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Incidental Finding and Management of Mesenteric Fibromatosis

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 45
Final Diagnosis: Mesenteric fibromatosis
Symptoms: —
Medication: —
Clinical Procedure: Surgical removal of the mesenteric fibromatosis
Specialty: Surgery

Objective: Rare disease





Background: Mesenteric fibromatosis, also known as mesenteric desmoids, is part of the clinical-pathologic spectrum of deep fibromatosis, which encompasses a group of benign fibro-proliferative processes that are locally aggressive and have the capacity to infiltrate or recur without metastasis.

Case Report: Case of a 45-year-old man, with a history of hypertension and lung fibrosis, presenting for a left abdominal mass, which was found incidentally during his lung fibrosis imaging. He complained of constipation due to pressure upon his bowel leading to difficulty in defecation.

Conclusions: Although there are many overlapping criteria between gastrointestinal stromal tumors and mesenteric fibromatosis, making it difficult to discriminate between the two, there are differences that are unique to mesenteric fibromatosis that should be noticed during the diagnosis. In this case, mesenteric fibromatosis was unusual as it is not associated with Gardner's syndrome, desmoid tumors, nor familial adenomatous polyposis, but was an incidental finding.

MeSH Keywords: Abdominal Neoplasms • Fibromatosis, Abdominal • General Surgery • Gardner Syndrome • Gastrointestinal Stromal Tumors • Adenomatous Polyposis Coli

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Background

Mesenteric fibromatosis (MF), also known as mesenteric desmoids, is part of the clinical-pathologic spectrum of deep fibromatoses which encompasses a group of benign fibroproliferative processes that are locally aggressive and have the capacity to infiltrate or recur without metastasis. They are classified according to their anatomical location, whether it is intra-abdominal, from the deep soft tissues of the abdominal wall, or deep within the extra-abdominal soft tissues [1].

Unusual cases of intra-abdominal fibromatosis that involve the bowel wall are commonly misdiagnosed due to their different biological behavior and rarity. [2] The two most commonly misdiagnosed conditions are between mesenteric fibromatosis and gastrointestinal stromal tumors.

Although mesenteric desmoids account for less than 10% of sporadic desmoid tumors, 70% are intra-abdominal, and most involve the mesentery [2]. They can arise spontaneously after surgical trauma or abdominal surgery [3]. Moreover, after extensive research, we found that a strong association has also been reported between desmoid tumors and familial adenomatous polyposis (FAP), especially Gardner's syndrome [2]. Thirteen percent of patients with mesenteric fibromatosis have FAP, specifically the Gardner syndrome variant of FAP [1]. Therefore, patients with FAP and a family history of mesenteric desmoids have a greater than 25% chance of developing this tumor [2]. Additionally, 83% of patients with FAP and mesenteric fibromatosis have a history of abdominal surgery, most commonly a total colectomy [1]. Mesenteric fibromatosis mainly occurs in female patients (80% of cases) from 14 to 75 years of age (mean: 41 years), without any preference in race [1]. The female predilection is due to the fact that estrogen, even exogenous estrogen, is a factor that predisposes one to mesenteric fibromatosis and plays a role in its formation. MF occurs more frequently during pregnancy and in premenopausal women compared to postmenopausal women [3]. The patient usually presents with signs and symptoms related to the small bowel such as abdominal pain or a palpable abdominal mass, or clinical complications like gastrointestinal bleeding, small bowel obstruction, fistula formation, or bowel perforation [1].

Although MF has characteristic morphologic features, when the lesion involves the wall of the stomach or bowel, which appears to be the site of origin, it mimics a primary gastrointestinal neoplasm, gastrointestinal stromal tumors (GIST) [4]. The differential diagnosis between MF and GIST is further complicated by reports of positive cytoplasmic immunostaining of MF with CD117, a key marker for the diagnosis of GIST [4].

GISTs originate from the gastrointestinal pacemaker cells of Cajal, the primary effectors controlling gut motility [2]. GISTs

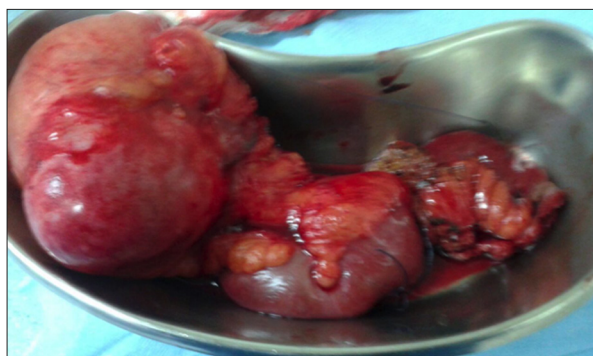


Figure 1. Abdominal MRI, vertical and transverse cuts.

can grow into the organ lumen, remain entirely on the serosal surface, or even become pedunculated within the abdominal cavity. They spread via direct invasion or blood-borne metastases. They may develop into a large size and present with bleeding. Radiologically, a central ulceration is present due to necrosis from the outgrowth of its blood supply [2].

Case Report

This is the case of a 45 year old man who presented for a left abdominal mass. He denied ever being in pain but complained of tension on his left side. He also experienced pressure on his bowel leading to difficulty in defecation. He denied fever, nausea, vomiting, and diarrhea. The mass was incidentally discovered four months ago upon imaging his lungs. His history goes back to 2011 when an idiopathic pulmonary fibrosis was diagnosed after recurrent pleuro-pericardial effusion, which was treated with pleural tap then decortication of the left lung in 2013. No medication was needed since the patient remained stable and the disease was stabilized without further progression, but continued checkups were scheduled. A pleural biopsy and pleurectomy was negative for malignancy and showed normal pulmonary functions and arterial blood gases. He is currently on valsartan, amlodipine, and pantopren. His known allergy is to contrast dye. Upon asking about his daily complaints, he stated that he experiences palpitations and shortness of breath as well as difficulty swallowing.

Concerning his family history, his father is known to have coronary artery disease as well as colon cancer, and his mother has hypertension.

The patient is married with 2 healthy children. He drinks only 2 cups of coffee a day and denies drinking alcohol, smoking, or any other substance abuse.

The investigation of the mass began with an abdominal MRI (magnetic resonance imaging) as shown in Figure 1. A left mesenteric mass at the level of L5–S1 was seen anterior to the



Figure 2. Gross view of excised retroperitoneal mass.

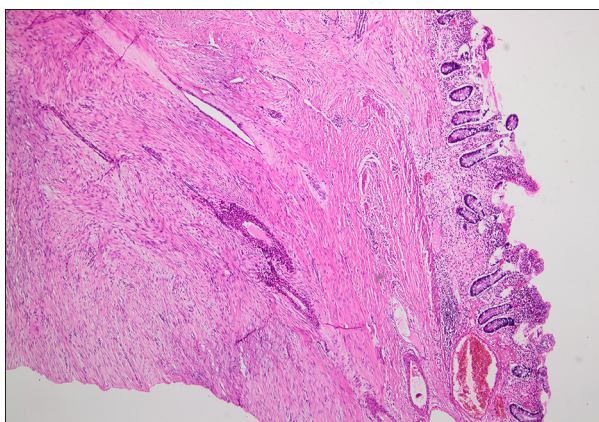


Figure 3. Pathology slide of the mesenteric fibromatosis.

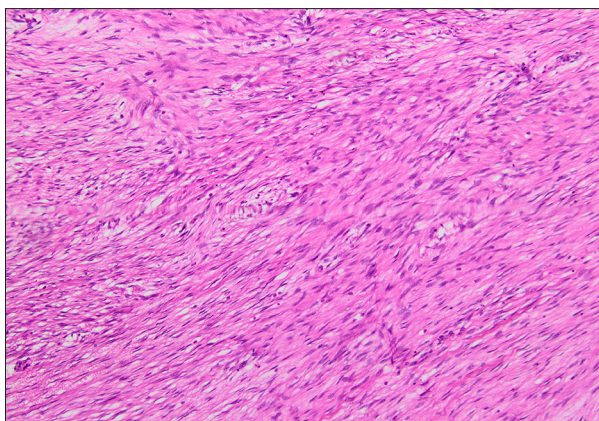


Figure 4. Pathology slide of the mesenteric fibromatosis – homogeneously distributed bland spindle cells.

iliopsoas muscle, which measured approximately 88 mm (crano-caudal) ×76 mm (anteroposterior) ×62 mm (transverse). On its proximities, it was in contact with the adjacent jejunal

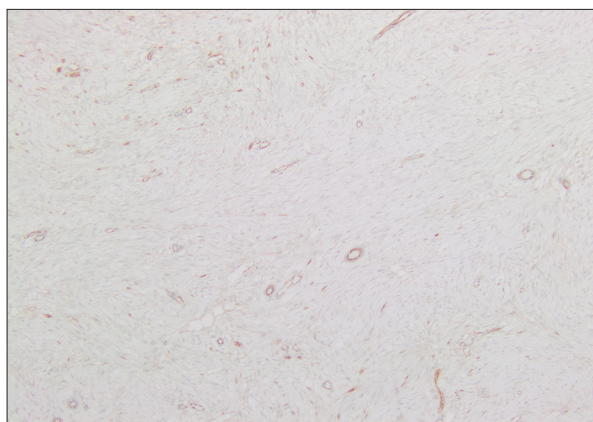


Figure 5. Cytoplasmic positivity of the B-catenin protein in tumoral cells.

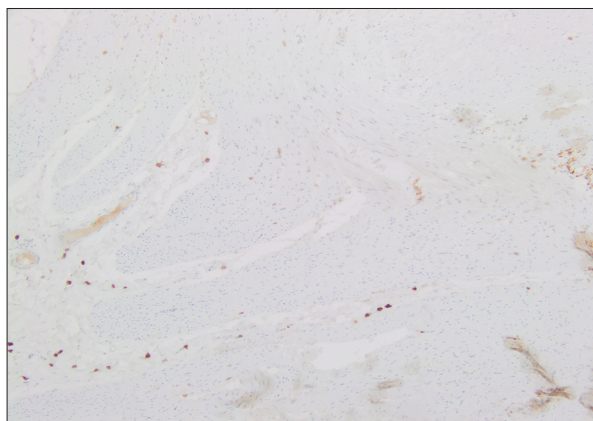


Figure 6. Negative CD117.

loops and the transverse colon. While evaluating the images, no signs of diffusion restriction were noted, whereas diffused enhancement was evident after administering an intravenous injection of gadolinium. Moreover, there was a finding of liquid in the pelvis without any retroperitoneal or mesenteric adenopathies, but rather a 14 mm mesenteric cyst posterior to the umbilicus.

Thus, a surgical approach was considered to be the optimal treatment for this patient's condition. The retroperitoneal mass, as shown in Figure 2, was 10–15 cm from the angle of Treitz and adherent to the bowel.

The gross presentation mimicked a GIST and had clear margins. As a result, jejunal resection and hand sown end to end jejuno-jejunal anastomosis was performed and a retroperitoneal drain was inserted.

After sending the specimen to pathology, the report revealed tumoral proliferation of homogeneously distributed bland spindle cells arranged in long fascicles with elongated nuclei and eosinophilic cytoplasm, as seen in Figures 3 and 4. The stroma

was fibrous and focally myxoid with many thick walled vessels. The tumor was seen to dissect the muscular wall and invade the peri-intestinal fat. The pathology reports for 26 lymph nodes were negative.

The characteristics of this bifocal spindle cell tumor correlated with and were suggestive of mesenteric fibromatosis.

Immunohistochemistry (Figures 3 and 4) showed nuclear and cytoplasmic positivity for β -catenin protein in tumoral cells (Figure 5), and were negative for S100, DOG1, CD117 (Figure 6) and alpha smooth muscle actin, hence confirming the diagnosis of mesenteric fibromatosis.

Discussion

Mesenteric fibromatosis is a fibroblastic growth of the mesentery due to surgical trauma or that may occur spontaneously. Mesenteric fibromatosis of the intestine is the most common site, followed by the omentum and mesocolon [5]. The fourth decade is the typical age of onset of the disease, but there have been cases reported where the disease was found in infancy as intestinal thickening not in the mesentery [6–10]. Usually, familial adenomatous polyposis patients (FAP, Gardner's syndrome) develop more MF [11]; however, this was not true for this case. The patient was previously healthy without a history of surgical or direct abdominal trauma. Ten percent of FAP patients develop desmoids, mainly intra-abdominally. Similarly, fibromatosis associated with FAP is more aggressive and can recur.

Although the clinical course and nature of a mesenteric fibromatosis cannot be predicted, some of them remain stable for years while others may undergo regression. [11–13]

Abdominal pain and bloating are the symptoms with larger MF tumors and these symptoms appear suddenly. Large tumors can also cause weight loss. In our case, the patient was not experiencing abdominal pain and the discovery of the mesenteric fibromatosis was incidental.

Mesenteric fibromatosis is characterized by a high propensity to invade adjacent organs, but with low mitotic activity and no metastatic potential [14]. The excess expression of β -catenin is the result of adenomatous polyposis coli (APC) gene mutations in myofibroblasts [15]. Nevertheless, invading tumors can cause diffused invasion of the intestines and mesentery resulting in occlusion and ischemia.

There are no reported data where the differentiation between GIST and MF is possible when using only radiological tools, even with MRI. Immunohistochemistry is essential for proper diagnosis.

Positive immunohistochemistry with c-kit/CD117 antibody is diagnostic for GIST. If c-kit immunohistochemistry is negative but positive for DOG1, PDGFRA or CD34 genes, we can diagnose gastrointestinal stromal tumor. But if c-kit, DOG1, CD34, PDGFRA, and S100 genes are negative and β -catenin is positive we are able to confirm that the tumor is MF and not GIST.

The management of desmoids has multiple modalities. Surgical resection is the only indication in non-invading tumors. In 53% to 67% of cases, desmoids of the abdomen can be excised [16]. Resection is the first line treatment, while hormonal therapy (for example, tamoxifen: an estrogen receptor blocker), interferon, and NSAIDs (non-steroidal anti-inflammatories) with chemotherapy can play a role in the treatment due to the fact that local recurrence is high, mainly in Gardner's syndrome patients. Others modalities could also be applied especially in patients unfit for surgery due to multiple comorbidities or if the tumor is unresectable [14]. Indomethacin or sulindac (an NSAID) is the first logical step for unresectable tumors, but in this case we treated the patient surgically. If the tumor is still growing, we could treat with tamoxifen with vinblastine and methotrexate (chemotherapy agents that act on cells division and proliferation). Radiotherapy has been shown to have a small role in mesenteric fibromatosis treatment.

As during any abdominal surgery, many complications can occur post op like short bowel syndrome or fistulas between bowel and skin. Fibromatosis tends to recur if incompletely resected because they are locally aggressive tumors. Recent studies showed the same outcome in an R0 and R1 resection of extra-abdominal desmoids [17], thus we treated our patient with an R0 resection. Advanced tumors are best treated with cytotoxic chemotherapy. The risk of radiation enteritis makes radiotherapy less useful and effective in intra-abdominal desmoids. Doxorubicin is the chemotherapy agent most used [18,19]. If there is no response with usual chemotherapy drugs we can use the more recently approved drug imatinib, a tyrosine-kinase inhibitor used in the treatment of multiple cancers with good success [20].

In many cases reported by Bertagnolli et al. [8], the majority of patients with MF had similar radiological results or non-recurrent disease while using a multimodal treatment that combined observation, chemotherapy, and surgery for about 50 months. The proliferation of the tumor usually does not invade past the mesentery, but it could invade the small bowel. MF could invade the muscularis propria or sometimes the submucosa [21,22]. Desmoid tumors encroaching on the bowel wall might be misdiagnosed as primary intestinal tumors, especially gastrointestinal stromal tumors, inflammatory fibroid polyps, or fibrosarcomas [23]. Desmoid tumors are grayish and grossly homogenous on cross section whereas GISTs appears soft and fatty. These tumors can also be differentiated by their areas of

hemorrhage and necrosis. In contrast to MF, GIST depends on the tumor size, with large tumors being more heterogeneous. Thus, a large, firm, homogenous tumor without hemorrhagic and necrotic regions is a great indication of abdominal fibromatosis. The difficulty of distinguishing these two tumor types is due to their similar presentation. Basically, the diagnosis of MF is proven through histological examination and immunohistochemistry. CD117 antigen, grossly in GISTs, can also be present in 75% the cases of MF [23–25]. However, although both tumor types express vimentin, the two types of tumors stain in different ways for smooth muscle desmin and actin. There is no expression of CD34 and S100 protein in MF in contrast to GIST. β -catenin is expressed in fibromatoses guiding the diagnosis in uncertain cases [22–24]. MF may present as a mass-like or infiltrative lesion radiologically [5,26,27]. Nonetheless, a recurrence following resection strongly guides us to a desmoid tumor diagnosis rather than metastasis [23,24,28–31]. Mass-like desmoids are mistaken for other tumors. However, the differentiation between the two tumor types is obligatory for prognosis and treatment [28].

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Conflicts of interest

None.

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