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Case Report

AGGRESSIVE MESENTRIC FIBROMATOSIS: A RARE CASE REPORT AND REVIEW OF LITERATURE.

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Abstract :

Mesentric fibromatosis is a proliferative fibroblastic neoplasm of the small intestinal mesentery with varied clinical presentation. Giant mesentric fibromatosis is uncommon and its rarity poses a diagnostic and therapeutic challenge. This paper presents a recurrent aggressive fibromatosis in a 38 year old male patient, who had initially undergone a laparotomy outside for mass abdomen but only pus was evacuated and definitive diagnosis was not made.

Keywords : Mesenteric fibromatosis, mass abdomen, abdominal abscess, benign neoplasms.

Introduction :

Fibromatosis are heterogenous group of condition which includes mesentric fibromatosis. Mesentric fibromatosis are benign proliferation of mesentric tissue that tends to recur without distant metastasis[1]. These tumors are histologically benign but they behave like malignant tumors. They might invade locally and recur after excision[2]. Giant mesentric fibromatosis are uncommon by itself (2-4 cases/million/year). Some cases may be associated with other pathologies like, association with Gardener's syndrome (3- 45%), and Familial colonic polyposis (10%)[3]. Most reported cases have been associated with Gardener's syndrome, previous trauma and prolonged estrogen intake[4]. However primary mesentric fibromatosis, without any predisposing factors or association is also known. Patients presents to hospital with mass effects or obstruction of surrounding structures like small intestine and ureters[5].

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This report presents a case of aggressive mesentric fibromatosis in a young male who presented with history of pain abdomen, abdominal distension and previous history of laparotomy, which was managed in Department of surgery in K.S.Hegde hospital, Mangalore, India.

Case Report :

A 38 year old male presented with history of abdominal pain and distension of abdomen since 2 months. Patient also had history of previous laparotomy 1 year back for mass abdomen. Operative notes showed, intra abdominal abscess adjacent to the splenic flexure of colon and root of the mesentery without any other abnormality. Pus was evacuated and definitive histopathology was not possible then. Now, on examination patient had large mobile mass per abdomen, occupying most of the abdomen and a large laparotomy scar was also seen. Laboratory investigations were within normal limits. CT scan showed large mass lesion between anterior abdominal wall and mesentery measuring 11.8 X 11.3 X 12.9 cm which was surrounding by bowel loops (Fig 1,2). USG guided FNAC was inconclusive. With clinical diagnosis of GIST, patient underwent laparotomy. Intra operatively, large mass at the root of jejunal mesentery with loop of jejunum adherent to it was seen, along with small bowel adhesion due to previous surgery. Patient underwent resection of the tumor along with resection of 90 cm of jejunum and proximal ileum with end to end anastamosis (Fig 3). Patient recovered well without any post operative complications. Histopathology

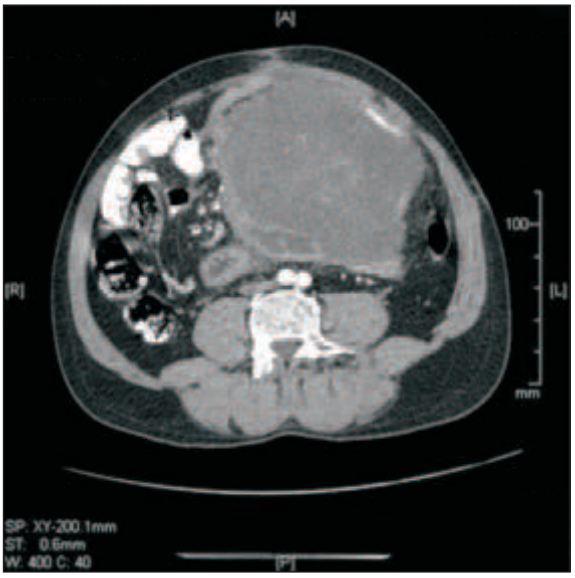

Fig 1 : CT scan picture showing large mass lesion in the mesentery surrounded by small bowel loops.

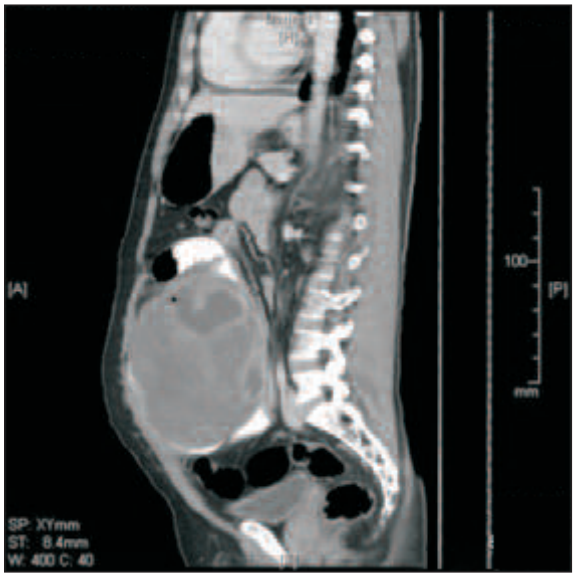

Fig 2 : CT scan sagital view.

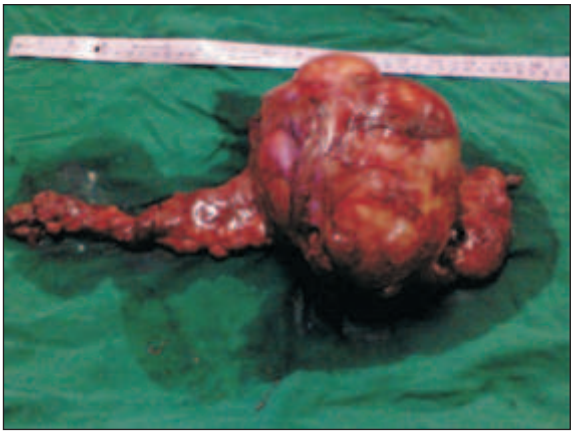

Fig 3: Large mesentric mass measuring about 15 X 12 cm with approximately 90 cm of jejunum adherent to it.

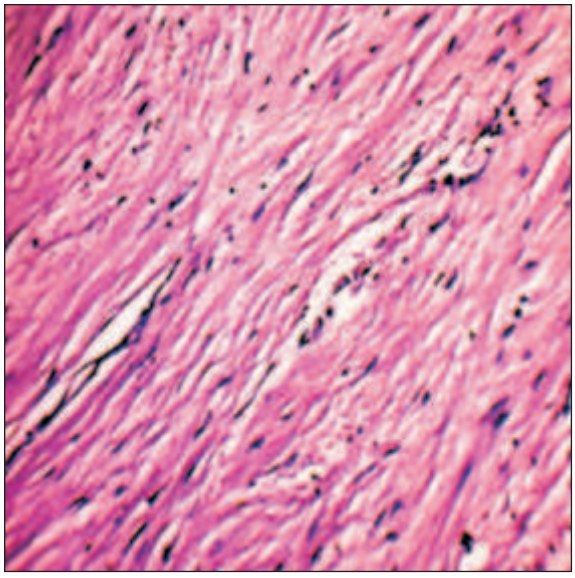

Fig4 : Microscopic picture showing spindle shaped cells with stroma containing eosinophilic collagen.

showed moderate cellularity with elongated slender spindle shaped cells. Stroma showed few eosinophilic collagen fibers. Thin walled vessels were seen with perivascular hyalinisation. Masson trichrome staining was negative for smooth muscle. Sections studied from jejunum overlying tumor showed mildly flattened villi, lamina propria showed lymphoplasmacytic infiltrate. Features were suggestive of mesentric fibromatosis (Fig 4). Immunohistochemistry was done, which stained negative for CD 117 and CD34. Post operatively colonoscopy was done which was normal and ruled out association with FAP syndrome.

Discussion :

Mesenteric fibromatosis are uncommon and often presents a diagnostic challenge. Its biological behavior is intermediate between benign fibrous tissue proliferation and Fibrosarcoma. The differential diagnosis are, a bland spindle cell tumor involving the GIT and from the mesentery includes GIST, fibromatosis and inflammatory myofibroblastic tumor[6]. Even though mesentric

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fibromatosis tends to be aggressive, there is considerable variability in their growth rate during the course of the disease. Usually they are characterized by initial rapid growth followed by stability or even regression. Commonly, patient presents with vague pain abdomen and distension. However, because of their ability to infiltrate they can present with intestinal obstruction, ischemia, perforation, hydronephrosis[7,8]. In spite of this overall 10 year survival rate for mesenteric fibromatosis can be as high as 60-70%[9].

The radiological features of these tumors are variable, there are no specific imaging features to distinguish between mesenteric fibromatosis and other mass lesions in the abdomen. CT scan and MRI scan are useful for evaluating the size of the tumor and involvement of adjacent structures[10].

Histopathological examination is essential for diagnosis. On gross examination tumor is well circumscribed and white and coarsely trabeculated on cross section. Microscopically mesenteric fibromatosis is characterized by a spatially homogenous proliferation of wavy spindle cells without atypia, associated with collagen among dilated vessels. The mitotic count is relatively low with no evidence of necrosis and nuclear differentiation[11].

Immunohistochemistry can differentiate mesenteric fibromatosis from GIST and inflammatory myofibroblastic tumors. Mesenteric fibromatosis positively stain for nuclear beta catenin and negative for CD 117 and CD34.Sometimes weak positivity or focal positivity for CD117 may be seen. GIST will stain positive for CD117 and CD34 but negative for beta catenin. Inflammatory myofibroblastic tumor stain negative for all the above three[12].

Many authors believe, complete surgical excision with a margin of uninvolved tissue is the treatment of choice for mesenteric fibromatosis. Unfortunately, radical surgery is not always a straight forward procedure because of the extent and invasiveness of the tumor. Involvement of base of mesentery and major portion of mesenteric blood supply makes surgery difficult and increases morbidity and mortality[13]. Debulking has no role, as it leads to more aggressive and infiltrative growth[14]. Inoperable growth should only be biopsied and other options of treatment need to be planned.

NSAIDs like Sulindac used post operatively and antiestrogen therapy with tamoxifen and testolactone can be used for many years. For clearly inoperable cases, cytotoxic chemotherapy, especially doxorubicin based or low dose vinblastin and methotrexate has been proposed. The evidence in the literature supports the opinion that both cytotoxic and non cytotoxic chemotherapy are effective against fibromatosis. However, lack of sufficient patients and randomized trials compromises the validity of the reported results[15].

In our patient, he was not put on any medications after surgery and was asked to be on regular follow up.

Conclusion :

There are no designed treatment protocols for mesentric fibromatosis. All modalities including surgery, chemotherapy radiation therapy are required in some cases. The rarity of these cases in high volume oncological centres has limited the ability to study the disease. Probably it is worthwhile to follow up all reported cases prospectively and draw some conclusion.

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