



## Sporadic Mesenteric Fibromatosis Presenting as Intra-Abdominal Sepsis

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### Abstract

We report an unusual case of mesenteric fibromatosis, a desmoid tumor, mimicking a left upper quadrant abscess, presenting as sepsis and colonic pseudo-obstruction. The authors review the presentation, workup and suggested treatment strategies for this rare tumor.

### Case Presentation

A 26 year old woman with a past medical and surgical history remarkable for a cesarean section one year prior presented to the emergency department for the third time in less than one week with severe left upper quadrant abdominal and left lower chest pain associated with shortness of breath. She had been discharged from a recent hospitalization with a diagnosis of constipation for similar complaints two days earlier. On arrival to the emergency room, the patient was afebrile, hypotensive (systolic blood pressure was 76 mmHg), and tachycardic (heart rate of 119). She reported having severe left upper quadrant abdominal and left lower chest pain that began 1 week prior, associated with nausea and vomiting. She had peritonitis on physical examination. Chest x-ray on admission showed the patient to have bilateral pleural effusions, but no other abnormalities were noted. Labs were unrevealing aside from leukopenia (white blood cell count 2.0 K/CMM, 75.1% neutrophils), lactic acidosis (3.6mmol/L), and hypoalbuminemia (2.7 g/dL).

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Review of previous imaging obtained including computed tomography (CT) of the chest, abdomen and pelvis revealed an approximately 8 cm round intra-abdominal mass in the left upper quadrant abutting the diaphragm, stomach, colon and spleen (Figure 1). Due to the patient's presenting symptoms and the corresponding findings seen on CT, the patient was emergently taken to the operating room for a diagnostic laparoscopy with presumptive suspicion for a left upper quadrant abscess versus mass of questionable etiology.

Upon entering the abdomen, ascites was noted in the right upper quadrant. The spleen was visualized and noted to be well adherent to a left upper quadrant mass. The omentum was subsequently dissected off of the mass. The capsule of the mass was noted to be very firm. A portion of this tissue was resected and a frozen section was concerning for a possible spindle cell neoplasm. Upon conversion to formal laparotomy, approximately 200-250 mL of purulent fluid was suctioned from the pelvis. The mass was resected en bloc, requiring partial colectomy (splenic flexure) and splenectomy (Figure 2). A primary colocolostomy was performed. The patient's post-operative course was prolonged secondary to delayed return of bowel function as well as poor pulmonary toilet secondary to sympathetic left pleural effusion (requiring thoracentesis x 2). She had no infectious complications.

Final pathology revealed a 10.3 x 8.8 x 7.0 cm mass consisting of bland spindle cell proliferation without necrosis. Mitoses were noted to be 0 per 50 high power fields. The tumor cells were found to be positive for vimentin, beta catenin, and CD10 and negative for pancytokeratin, S100, HMB45, CD34, smooth muscle actin, desmin, GFAP, estrogen receptor, CD117, and DOG-1. Testing for c-KIT and PDGFR mutations were performed, in order to completely exclude a gastrointestinal stromal tumor, and was found to be negative. Three mesenteric lymph nodes were sampled and were negative for any abnormality. These overall features were consistent with mesenteric fibromatosis.

Post-operatively, the patient has done well. Of significance, the patient has no significant family



**Figure 1:** Computed tomography (CT) of the abdomen revealed a large mass (arrow) in the left upper quadrant abutting the spleen, splenic flexure, diaphragm and the stomach.

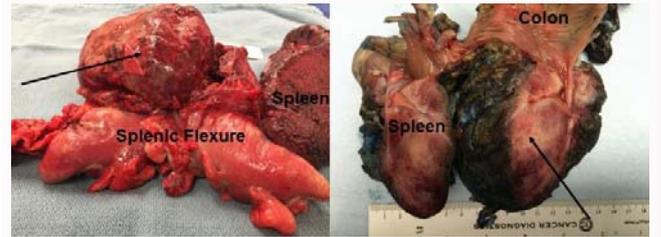
history of polyposis syndromes or colon/gastrointestinal cancer. Genetic testing for familial adenomatous polyposis (FAP) was negative.

## Discussion

Mesenteric fibromatosis, also known as desmoid tumors, are rare soft-tissue tumors, characterized by deregulated fibroblastic proliferation. They are usually slow growing and locally aggressive. Although they lack the capacity to metastasize, they are associated with a high rate of recurrence after resection [1]. Desmoid tumors account for roughly 0.03% of neoplasms and are not known to have any significant racial or ethnic predilection [2]. However, some studies have shown that there is a slight increase incidence for desmoid tumors in women [3]. This may be due in part to the potential influence of estrogen on the local environment, since studies have linked higher incidences of desmoid tumors in pregnant women and women using oral contraceptives [1]. The fact that estrogen plays a role in the development of desmoid tumors has further been strengthened by the fact that spontaneous regression of desmoid tumors have been noted in women undergoing menopause [4]. Desmoid tumors have also been linked to patients who have undergone surgery or experienced soft tissue trauma [5].

Desmoid tumors have been reported to affect all sites of the body, however, they are generally classified into three main anatomic locations: extra-abdominal, intra-abdominal, and along the abdominal wall [6]. Although there is an association between desmoid tumors and FAP, the majority of desmoid tumors arise sporadically [7]. It is important to note that the most common location for FAP associated desmoid tumors is intra-abdominal, while sporadic desmoid tumors are more commonly found deep in the shoulder girdle, the extremities, and the buttock region [8]. Desmoid tumors that arise intra-abdominally most commonly originate from the small bowel mesentery, thus they have been routinely named mesenteric fibromatosis or mesenteric desmoid tumors in the literature [9]. One important risk factor for the development of intra-abdominal mesenteric desmoid tumors is a history of abdominal surgery. This risk factor is especially linked to patients who have been diagnosed with FAP, specifically those patients who have the Gardner's syndrome variant [9,10]. Although a history of trauma has also been linked to the development of desmoid tumors, it is usually associated with the extra-abdominal variant. One study showed that a history of trauma was seen in 19-63% of patients who developed extra-abdominal desmoid tumors [11]. Typically, desmoid tumors are isolated single tumors; nonetheless, patients with multiple desmoid tumors have been reported, specifically in patients with Gardner's syndrome [12].

Most patients with desmoid tumors are clinically asymptomatic;



**Figure 2:** Photographs of the specimens revealed a large mass arising from the splenic flexure of the colon, consistent with mesenteric fibromatosis on final pathology. Demonstrated is the excised specimen as well as cross section demonstrating the mass to be arising from the splenic flexure of the colon.

however, patients who present with symptomatic mesenteric desmoid tumors usually present late [13]. Patients who present with intra-abdominal desmoids usually present with symptoms that can be attributed to the tumor's mass effect [14]. These symptoms include a palpable abdominal mass, abdominal pain, vomiting, weight loss, fever, malaise, small bowel obstruction, small bowel perforation, gastrointestinal bleeding, peritonitis, and ureteric obstruction [15-17]. Desmoid tumors have unpredictable clinical courses that range from small, asymptomatic tumors that remain static to small tumors that rapidly enlarge causing severe disability and possible death within months or years. The morbidity and mortality associated with intraabdominal desmoid tumors are directly related to the tumor's location, and therefore, the abutting structures the tumor may infiltrate or compress [18].

Mesenteric desmoid tumors have variable radiological features which make them difficult to diagnose with imaging alone [19]. Desmoid tumors can appear as either ill or well circumscribed masses with variable attenuation and contrast enhancement when viewed on CT and magnetic resonance imaging (MRI). Most mesenteric desmoid tumors present as large (>15cm in diameter) abdominal masses that have a generally higher attenuation, when compared to muscle, on contrast enhanced CT [19]. MRI has been described as the imaging modality of choice in patients suspected to have a mesenteric desmoid tumor. MRI typically reveals masses with a low signal intensity on T1 weighted images and variable signal intensity on T2 weighted images when comparing its attenuation to muscle [20,21]. Ultrasound is another imaging modality that has been used in the clinical work up of desmoid tumors. Although it does not have the multi planar capabilities of both MRI and CT, it is an important tool in the evaluation of abdominal discomfort, one of the common complaints seen in patients presenting with mesenteric desmoid tumors [15-17]. Like MRI and CT, desmoid tumors can have multiple presentations when seen on sonography, ranging from smooth, well circumscribed masses with a high, medium, or low echo texture, to ill-defined masses with multiple echo textures [22].

The most specific and conclusive diagnostic test for desmoid tumors is a tissue biopsy [23]. The pathophysiology of desmoid tumor formation has been linked to excess production of the protein Beta-catenin [24]. This excess production is believed to be caused by different mutations in the APC gene and/or the Beta-catenin (CTNNB1) gene [25]. In one case series, done by Huss et al, 91.6% of all desmoid tumors showed nuclear expression of Beta-catenin [26]. Those desmoid tumors that were shown to not express Beta-catenin either carried a CTNNB1 mutation or had an association with patients diagnosed with Gardner's syndrome [26]. It is good practice to test all patients diagnosed with a desmoid tumor for mutations in the

APC gene, even though APC mutations are uncommon in sporadic desmoid tumors [27,28]. This is due to the fact that patients with APC gene mutations have an increased risk of colorectal cancer [28]. Similarly, Wang et al. proposed that all pediatric patients diagnosed with a desmoid tumor should be referred for FAP evaluation, APC gene mutation testing, and genetic counseling [29].

The treatment of choice for desmoid tumors is surgical excision with wide margins [30]. However, wide margins may not always be possible given that some desmoid tumors are inoperable due to their large size or their involvement with vital structures. Another common problem seen when treating mesenteric desmoid tumors with surgical excision is the not insignificant recurrence rate (25-50%) [31]. Neoadjuvant radiotherapy has been utilized prior to surgical excision in order to shrink tumors to an operable size and to reduce the recurrence rate [31]. For patients who are not good surgical candidates or for those with local recurrence, cytotoxic pharmaceutical agents, estrogen receptor antagonists, non-steroidal anti-inflammatory drugs, and tyrosine kinase inhibitors have been shown to have some efficacy in the medical management of desmoid tumors [32-36]. However, these agents are not considered first line therapy since only a small number of patients have been treated with these agents, and the responses seen from these agents have been poorly documented [37].

Because of the high rate of recurrence, despite the achievement of histopathological negative margins during surgical resection, long term follow up is essential in the post-operative management of desmoid tumors. The median time to tumor recurrence has been found to be around 23 months, with more than 90% of recurrences evident by 5 years [37]. Currently, there are no evidence based protocols for tumor surveillance following surgical treatment. However, the National Comprehensive Cancer Network suggests completing a history and physical, with appropriate imaging, every 3-6 months for the first three years. After the first 3 years of post-operative follow up, an annual history and physical with appropriate imaging is sufficient for surveillance of tumor recurrence [38].

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