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Case Report

A rare case of spindle cell tumour of mesentery

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ABSTRACT

We herein reported a rare case of a patient who developed a spindle cell tumor of the mesentery. A 45 years old female with a previous history of hysterectomy presented with pain in the abdomen for two weeks. CT scan was suggestive of a well-defined solid, enhancing lesion in the right pelvic region and iliac fossa measuring 10.9×13.9×15.1 cm, abutting right ovary, ileocaecal junction, caecum, appendix and rectosigmoid junction (possible differentials) are right ovarian neoplastic lesion or desmoid tumor. The patient was planned for surgery underwent laparotomy, where the mass appeared to originate from the mesentery (15×12) in dimensions and was adherent to distal ileum and caecum. The mass was inseparable, hence, the right quadra colectomy was taken and the whole mass with distal ileum and caecum and right colon was excised and ileocolic anastomosis was done. The tumor was histologically diagnosed as a spindle cell tumor. Follow up of the patient had been uneventful.

Keywords: Spindle cell tumor, Mesentery, Quadracolectomy

INTRODUCTION

Mesenteric fibromatosis or spindle cell tumor of the mesentery is a rare benign fibrous lesion that is commonly found in extrapleural locations such as lung, mediastinum, pericardium, mesentery, peritoneum, peritoneal spaces, nose, paranasal sinuses whose incidence is 2-4 per million. Its biological behavior lies in between benign fibrous lesions and fibrosarcoma. These tumors have a peculiar character of being locally aggressive but do not metastasize and they have a well-known local recurrence tendency.[1] Despite its rarity, mesenteric fibromatosis is the most common mesenteric tumour.[2] These tumors are usually benign in their course and remain asymptomatic and present late with abdominal pain symptoms, lump in the abdomen or symptoms arising due to pressure on adjacent tissues. Both computed tomography and magnetic resonance imaging can be used to diagnose these tumors. However, a definitive diagnosis relies on immunohistochemistry. Macroscopically most of the spindle cell tumor of the mesentery is round, well-circumscribed, encapsulated mass. They are typically tan white, firm and bosselated in appearance. It might be attached to the surrounding structures and hence surgical resection of the tumor is advised.

CASE REPORT

A 45 year old female with a history of hysterectomy done five years ago presented with chief complaints of pain in

the abdomen and a lump in the right iliac fossa. On clinical examination, a huge mass was palpable in the right iliac fossa extending to the right lumbar and suprapubic region and umbilical region measuring approximately 12×15 cm, which was partially mobile side by side. On routine investigations, the hemogram was suggestive of anemia, rest all reports were within normal limits. Contrast-enhanced computed tomography demonstrated a well-defined, solid enhancing lesion is noted in the right pelvis region and iliac fossa measuring approximately 10.9×13.9×15.1 cm in maximum dimensions. The lesion is seen abutting the right ovary, ileocaecal junction, caecum, appendix and rectosigmoid junction. The lesion is seen causing mass effect on right distal ureter with a prominence of the right proximal mid ureter and pelvic calyceal system.

Post hysterectomy status was noted. No evidence of any significant lymphadenopathy, ascites or omental thickening. The patient was planned for surgery. A lower midline incision of the abdomen was taken. The mass was identified as mesenteric in origin, 15×12 cm in size and adherent to distal ileum and caecum. It was inseparable, hence the decision of the right quadra colectomy was made and the whole of the mass with distal ileum and caecum and right colon was excised and ileocolic anastomosis was done. Both the ovaries were found to be normal. The mass was sent for the frozen section, which was reported as a benign spindle cell tumor. Hence omentectomy was done. Frozen section was reported to be a low-grade spindle cell tumor? desmoid fibromatosis? GIST. On gross appearance, the mass appeared bosselated with congestion and dilated vessels on the capsule. The mass measured 18×15×9.5 cm covered by an intact capsule. On one side, the mass was seen to be attached to the ileum and caecum with serosa and fat. On serially slicing the mass, it was seen to be a firm, white fleshy tumor having a homogeneous, white, pale-cut surface. The cut surface showed some pink and hemorrhagic spots with no evidence of necrosis. The ileum and caecum both appeared to be attached to the capsule of the tumor by their serosa. The appendix was found to be attached to the caecum, unrelated to the tumor capsule. The ileum and caecum's cut surface show thinning of the mucosa at the points where the tumor lies under it.

The histopathology report was suggestive of mesenteric fibromatosis (desmoid tumor) seen in the serosa, attached to muscularis of the caecum and ileum. The tumor shows a uniform, fascicular growth with low to moderate cellularity. The cells are wavy to spindly and show little to no atypia. The stroma is myxoid at places with small blood vessels. Tumour size was noted to be 18×15×9.5 cm with mitotic rate <1/10 hpf, no necrosis was reported. Lymphovascular/perineural invasion was not identified.

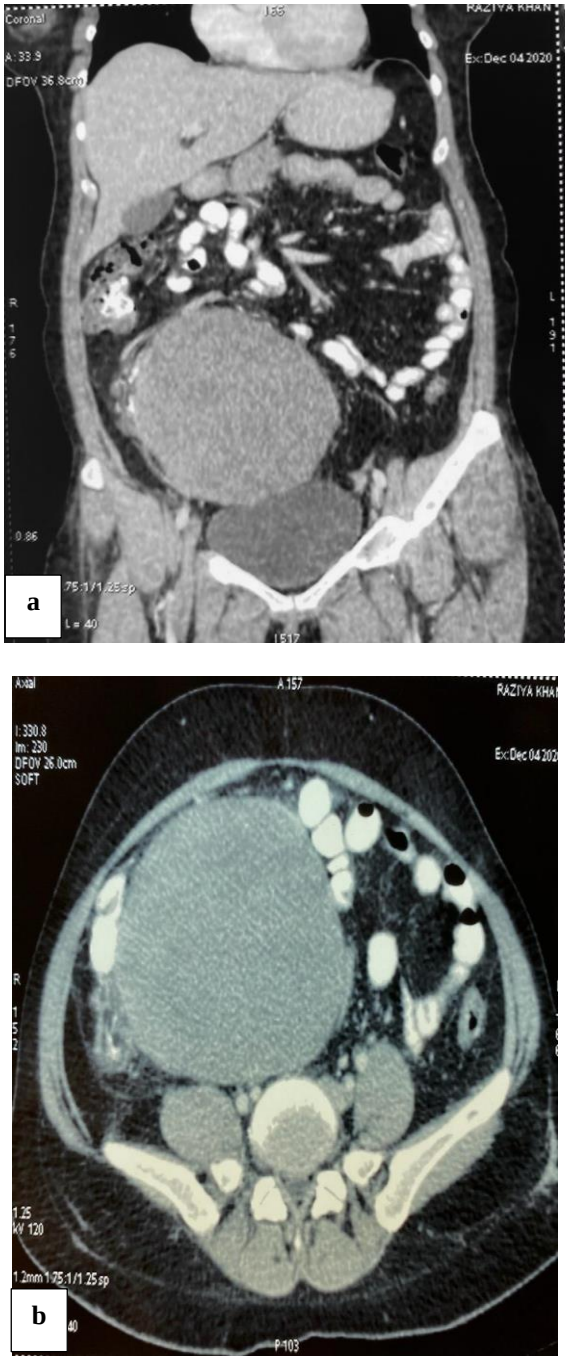

Figure 1 (a and b): A well-defined, solid, enhancing lesion is noted in the right pelvic region and iliac fossa, measuring 10.9×13.9×15.1 cm (AP×TR×CC) in max dimensions; the lesion is seen abutting right ovary, ileocaecal junction caecum, appendix and rectosigmoid junction.

Adjacent ileal and caecal mucosa show melanosis coli with both ileal and caecal cut margin are free. The appendix shows reactive hyperplasia of lymphoid follicles with no evidence of tumor. The omental tissue was found to be unremarkable. The abdomen's fluid shows clusters of mesothelial cells with hemorrhage and blood-derived inflammatory cells with no evidence of atypical cells. Immunohistochemistry was suggestive of

spindle cells are positive for B-catenin and negative for CD-34, SMS, S-100, C-kit, DOG-1 and desmin, ki-67 was found to below (2-3%).

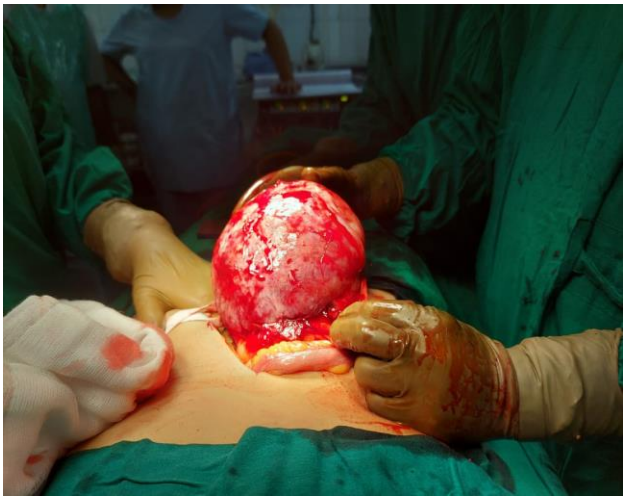

Figure 2: Intraoperatively the mass was 15×12 cm mesenteric in origin and was adherent to the distal ileum and caecum.

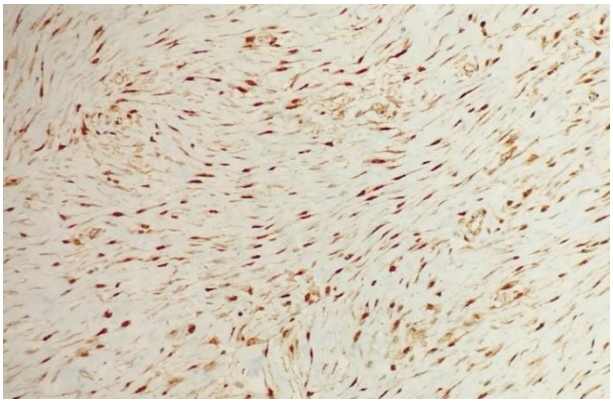

Figure 3: Cut section of the tumor showing beta catenin.

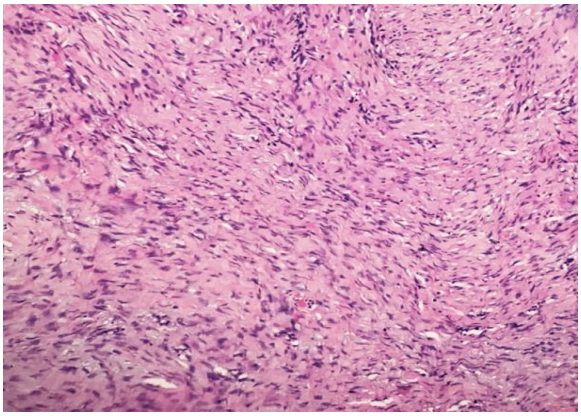

Figure 4: Section from mesenteric mass showing interlacing fascicles of bland spindle cells with thin vascular channels traversing.

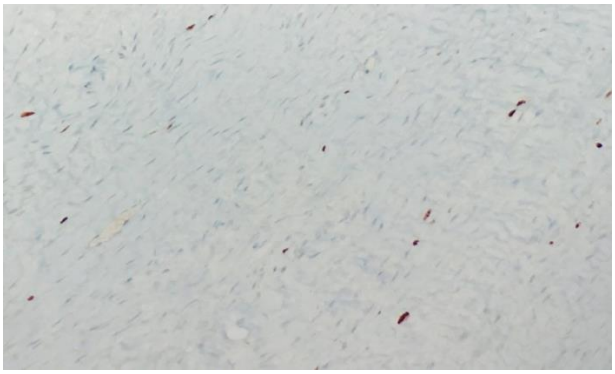

Figure 5: Image showing low ki-67 expression in the mesenteric tumor.

DISCUSSION

A solitary fibrous tumor is a rare mesenchymal spindle cell neoplasm, first described by Klemperer et al in 1931.[3] The etiology of solitary fibrous tumors is still unclear. It is commonly found in extrapleural locations such as lung, mediastinum, pericardium, mesentery, peritoneum, extraperitoneal spaces, nose, paranasal sinus.[4] Out of the rare cases of SFT of the mesentery reported in the literature, it was found that there is a male prevalence with a medium age of 54 years. Though here, we reported an unusual case of a female presenting with a mesenteric solitary fibrous tumor. SFTs exhibit a wide spectrum of clinical findings ranging from pain in the abdomen, abdominal discomfort, pressure symptoms on adjacent organs, lump in the abdomen. The tumor also may be presented with frightening symptoms resulting from complications such as torsion, hemorrhage or infarction of the mass.

Surgical resection is necessary to arrive at a final histological and immunohistochemical diagnosis. These mesentery SFT seem to occur more commonly in the ileum, as in our case in which the mass was adherent in the distal ileum and caecum. CT scan and MRI are effective in the diagnosis of SFTs. These tumors are generally round, well-circumscribed, completely enveloped, as reported in the literature.[5] Similarly, in our case, the surgically resected mass was rounded, well-circumscribed, bosselated with congestion and dilated vessels on the capsule. The mass was seen to be attached to the ileum and caecum with serosa and fat. However, it is considered a benign tumor usually localized in the small bowel mesentery, local recurrence after tumor resection was reported. As a result of its biological property and tendency towards involving adjacent visceral abdominal structures, an aggressive surgical approach is preferred including wide local excision with surgical excision of the adjacent involved structure as was advocated in our case. Most cases are sporadic, but there are associations with previous intra-abdominal surgery, estrogenic stimulation, Gardner syndrome, familial polyposis coli and bilateral ovarian fibromatosis.

Table 1: Comparison between different studies done on the mesenteric tumor.

Case no.	Age	Sex	Complaints	Locations	Tumour size (cm)	Treatment	Follow up (months)	Outcomes	References
1	33	M	NP	Mesentery	NP	Surgery	NP	NP	9
2	68	M	Abdominal pain	S-colon mesentery	18	Surgery	NP	NP	10
3	53	M	Abdominal pain	Distal ileum mesentery	22	Surgery	1	Alive	11
4	73	M	Abdominal pain	Mesentery	25	Surgery	NP	NP	12
5	71	M	Painless mass	Small bowel mesentery	15.5	Surgery	12	Alive	13
6	41	M	Abdominal pain	Mesentery	23	Surgery	7	Alive	14
7	26	M	Abdominal fullness	Proximal ileum mesentery	12	Surgery	18	Alive	15
8	36	M	Abdominal pain	Rectum mesentery	15.5	Preoperative RT surgery	NP	NP	16
9	59	F	Abdominal pain	Mesentery	21	Surgery	9	Recurrence	17
10	65	F	Abdominal pain	Distal ileum mesentery	18	Surgery	12	Alive	Our case

Other therapy modalities include treatment with antiestrogenic agents, steroids, cytotoxic chemotherapy and postoperative irradiation. Surgical excision is mentioned to be the only curative method for the treatment of this kind of tumor. Medical therapy is of unclear benefit. Sclerosing mesenteritis, mesenteric panniculitis, inflammatory fibrosracoma of the mesentery and retroperitoneum must be considered in differential diagnosis during pathological evaluation. The lesion is composed of proliferated fibroblasts and its histological features are specific enough to allow definitive diagnosis in the majority of cases. Unfortunately, in some cases, the differential diagnosis between fibromatosis and well-differentiated fibrosarcoma is difficult.[6] The differential diagnosis included tumors originating from smooth muscle, neuronal, adipose and stromal tissue of the gastrointestinal tract and tumors of histiocytic origin. All cases were treated by surgery, as in our case. SFTs exhibited a broad spectrum of clinical findings, therefore, surgical resection is necessary to arrive at a final histopathological and immunohistochemical diagnosis.[7] If the tumor was not resected, it eventually could have caused pain and obstructive symptoms by mass pressure on neighboring structures such as the ureter, small or large intestines. Burke reported 23% of all mesenteric solitary fibrous tumors recurred and there was a strong association between patients with Gardner syndrome and recurrence.[6] Recurrent disease could be resected and patients may live for extended periods with recurrent disease, especially in cases of sporadic desmoid tumors (spindle cell tumor of mesentery). Smith indicated that prolonged survival is possible in patients with intraabdominal desmoid tumors, even for those with technically irresectable or recurrent lesions.[8]

In general, SFTs comprise various cell types, with an abundance of spindle cells exhibiting patternless growth on histopathological examination.[7,10,18] Immunohisto-chemically, SFTs are commonly positive for CD34, bcl-2 and vimentin, but rarely positive for S100 proteins, desmin, actin and cytokeratins.[7] In our case, the tumor exhibited characteristics of an SFT by both histopathological and immunohistochemical analyses as the tumor shows a uniform, fascicular growth with low to moderate cellularity. Although most SFTs are histopathologically benign, up to 20% may be malignant.[18] As shown in Table 1, recurrence was reported in only one case (number 9). Other therapy modalities include treatment with antiestrogenic agents, steroids, chemotherapy and postoperative irradiation. Surgical excision has been the only curative method for treating this kind of tumor in literature. Medical therapy is of unclear benefit until now and should be kept in mind as complementary methods in managing these patients. Still, they are no substitute for surgical resection.

The role of other treatment modalities in the management of SFTs is unclear. Some reports in the literature have demonstrated the effectiveness of radiotherapy for the control of SFTs.[19,20] Chemotherapeutic drugs and novel targeted drugs seem to exert some activity on SFTs. However, a consensus has not been achieved concerning

the effectiveness of radiotherapy and chemotherapy against SFTs. Long term follow up has been advocated in literature due to incidences of recurrence. Our case was followed for six months with no recurrence to date.

CONCLUSION

Although it is difficult to make a definitive diagnosis of a solitary fibrous tumor before surgery and histopathological diagnosis, CT or MRI may help to identify the tumor. Imaging techniques can be used as the first-choice method of diagnosing this tumor and for analyzing its morphology, density and composition. Enhancement is also essential. The most effective therapeutic modality for SFTs is surgical resection with clean margins. In our case, the tumor was removed entirely during surgery without any adjuvant chemotherapy or radiotherapy. Long term careful follow up is necessary because patients with a large tumor and a high mitotic index may have malignant potential and an increased propensity for recurrences.

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