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Fibromatosis of Mesentery

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Abstract
Introduction: *Fibromatosis of mesentery are rare tumor frequently associated with Gardner syndrome. They are usually asymptomatic and are usually diagnosed post-operatively. The clinic-pathological features of these patients are retrospectively reviewed in this study.*
Method: *Study period was August 2014 to July 2015 done at NIMS, Hyderabad in department of surgical oncology. All patient with histological proven cases of mesenteric fibrosis were included in the study.*
Observation: *A total of 2 patient were proved to be case of mesenteric fibrosis. Both presented with abdominal lump. CECET abdomen showed intra-abdominal mass with heterogeneous enhancement. Both patient underwent surgical excision. Histopathological examination and IHC was done. One was diagnosed to be case of Gardner syndrome latter.*
Conclusion: *surgical resection is the standard treatment for mesenteric fibrosis. Endocrine therapy is usually recommended in in-operable cases.*
Key words: *fibromatosis, mesentery, Gardner syndrome*

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I. Introduction

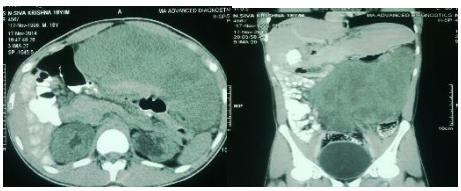
Mesenteric fibromatosis are rare benign tumor occurring most frequently in young adults with peak incidence at 30 years. In many cases the tumors are related to Gardner's syndrome. The majority of the patient remains clinically asymptomatic, until later in their course when they present with pain abdomen, distension and discomfort, vomiting, or other organ compressive symptoms. The diagnosis is only confirmed after the histopathological examination of the surgical specimen/ biopsy. Surgical excision is the standard treatment. We have reviewed our data and presented our experience.

II. Methods

The study was done at NIMS; Hyderabad from August 2014 to July 2015. The data was reviewed retrospectively and all the patients with histological prove cases of mesenteric fibromatosis were included in the study.

III. Observation

A total of two patients were included during the study period. All the patient presented with lump abdomen and mild abdominal pain. Some had symptoms of early satiety. Average age of presentation was 36 years with range from 18 years to 50 years. One patient was later diagnosed with Gardner syndrome. This patient was 18 year old and presented with lump abdomen with mild pain of 4 month duration. History of loss of appetite was present. 2 months back excision of a small occipital swelling was done which was reported as fibromatosis. There was no history of bleeding per rectum or melena. No history of vomiting or constipation.Clinically anill-defined abdominal lump approximately 25*25 cm occupying almost whole of abdomen was present along with scar over occipital area. External genitalia examination was normal. No supraclavicular lymph adenopathy. No other swelling. Per rectal examination was normal.Core biopsy from the lump done outside was reported as myxoid neoplasm, IHC inconclusive.Contrast abdominal CT revealed large intra-abdominal heterogeneously enhancing retroperitoneal mass/? Peritoneal, solid cystic mass. Bilateral hydroureteronephrosis (mass effect), few enlarged lymph nodes in pericardial fat on left side.



Contrast enhanced CT picture of the patient

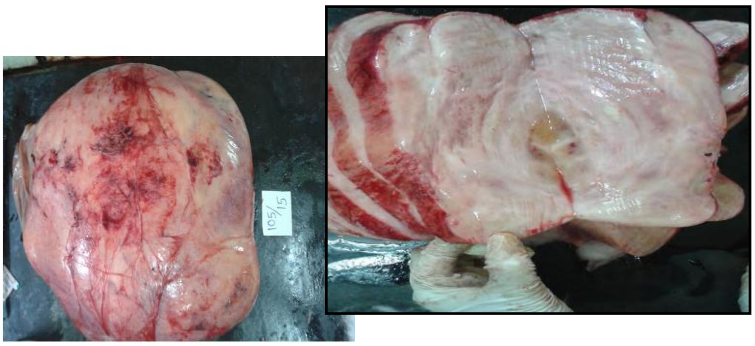
With the clinical diagnosis of retroperitoneal tumor? lipoma ? Fibroma patient was subjected for exploratory laparotomy.

Intra- operatively we found:

- 40*30*30 cm firm lump adherent to the stomach and transverse colon
- Multiple enlarged mesenteric and para aortic lymph nodes
- Multiple polyps in transverse colon (polyp sent for HPE)

En bloc removal of lump along with wedge resection of stomach+ segmental resection of transverse colon was done.

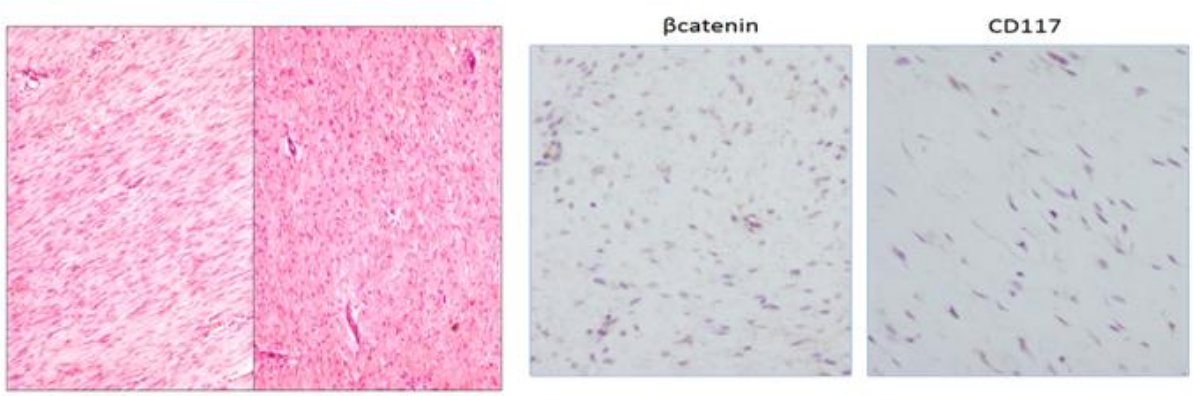
Gross specimen



Microscopic and IHC

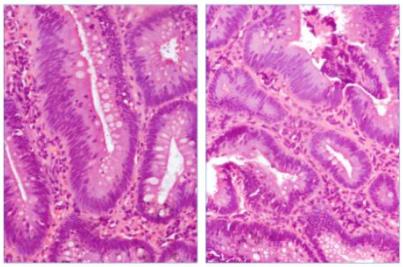
Microscopically it was characterized by spindle cells in abundant collagen stroma. Focally the spindle cells were arranged in short fascicle. Areas of myxoid changes and infarct seen. No evidence of significant pleomorphism or mitosis. No evidence of invasion to bowel wall.

Immune-histochemistry with CD117 rules out GIST. The lesion shows focal positivity with b-catenine. The tissue diagnosis of desmoid tumor was made.



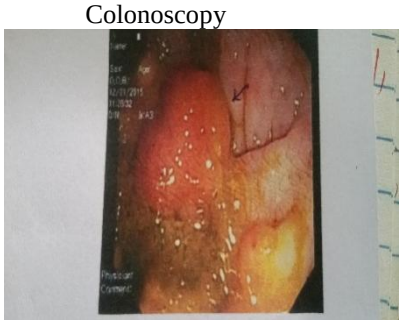
Colonic polyp

The polyp shows features of adenomatous polyp.

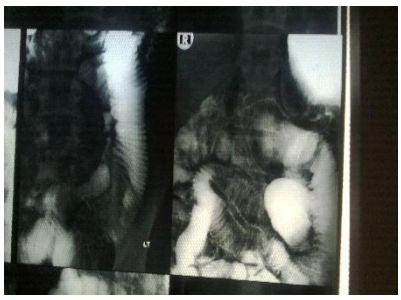


Retrospectively a colonoscopy was done which showed multiple polyp. Barium meal follow through was also done which was normal.

Colonoscopy



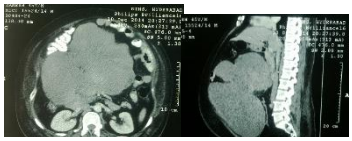
BMFT



Upper GI endoscopy was done which showed esophageal and duodenal polyps

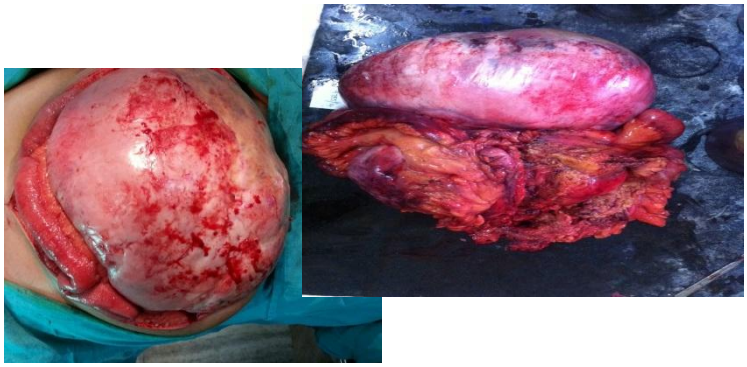
The patient was later labelled to be a case of Gardner's syndrome.

The other patient also presented with asymptomatic abdominal lump. Clinical examination revealed abdominal lump occupying almost whole of abdomen which was firm in consistency. CECT examination revealed large lobulated intra-abdominal solid mass likely arising from mesentery.



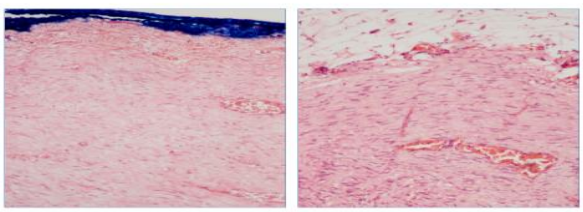
En bloc resection of mass was done which also required multiple intestinal resection.

Gross specimen

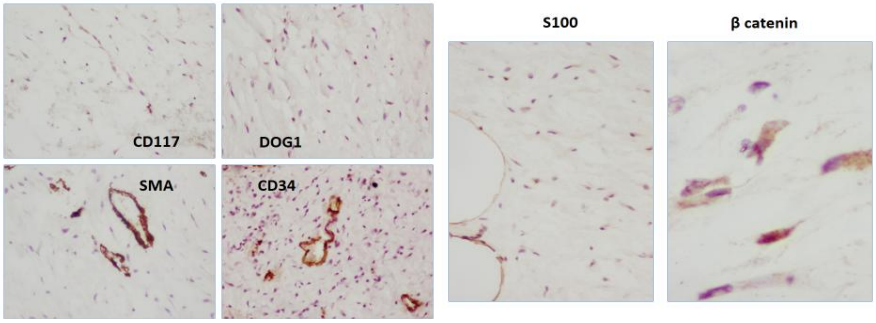


Microscopic

The microscopic picture and immunohistochemistry pictures



IHC



IV. Discussion

Fibromatosis are a group of similar lesion first coined by Stout in 1961 (6). **D**esmoid tumor represents a rare monoclonal neoplasm arising from deep musculoaponeurotic structures.[1-3]

Reported to affect approximately 2–4 per million persons annually. The deep variant involves the abdominal wall, mesentery, retro peritoneum, mediastinum and abdominal cavity. Mesenteric fibromatosis is very rare so there are no clear data of its incidence and presenting features

In many cases they are related to Gardner's syndrome where they occur as a part of the syndrome manifestation (Colonic adenomatous polyposis; Osteomas usually present in skull, mandible, and tibia;Soft tissue tumours like epidermoid cysts, fibromas, desmoid tumors.[4] Desmoid tumors occur at a rate of 10-15% in patients with FAP. Women of childbearing age are reported to be affected most often[5,6]. The intrabdominal desmoid are uncommon and together they are 0.03% of all tumors[7]. Sex incidence varies, with slight female predominance as documented by Dong Heup et all, whereas others have reported equal incidence among male and females[8,9].

The association ofintra-abdominal desmoid tumors with familial polyposis coli was made in 1923 by Nichols [10] and later reinforced in a series by McAdam and Goligher in 1970[11].The tumor is characterized by the mutation of the APC gene; a tumor suppressor gene located on the chromosome 5q21.Patients with FAP have a 1000-fold increased risk of developing desmoid tumors, comparedwith the general population.

An endocrine cause has been suggested in other patients due to its relative prepordance in perinatal women, with few tumor showing regression after tamoxifen therapy or after menopause. Furthermore estradiol receptors has been demonstrated in these tumors in high amount. Trauma especially post-surgery may also be contributing factor.[12]

Patient usually present late in the course of disease with lump abdomen, pain or symptoms due to mass effect like loss of appetite, bowel obstruction, hydronephrosis. Mesenteric fibrosis may show spontaneous regression, remain stable or depicts progressive growth. Some shows variable growth while some may have aggressive pattern of growth.[13]

CECT abdomen and pelvis is the imaging modality of choice which reveals soft tissue mass with adjacent structures involvement or displacements. The CT iconographic aspect of fibromatosis is directly related to the histology of the lesion, and in particular to the degree of cellularity and stromal component, as well as its vascularization. Lesions with abundant collagenous stroma will be homogeneous, with soft-tissue attenuation on CT.On MRI these tumors appears hypointense on T1- weighted images due to fibrous components, shows variable signal intensity on T2 images.[14,15] The predominance of myxoid component, will result in a CThypo-attenuation density.[16]

Microscopically mesenteric fibromatosis consists of spindle cells showing homogenous proliferation without atypia, associated with collagen among dilated vessel. The mitotic counts are low without any evidence of necrosis and nuclear pleomorphism or dedifferentiation. Immunohistochemistry demonstrate tumor cell being positive for vimentin and smooth muscle actin. Histologically it is differentiated from fibro sarcoma on the basis of absence of pleomorphism, significant mitotic activity or atypical mitosis. Absence of inflammation, fat necrosis and vascular invasion differentiate it from retroperitoneal fibrosis.On the basis of the degree of aggressiveness two pattern can be seen a 'nodular' form, characterized by 'mass-expansion' and a diffuse infiltrative one.[16]

The differential diagnosis can be gastrointestinal tumor for nodular desmoids. For diffuse pattern the differentials are lymphoma, adenocarcinoma. Other differentials are retroperitoneal soft tissue sarcomas/lipoma.

Wide field surgical excision is the first-line treatment for most mesenteric fibromatosis[17]. M .N Kulaylat and colleague studied 12 patient of desmoid tumor of which 4 were localized to abdominal wall and one in pelvis and one in retro peritoneum. One had mesenteric fibromatosis. They concluded that complete excision is the main modality of treatment for primary and recurrent disease. This is feasible in most of the cases except for tumors involving the base of bowel mesentry.[18]

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.[19]

Several medical approaches are also tried with or without surgical resection with mixed results These include chemotherapy with doxorubicin-based combinations[20], antiestrogen therapy with Tamoxifen[21,22],testolactone (which inhibits steroidaromatase activitiy) and its consequent reduction in estrogen synthesis[23], nonsteroidal anti-inflammatorydrugs (NSAIDs) such as indomethacin and sulindac.

Sulindac has been used in some studies with result showing evidence of partial reduction. Anika Hansmann and colleague 25 patient of desmoid tumor of which 17 were FAP associcated. Of the group of patients who received tamoxifen and sulindac as a primary treatment, all three patients with sporadic desmoid tumors demonstrated cessation of growth, and 10 of the 13 patients with FAP-associated tumors achieved either a PR or CR. Desmoid tumor recurrence after surgery was high and in the FAP-associated tumor group, therapy with tamoxifen and sulindac was found to be less successful. Based on this experience, the authors recommended high-dose tamoxifen and sulindac as the primary treatment for patients with FAP-associated desmoid tumors. The author concluded that the best approach after surgical intervention for patients with sporadic desmoid tumors remains to be determined.[24]

To conclude there is no established or evidence-based approachfor the treatment of this neoplasm. Surgical resection is standard approach. Endocrine therapy is usually recommended in inoperable cases, but might also be employed in postsurgical recurrences and in the preoperative setting.

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