

RARE CASE REPORT OF MESENTERIC FIBROMATOSIS

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Mesenteric fibromatosis is a part of the clinical-pathologic spectrum of deep fibromatoses. We report this rare case of primary mesenteric tumor that was diagnosed to be a mesenteric fibromatosis on histopathological examination.

In majority of patients it may remain asymptomatic and the management of these tumors depends on histopathological examination. Postoperatively, patient was well and subsequent follow up showed normal recovery.

Key words: mesenteric fibromatosis, abdominal tumor, surgery

Mesenteric fibromatosis is a part of the clinical-pathologic spectrum of deep fibromatoses. The deep fibromatoses encompass a group of benign fibroproliferative processes that are locally aggressive and have the capacity to infiltrate or recur but not metastasize.

The deep fibromatoses are classified by anatomic location because they may arise from intraabdominal sites (mesenteric, pelvic, and retroperitoneal fibromatosis), the deep soft tissues of the abdominal wall (abdominal fibromatosis), and deep within extraabdominal soft tissues (extraabdominal fibromatosis).

Mesenteric fibromatosis occurs in a wide age range of patients, 14–75 years of age (mean, 41 years), and has no gender or race predilection.

Thirteen percent of patients with mesenteric fibromatosis have familial adenomatous polyposis (FAP), specifically, the Gardner syndrome variant of FAP. The small bowel mesentery is the most common site of origin of intraabdominal fibromatosis.

We report this rare case of primary mesenteric tumor that was diagnosed to be a mesenteric fibromatosis on histopathological examination.

CASE REPORT

A 59-year-old male came to the surgery opd with abdomen since 1 month. On examination, his vitals were normal. On per abdomen examination, there was a solitary swelling measuring 15 x 12 cm, spherical, non tender, freely mobile with a band of resonance heard over the mass. Aprobable diagnosis of mesenteric cyst was made.

Investigations:

- FNAC: malignant mesothelioma, well-differentiated adenocarcinoma;
- ultrasound abdomen and pelvis: there is a large hyperechoic mass lesion measuring 11.5 x 14.9 x 16.5 cm arising from the Right Iliac Fossa (likely Caecum). There is increased vascularity within the lesion – likely GIST.
- NCCT + CECT abdomen: a well defined, large (measuring 15.6 x 11.3 x 17.5 cm) minimally enhancing, homogenous, solid mass lesion noted in the mesentery with adjacent distal ileal bowel wall thickening (6 mm), mesenteric fat stranding inferiorly, abutting and displacing the adjacent small bowel loops. The lesion is seen in the mid-line superiorly extending upto origin of

SMA, inferiorly up to common iliac bifurcation, anteriorly abutting the anterior abdominal wall. The lesion is supplied by branches of SMA that are seen transversing through out the lesion.

Imp: Large, mesenteric mass lesion as described with adjacent ileal wall thickening – suggestive of GIST, likely to be arising from distal ileum.

Patient underwent Exploratory Laparotomy with Intestinal resection and anastomosis under GA on 13/03/2014. A large, solitary mass was present at the root of the mesentery and adherent to recto sigmoid colon and small intestine. Specimen weighed 2.4 kgs and was sent for HPE.

Operative findings



Fig. 1. Large abdominal mass present at the root of mesentery



Fig. 2. Mass adherent to recto sigmoid colon and the small intestine



Fig. 3. Specimen resected



Fig. 4. Anastomosis and resection done

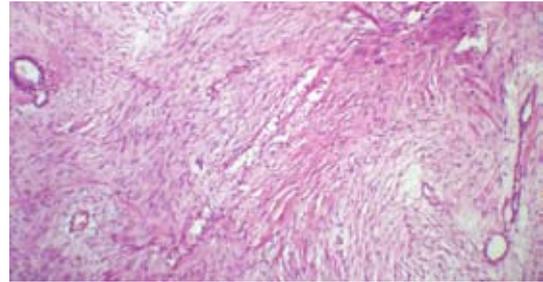


Fig. 5. Mesenteric fibromatosis showing cytologically spindle cells in a collagenous stroma

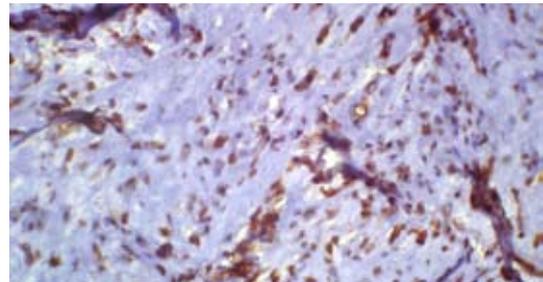


Fig. 6. β -catenin immunohistochemistry separates mesenteric fibromatosis from GIST

DISCUSSION

Mesenteric fibromatosis is a type of fibroblastic proliferation affecting the mesentery that develops usually as a consequence of surgical trauma, but it may occur spontaneously. It is sufficiently rare for no clear data of incidence and presented features to be known. The majority of patients with mesenteric fibromatosis remain clinically asymptomatic, with little or no focal symptoms until later in their course, at which stage they complain of abdominal pain and discomfort, constipation, vomiting, and organ compression symptoms, such as small bowel obstruction and hydronephrosis.

The etiology is unknown but an endocrine cause is suggested by: (i) the relative prevalence in perinatal women; (ii) tumour regression after the menopause; and (iii) regression with tamoxifen therapy (1).

As there is no classical symptomatology related to mesenteric fibromatosis, the diagnosis is confirmed only after the histological analysis of the tumor. Desmoidtumours are firm masses. On cross section these tumours are grayish and grossly homogenous.

Microscopically, mesenteric fibromatosis is characterized by a spatially homogenous proliferation of wavy spindle cells without atypia, associated with collagen among dilated vessels (2). The mitotic count is relatively low with no evidence of necrosis and nuclear dedifferentiation.

The diagnosis of mesenteric fibromatosis is based on the microscopic examination and immunostaining. It is noteworthy that a CD117 antigen, expressed commonly in GISTs, can be positive in up to 75% cases of mesenteric fibromatosis. In contrast to GISTs, MF does not express CD34 and S100 protein. Recently, the expression of beta-catenin was revealed in fibromatoses that might prove helpful in the differential diagnosis in doubtful cases (3).

The sonographic features of mesenteric fibromatosis are nonspecific and chiefly dependent on collagen and fibroblast content and intralesional vascularity of the tumor. The results of the treatment might be biased due to the unpredictable course of this disease with

some tumours regressing or remaining stable without any treatment.

The management of desmoids should be individualized and multimodal. The surgical resection should be performed only in localized tumours that do not invade the root of the mesentery. Intra-abdominal desmoids can be resected in 53–67% of cases. Fibromatoses are locally aggressive tumours that tend to recur when incompletely resected (4, 5).

A high rate of recurrence after the surgical resection results from the incomplete resection, multicentric disease or surgical trauma as a new precipitating factor.

Radiotherapy may be used before surgery in cases of recurrence and inoperable lesions to shrink the tumor and make it operable. Adjuvant radiation therapy reduces recurrence of mesenteric fibromatosis to 20-40%, compared to 40-70% with resection only (5).

Recently, a successful therapy of a desmoids tumour resistant to traditional chemotherapeutic regimens was reported with imatinib, a tyrosine kinase inhibitor that is successfully used in advanced gastrointestinal stromal tumours.

CONCLUSION

Mesenteric fibromatosis is a rare tumour that in majority of patients may remain asymptomatic and the management of these tumors depends on histopathological examination.

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