Mesenteric fibromatosis, or intra-abdominal desmoid tumor, is a rare proliferative disease affecting the mesentery that can pose a diagnostic and therapeutic challenge. It is a locally aggressive tumor that lacks metastatic potential but often recurs locally. Mesenteric fibromatosis with intestinal involvement can be easily confused with other primary gastrointestinal tumors, especially with that of the mesenchymal origin. We report a case of a 46-year-old male with no significant medical or surgical history, complaining of left side abdominal pain and fever for 3 days. The patient was thoroughly investigated and contrast-enhanced CT abdomen revealed a well-defined mass over the left upper abdomen near the splenic flexure. Exploratory laparotomy was underwent and a mass measuring 5.6 cm in diameter arising from the descending mesocolon was identified. Complete removal was accomplished with no need of small bowel or colon resection. Histopathological examination showed mesenteric fibromatosis. Postoperatively, patient was well and 3-month followup showed normal recovery.
identified over left upper abdominal quadrant above the level of kidney (Fig. 2). Subsequent exploratory laparotomy was performed which revealed a greyish white, firm, and well-circumscribed mass adherent to the descending mesocolon at the splenic flexure. The characteristics of the mass allowed its complete removal with no need of small bowel or colon resection. Postoperative recovery was uneventful, and there was no evidence of residual or local recurrent tumor at 3-month follow-up abdomen CT study.

The microscopic examination of the resected specimen showed a picture of fibromatosis characterized by fibroblasts dispersed throughout dense collagen. Hemorrhage and chronic inflammatory cell infiltrations are also noted at the interface of the mass and peripheral tissues. At immunohistochemical study, the cells stained positive for beta-catenin but negative for CD117, CD34, and S100 protein. Final histological diagnosis of mesenteric fibromatosis was thus established.

**Figure 1.**

1a

1b

1c

**Figure 1.** a. Non-contrast axial section computed tomographic (CT) scan shows a well-circumscribed solid mass (arrow) located in the left abdominal quadrant near the splenic flexure. A gallstone is also noted. b. Contrast-enhanced axial section CT scan shows a mixed-attenuation mass (arrow) in the left abdominal quadrant near the splenic flexure with well-defined borders and a small regional lymph node (arrowhead) nearby. c. Contrast-enhanced coronal section CT scan shows a mixed-attenuation mass (arrow) in the region of descending mesocolon.

**Figure 2.**

**Figure 2.** Oblique sonogram of the left upper abdomen shows a well-defined mass with mixed echotexture above the left kidney.
DISCUSSION

The fibromatoses are a broad group of histologically similar fibroblastic proliferations, which contain spindle-shaped myofibroblastic cells, dense deposits of intercellular collagen fibers, variable amounts of extracellular myxoid matrix, and compressed and elongated vessels [6]. They demonstrate infiltrative growth and local recurrence but do not metastasize. Fibromatoses are rare, accounting for 0.03% of all tumors.

The fibromatoses can affect both superficial and deep parts of the body. Superficial fibromatosis involves the face and neck (fibromatosis coli), palms (Dupuytren's contracture), feet (Ledderhose's disease), penis (Peyronie's disease), shoulder, thigh, buttock and trunk [7]. The deep fibromatoses arise within the deep soft tissues and are usually divided into abdominal wall, extraabdominal, and intraabdominal types, which has been reported with a relative frequency of 49%, 43%, and 8% respectively.

Intraabdominal fibromatosis can occur in the pelvis, mesentery, and retroperitoneum [8]. The small bowel mesentery is the most common location for intraabdominal fibromatosis. Consequently, the terms mesenteric fibromatosis or mesenteric desmoid tumor are most often used interchangeably with intraabdominal fibromatosis. However, other mesenteric structures within the abdomen, such as the omentum, transverse or sigmoid mesocolon, and ligamentum teres, may be the site of origin for intraabdominal fibromatosis.

MF is also the most common primary tumor of the mesentery [5]. MF can occur in a wide age range of patients, ranging from 14 to 75 years of age with a peak incidence at 30 years old, and has no gender or race predilection [2]. The etiology of MF is still unclear. MF demonstrates sporadic occurrence, but the incidence is increased in patients with familial adenomatous polyposis (FAP), especially the Gardner syndrome variant of FAP [5]; trauma [9]; previous abdominal surgery [10, 11]; or hormonal stimulation [12-14]. Patients with Gardener's syndrome are estimated to have an 850 times increased risk for fibromatosis compared to the general population [15, 16]. Prior abdominal surgery is an important risk factor for the development of MF in patients with FAP. Eighty-three percent of patients with both FAP and MF have a history of abdominal surgery, most commonly a total colectomy [17]. In contrast, only ten percent of patients with sporadically occurring MF have previously undergone abdominal surgery [18].

The majority of patients with MF remain clinically asymptomatic, with little or no focal symptoms until later in their course, at which stage they complain of abdominal pain or discomfort, constipation, vomiting, and organ compression symptoms, which is related to size and location, such as small bowel obstruction and hydronephrosis. Otherwise, MF may be an unexpectedly incidental finding during abdominal exploration or imaging study for other causes.

Imaging remains the mainstay of preoperative investigations to establish a working diagnosis of mesenteric fibromatosis [19], but the definitive diagnosis is confirmed only after the histological analysis of the tumor [2]. Plain radiographs may show evidence of mass effect with displacement of adjacent bowel loops or a prominent soft-tissue mass centered within the abdomen. Calcification is uncommon. Colon may be involved occasionally, especially when the site of origin is the mesocolon [5]. The sonographic appearance of MF is a solid, well-circumscribed mass of variable echotexture and homogeneity, which is nonspecific and chiefly dependent on the collagen and fibroblast content within the tumor as well as the intralesional vascularity [20, 21]. The CT and magnetic resonance imaging (MRI) appearances of MF are directly related to its underlying histologic characteristics and vascularity, and CT is frequently the modality of choice for detection and followup.

On CT, MF usually demonstrates a soft tissue density and attenuation directly related to the underlying histology with radiating strands projecting into the adjacent mesenteric fat. Collagenous stroma predominant MF will be homogeneous with soft-tissue attenuation, while myxoid component predominant MF will result in a hypoattenuation density and do not enhance with intravenous contrast material. When a MF has both collagenous and myxoid stroma, it may appear striated or whorled because of the alternating collagenous and myxoid areas. The blood supply and contrast enhancement of MF are very variable but more frequently show little contrast material enhancement on CT scans [5].

On MRI, MF are low or intermediate signal intensity on T1-weighted images due to their major fibrous composition and have heterogeneously intermediate or high signal intensity on T2-weighted images. The relative amount of hyperintensity on T2-weighted images reflects the degree of high cellularity areas and myxoid stroma within. The intravenous contrast enhancement pattern is variable, similar to the pattern observed at CT [8]. MF that do not significantly enhance on CT scans have been shown to enhance with intravenous gadolinium on MR imaging [22, 23].

On the basis of imaging findings alone, MF is difficult to differentiate from GIST and malignant neoplasms of the mesentery, such as lymphoma, metastatic disease, and soft-tissue sarcoma. MF may closely resemble lymphoma on CT and MRI whenever the lesions manifest with well-defined margins, homogeneous attenuation, and involvement of adjacent small bowel segments [5]. GISTs typically contain areas of hemorrhage and necrosis, which manifest as focal areas of hypoattenuation [24]. The immunohistochemical profile of MF is helpful for differentiating it from a GIST. In contrast to GISTs, MF does not express CD34 and S100 protein. CD117 antigen, which is expressed commonly in GISTs, can be positive in up to 75% cases of MF [4]. Nuclear [beta]–catenin that MF expresses is also
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a distinctive characteristic that may be of diagnostic value [25, 26]. As radiological findings are indefinite, a surgical approach is required for diagnostic purposes to take a biopsy by a laparoscopy or by a laparatomy, resection of the tumor if possible, or a palliative surgery in a symptomatic patient with unresectable tumors [27].

The management of patients with MF is controversial. Complete resection with negative microscopic margins, when feasible, is the standard care for most MF [28, 29]. Surgery for patients with sporadically occurring MF is frequently curative. Nevertheless, local recurrences are frequent especially in cases of MF associated with Gardner’s syndrome [1], and complications such as small bowel obstruction and fistula formation may sometimes develop. Therefore, in some cases, reexcision and postoperative irradiation, endocrine therapy, or chemotherapy may be necessary [5].

In summary, we present a rare case of mesenteric fibromatosis presenting as a soft tissue mass over the left upper abdominal quadrant mesentery without definite direct involvement of the small bowel visible on imaging studies. Mesenteric fibromatosis is usually asymptomatic and presents with a variety of clinical features. The diagnosis of mesenteric fibromatosis is difficult to establish preoperatively, especially in patients without a significant medical or surgical history. Treating physicians should always be suspicious and include such a differential diagnosis when encountering a mesenchymal tumor that diffusely infiltrate the mesentery.

REFERENCES
