, which remains the gold standard for diagnosis. Differentiating between the two types of tumours is important because while MF tumours are usually benign, GISTS have malignant potential.

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