Case Report

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Case Report: Mesenteric Fibromatosis

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ABSTRACT

A 28-year-old patient reported a lump in the right lower abdominal and pelvic area for the last 21 days along with pain, anorexia, fever, and nausea. After a thorough investigation, an AP scanogram of the patient showed a clearly defined lump in this area. Histopathological examination showed mesenteric fibromatosis. Resection and anastomosis were done.

Our case is one of the few known cases of the extremely rare condition fibromatosis of the mesentery. These tumors present a diagnostic and therapeutic difficulty due to their rarity. These tumors are still asymptomatic; however, they frequently appear as an asymptomatic mass. There aren't any obvious therapy options. The preferred method of treatment is surgical excision with a wide margin; we underwent the same technique, and the prognosis was good. Numerous single case reports with various and peculiar appearances and their difficulties exist. This case report aims to offer a current understanding of mesenteric fibromatosis as well as our clinical experience with the presented patient. Following surgery, the patient did well, and a three-month follow-up revealed a typical recovery.

Keywords: Mesenteric Fibromatosis; Lump; Surgical Excision

INTRODUCTION:

Small intestine mesentery affected by a proliferative fibroblastic lesion is known as mesentery fibromatosis (MF). In 8% of desmoid tumors, it is present. It makes up 0.03% of all tumors. The clinical pathologic spectrum of deep fibromatosis includes mesenteric fibromatosis [1]. In terms of biology, this uncommon tumor is capable of infiltration or recurrence and is locally aggressive without the ability to metastasize. It resembles mesenchymal tumors of the digestive tract called gastrointestinal stromal tumors (GISTs) [2]. The majority of instances that have been recorded have been linked to familial adenomatous polyposis (Gardner Syndrome), prior trauma, and prolonged estrogen use. Although mesenteric fibromatosis accounts for 80% of intra-abdominal desmoids, it is extremely uncommon, occurring in about 2–5 instances per million people [3]. With a reported incidence of 10–15%, familial adenomatous polyposis has a strong correlation. Mesenteric fibromatosis's true origin is uncertain. Patients with Mesenteric Fibromatosis typically experience sluggish growth before developing symptoms [4]. Activated surveillance medical therapy (non-steroidal), anti-inflammatory medicines, (anti-estrogenic) agents, chemotherapy, radiation, and surgery can all be used in a center with experience treating sarcomas. Reporting of Mesenteric Fibromatosis carries a lot of significance as these are often misdiagnosed due to their common resemblance to many other tumors clinically, radiologically, and in rare cases, pathologically as well.[4] Misdiagnosis may lead to a poor prognosis. This instance acts as a teaching moment to emphasise Mesenteric Fibromatosis' irrational nature [5].

The present report outlines a case of mesenteric fibromatosis, with the patient presenting with a complaint of pain in the abdomen, which was managed at Dbp-Dr Anant Pore, in front of APMC, Achalpur Road, Paratwada dist, Amravati.

Keywords: Desmoid Tumors; Mesenteric Fibromatosis; Gastrointestinal stromal tumor (GIST); Fibromatosis, Familial adenomatous polyposis; Cancer recurrence; Medical oncology.

Case report

An examination by CT revealed a big tumor coming from the mesentery that was on the verge of rupture in a 28-year-old male patient's central and lower right abdominal and pelvis region. The pain was dull and lasted for 2 months. There were no additional symptoms, and it was steadily getting bigger. There was no substantial family history or medical history in the past. Following a preliminary A.P. scanogram of the abdomen, a histological study was conducted.

Examining the A.P. Scanogram reveals a large solid cystic lesion that originates from the small bowel mesentery and invades the jejunal loop by about 15-20 cm. This lesion is about 30 cm long and 18 cm wide, and it extends up to the superior part of the urinary bladder. It is firm, mobile, and mildly tender, with a smooth surface and margin. The lesion exuded from a congested area of the external surface. The gross examination of the cut section shows firm to myxoid areas with a cyst measuring 5×3×2cm. The distance from the proximal margin is 12 cm, and from the distal margin is 8 cm. The gross examination revealed that the tumor did not cause any perforation. The hypodense collection in the cystic areas contains multiple air foci within. The walls of the lesion appeared indistinct along the lateral aspect.

The right lower ureter, common and adjacent part of the external iliac artery, were seen closely abutting the lesion and sigmoid colon. The adjacent anatomic organs (liver, gall bladder, spleen, pancreas, kidneys, retroperitoneum, G.I tract, bladder, prostate, lungs 'pleurae) showed no gross morphological and anatomic changes. Mild ascitic fluid collection was seen in the abdomen, and visualized bones did not show any significant focal lesion. On rare occasions, bowel obstruction, perforation, or gastrointestinal bleeding can coexist with mesenteric fibromatosis. Very seldom does mesenteric fibromatosis manifest as an abscess.

Along with the ileum resection, an omentum specimen measuring 18×5×2 cm was received. Omentum's external surface had exuded from the congested area, and a solid nodular area was absent. There was an unremarkable status of residual bowel (i.e., stricture, polyp, ulcer, diverticula). Peri–colonic lymph nodes were present. There was the presence of the largest lymph nodes, and in total, 6 lymph nodes were examined.

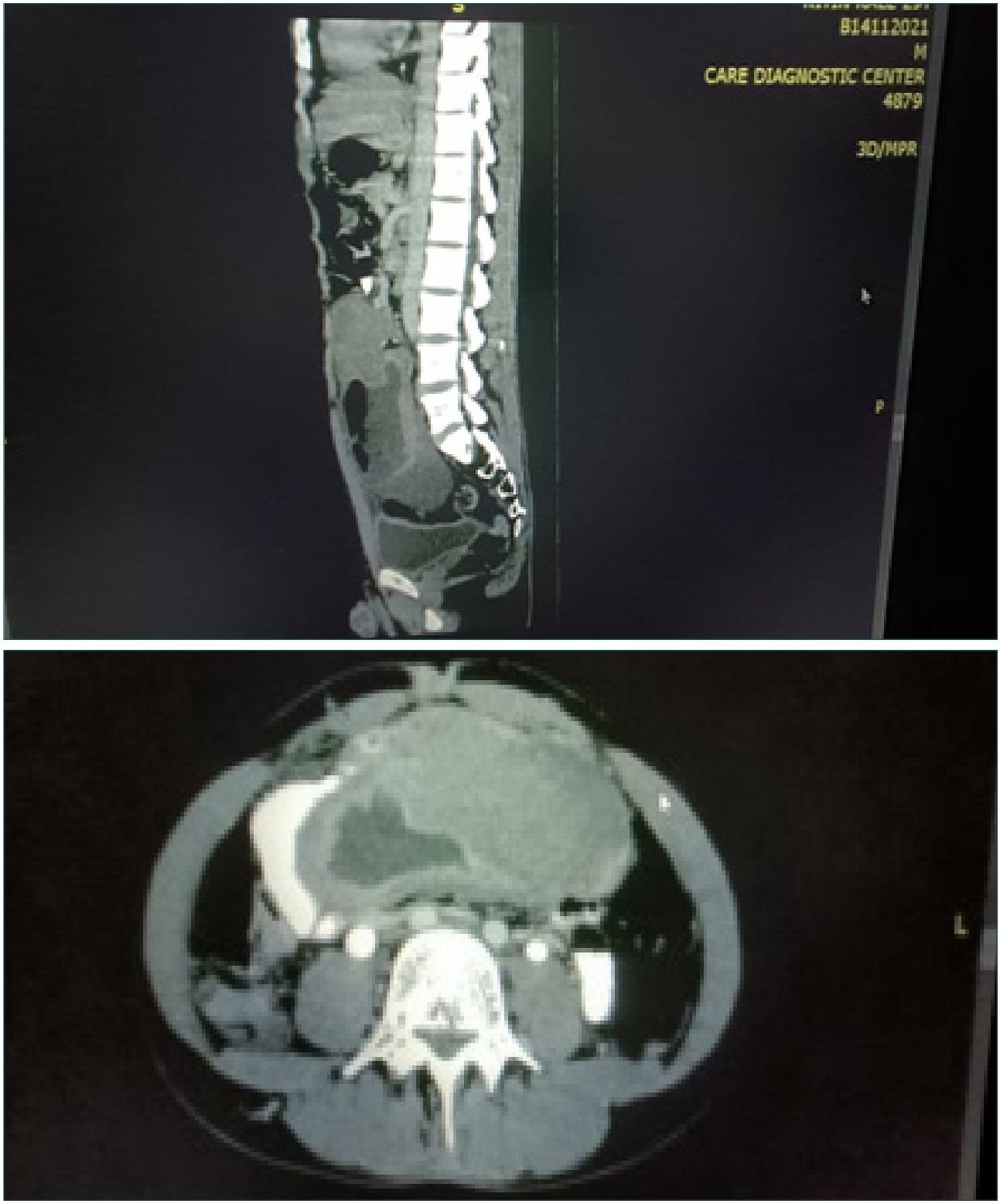
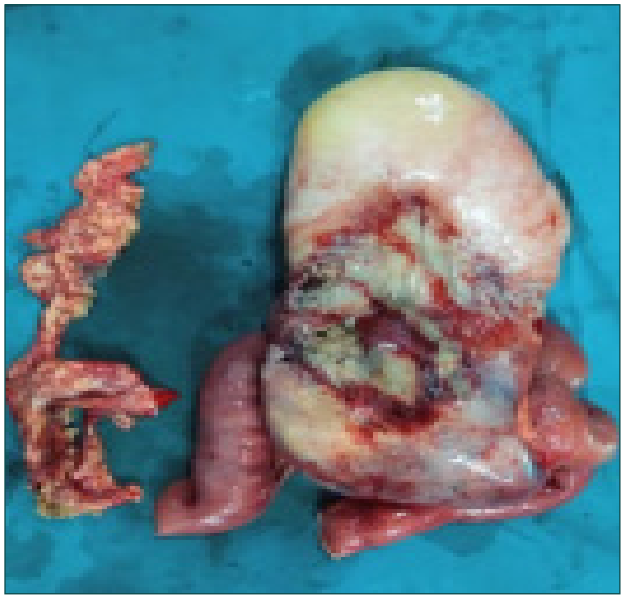


Figure 1-2: Figure 1, figure 2 showing a large well-defined intra-abdominal mass in the right abdomen



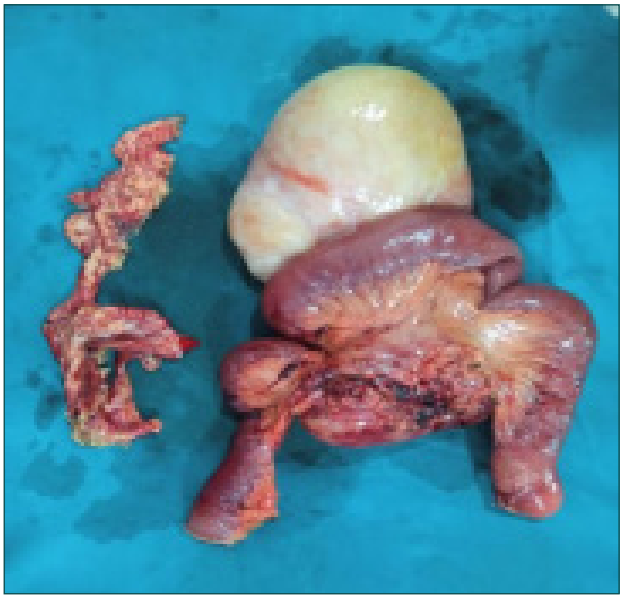


Figure 3-4: Figure 3, 4 represent the resected mesentery of the abdomen

Histologically, multiple sections were examined. Fibromatosis is a fibroblast-derived tumor that is histologically benign but locally aggressive. These sections showed the presence of a tumor made up of long, sweeping fascicles of spindle cells with elongated nuclei and tapering ends, as well as eosinophilic cytoplasm that blends invisibly with collagen bundles. The tumor cells insinuate between the adjacent skeletal muscle bundles, entrapping individual muscle fibers, which show degenerative giant cell forms. A few typical mitotic figures were seen. Complementary IHC shows positive Beta Catenin. Ki-67 showed a low index. The histological study revealed no increase in mitotic activity and the absence of necrosis. The margins were free of tumor. Numerous blood vessels are seen along with areas of hemorrhage.

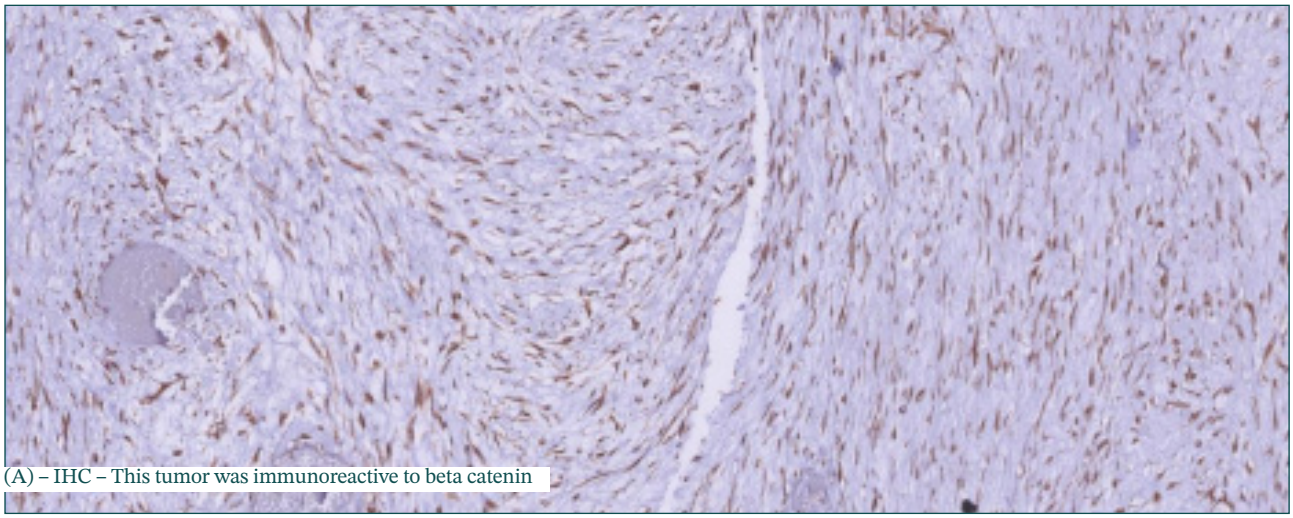
In the histological study, the omentum was devoid of tumor. A total of 15 lymph nodes were identified. All 15/15 show reactive lymphoid hyperplasia. Postoperative days were uneventful, and on the tenth day, patients were released. In the days following surgery, the patient received adjuvant therapy. The good prognosis requires follow-up.

Discussion:

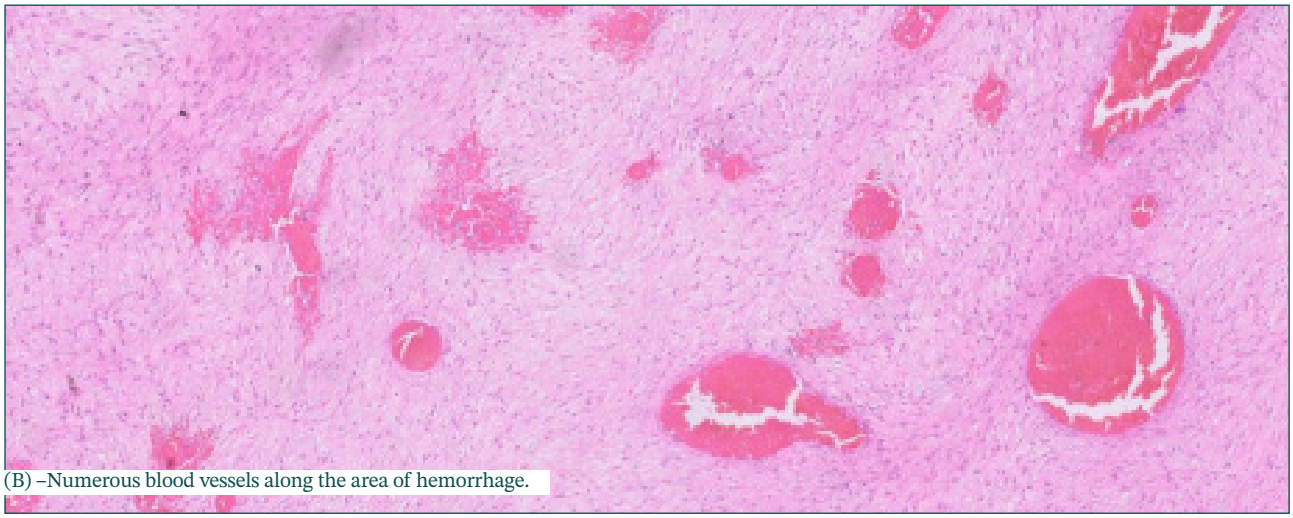
Muller first used the name “desmoid” in 1838, and it is derived from the Greek word “desmos,” which means band or tendon. These are extremely rare tumors that make up 3.5% of all fibrous tissue tumors and account for 0.03% of all neoplasms. They are caused by musculo-aponeurotic components.

Due to the low frequency of prevalence of mesenteric fibromatosis, the following chart depicts the cases reported in the past 10 years.

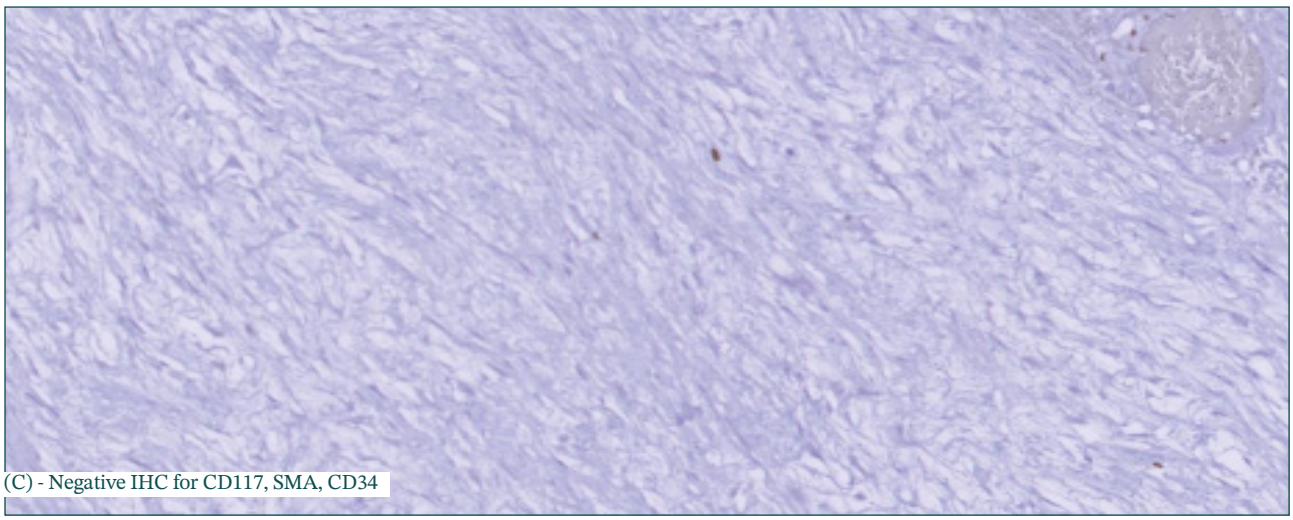
Desmoid tumors are connective tissue growths that are not malignant. Desmoid tumors typically develop in the arms, legs, and abdomen. Desmoid tumors are also referred to as aggressive fibromatosis. Some desmoid tumors grow slowly and don’t need to be treated right away. Desmoid tumors are locally invasive, do not spread, and significantly increase morbidity and mortality. DTs have a high rate of local recurrence despite surgical resection; as a result, the World Health Organization has classified them as an intermediate locally aggressive tumor. Desmoid tumor is classified into 3 types based on etiology: abdominal, extra-abdominal, intra-abdominal, multiple familial, associated with Gardner syndrome, and associated with Turcot syndrome. There are numerous masses in the central, lower, and pelvic areas, ranging from retroperitoneal to peritoneal cavity to abdominal wall-derived masses. These range from widespread conditions, including appendicular tumor or abscess, ileocaecal TB, carcinoma caecum, and lymphoma, to relatively uncommon diagnoses like retroperitoneal sarcoma, iliac artery aneurysm, or chondrosarcoma of the iliac crest.



(A) – IHC – This tumor was immunoreactive to beta catenin



(B) –Numerous blood vessels along the area of hemorrhage.



(C) - Negative IHC for CD117, SMA, CD34

Figure 5: A, B, C represents the histological features of Mesenteric Fibromatosis

Table 1. The chart depicts the cases reported in the past 10 years.

Sr	Author	Location	Management	Prognosis	Year
1	Masahiro Tawada[6]	Lower right	Complete resection without	Good.	2021
2	Yingying XU[7]	Entire left	Systemic therapy along with surgery.	Good.	2020
3	K. Seidenosaal, S.B. Harrabi, and M. Uhl.[8]	Abdominal	Radiotherapy and pharmacotherapy.	Excellent.	2020
4	Jian Li.et.al[9]	Right abdomen with compression of	Resection without chemotherapy or radiotherapy.	Good.	2019
5	E. Reza et al[10]	Right abdomen.	Surgical excision.	Good.	2019
6	Geeta Karbeet[11]	Right abdomen.	Laparotomy	Good.	2017
7	George Ap.Efthimiapoules.et.al	Lower right quadrant of the abdomen	Surgical approach	Good	2015
8	Anandaravi BN et al[12]	Lower abdomen.	Complete excision	Good.	2015
9	Poras Chaudhary et.al [13]	Right abdomen.	Surgical excision	Good.	2014
10	Zvi Peled et.al [14]	Lower right abdomen.	Surgical excision, chemotherapy, and radiotherapy.	Excellent.	2012

Trisomy of chromosomes 8 or 20 (sporadic form) [3], trauma, prior abdominal surgery, hormone stimulation, and occasionally de novo is some of the causes of DT.

A painful abdominal mass, an asymptomatic condition, bowel or ureteral blockage, mucosal ischemia, intestinal perforation, fistula, pyrexia of unclear cause, or functional impairment of the ileoanal anastomosis are all possible manifestations of mesenteric fibromatosis. Since mesenteric fibromatosis doesn't typically present with symptoms, the diagnosis is only made after a histological examination of the tumor. To make a working diagnosis of mesenteric fibromatosis, imaging continues to be the cornerstone of preoperative studies. The intralesional vascularity of the tumor and the collagen and fibroblast content are the main determinants of the non-specific sonographic characteristics of mesenteric fibromatosis. The CT scan is regarded as the first-line imaging modality for diagnosing, defining, and staging fibromatosis due to the propensity of aggressive mesenteric fibromatosis to invade neighboring structures.

Mesenteric fibromatosis is visible under a microscope as a spatially uniform proliferation of wavy spindle cells without atypia, accompanied by collagen in the vicinity of dilated arteries. The number of mitoses is rather low, and neither necrosis nor nuclear dedifferentiation is present.

It is commonly misdiagnosed as another tumor, so the following features differentiate mesenteric fibromatosis from other conditions.

Features to differentiate mesenteric fibromatosis from other tumors

- It is the primary tumor of the mesentery
    - The majority of occurrences are sporadic; however, some are caused by trauma, the Gardner syndrome, or hyperestrogenic conditions.
- Mean age – 35 years
    - Site - Small bowel mesentery, ileocolic mesentery, gastrocolic ligament, omentum, or retroperitoneum are examples of mesenteries.
- Most patients present with an asymptomatic abdominal mass.
- Gross – large ≥10 cm, well circumscribed
    - Complications can arise from intestinal perforation, small- or large-intestine compression, or compression of the ureter.
- Locally recurrent
    - Microscopic features – A highly collagenous stroma that is uniformly layered with spindle-shaped or stellate cells that exhibit cytological blandness.

    There is typically a range of cellularity, with some areas exhibiting dense fibrous tissue and others possibly exhibiting myxoid alteration.

    There are prominent dilated thin-walled blood vessels.

    The number of mitoses is rather low, and neither necrosis nor nuclear dedifferentiation are present.

    - IHC – Positive stain; vimentin, β-catenin, variable smooth muscle actin, and muscle-specific actin. Negative stain – CD34, keratin, s-100, desmin, CDu7
- Differential diagnosis
    1. Mesenteric panniculitis and mesenteric lipodystrophy (Sclerosing mesenteric); these conditions are characterized by fibrosis and chronic inflammatory infiltrate.
    2. Fibrosarcoma
    3. Idiopathic retroperitoneal fibromatosis
    4. GIST
    5. Leiomyoma
    6. Neurofibroma
    7. Schwannoma
    8. Keloid

Furthermore, germline mutations in the adenomatous polyposis coli (APC) gene, which cause familial adenomatous polyposis (FAP), have been linked to the onset of desmoid tumor, and it has been reported that the risk of desmoid tumor onset in FAP patients is 800 times higher than that in a healthy person. Spontaneous desmoid tumors are thought to be caused by somatic mutations of the catenin beta-1 (CTNNB1) gene in addition to the APC gene. It has been estimated that 50–85% of cases of desmoid tumor had CTNNB1 gene mutations. Both genes participate in significant signal transduction pathways that control the cellular level of -catenin, a protein that affects cell-cell junctions. Consequently, whenever a mutation is found in either APC or CTNNB1, β-catenin accumulates in cells, forming tumors. Desmoid tumors frequently affect women, especially those who are multiparous, as estrogen has been linked to their growth and development. Tamoxifen has recently been discovered to be beneficial as a treatment for desmoid cancers, which have strong oestrogen receptor expression. Desmoid tumors are thought to be influenced by a history of trauma or surgery as well as gene alterations that affect tissues' ability to mend wounds. A broad bundle of stromal fibrosis was formed as a result of bland spindle-shaped cells proliferating with little mitotic activity, according to histopathological findings. (Fig. 3a). Based on positive immunohistochemistry staining for -catenin (Fig. 3b) and negative staining for c-kit, CD34, platelet-derived growth factor (PDGFR), and Discovered on GIST-1 (DOG-1), the tumor was identified as a desmoid tumor. There were no aberrant findings during physical examination, serum biochemistry study, or upper or lower gastrointestinal endoscopy.

Its respectability and attachment to surrounding structures are revealed by CT and MRI scans. The only way to confirm is through a histological study. GIST, lymphoma, carcinoid tumor, fibrosarcoma, and inflamed fibroid polyp are examples of differential diagnoses.

Treatment modalities include antiestrogen, chemotherapy, and radiotherapy. According to Fotiadis C et al [6], DT in the abdominal wall is less difficult to remove and has a lower chance of returning than mesenteric or retroperitoneal DT. 25% to 50% of recurrences occur after undergoing major surgery and adjuvant radiation. In their study on the use of radiation in DT, Kriz J et al. demonstrated its value with a local control rate of 79% with long-term follow-up and suggested it as a substitute for mutilating Surgery. In the immediate aftermath of surgery, our patient had adjuvant radiotherapy (50 Gy).

In cases of recurrence and inoperable lesions, radiotherapy may be administered before surgery to reduce the tumor and make it operable. In contrast to the 40% to 70% recurrence rate with resection alone, adjuvant radiation therapy lowers mesenteric fibromatosis recurrence to 20% to 40%.

Surgical procedure

In our patient, the surgical modality was performed as a treatment option. The following surgical steps were followed precisely. As the tumor was close to the SMA (Superior mesenteric artery), the tumor was separated carefully from the SMA pedicle. As about 15 - 20 cm of the jejunal part was invaded by the tumor, so endobloc resection of the tumor was performed, followed by excision of 5 cm of the proximal and distal margins of the ileum. Along with the excision, side-to-side hand-sewn anastomosis was performed in two layers with Prolene 3-0. During the surgical procedure, approximately 50 ml of blood was lost. Romo ADK no.18 drain was kept. Closure of the excision was performed in layers with loop PDS on the skin with a stapler. After the post-operative procedure and histological examination, the report was referred to the medical oncologist (Dr Sushant Ikhar).

Pathology:

Although they are commonly linked to prior trauma or surgical incision, their specific cause is yet unknown. Desmoid tumors are defined by mutations in the -catenin, CTNNB1, or adenomatous polyposis coli (APC) genes at the genetic level.

Conclusion

Desmoid tumor differential diagnosis is frequently made in middle age, despite its rarity. To find tumor recurrence, a multidisciplinary approach and long-term monitoring are required. As our diagnosis was confirmed to be mesenteric fibromatosis, all resected margins were negative. The patient was kept on close follow-up of 2-3 months with CT SCAN. After regular follow-up of 6 months, no recurrence was observed. Despite the risk, there remains a role for surgery in the management of large mesenteric desmoid.

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