



Sporadic Mesenteric Fibromatosis with Colonic Entrapment Mimicking Gastrointestinal Stromal Tumor—A Rare Entity: Two Case Reports

Thara Pratap¹ Dhanya Shibi² Pushpa Mahadevan³ Muhammed Jasim Abdul Jalal⁴ Amritha Aravind²

¹Department of Radiodiagnosis, VPS Lakeshore, Kochi, Kerala, India

²Department of Radiology, VPS Lakeshore, Kochi, Kerala, India

³Department of Pathology, VPS Lakeshore, Kochi, Kerala, India

⁴Department of Internal Medicine and Rheumatology, Olive Healthcare, Thrissur, Kerala, India

Address for correspondence Muhammed Jasim Abdul Jalal, MBBS, DNB(Family Medicine), MNAMS, MRCP(UK), MRCP(UK), EULAR Certification in Rheumatology, Department of Internal Medicine and Rheumatology, Olive Healthcare, Thrissur 680614, Kerala, India (e-mail: jasimabduljalal@yahoo.com).

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Mesenteric fibromatosis (MF), also known as desmoid tumors of the mesentery, is rare, locally aggressive fibroblastic proliferation of mesentery that tends to recur locally without distant metastasis. The clinical behavior of desmoids is unpredictable varying from indolent to aggressive. Radiologically, mesenteric fibromatosis could have infiltrative margin or well-defined, lobulated margin at presentation. The commonest site of mesenteric fibromatosis is small bowel mesentery.

Though the diagnosis of MF with infiltrative margins is rather easy, mesenteric fibromatosis with well-defined margins can be confused radiologically with gastrointestinal stromal tumor (GIST).^{1,2} Histopathology confirmation is essential for a definitive diagnosis.

We, herein, present two cases with well-defined margins mimicking GIST arising from small bowel mesentery with entrapped segment of colonic loop within the lesion.

Case 1

A 42-year-old patient presented with acute onset abdominal pain with a palpable mass in the epigastrium. Ultrasound revealed heterogeneous density soft tissue mass with lobulated margin in the epigastric region. Contrast-enhanced computed tomography (CECT) was done (► Fig. 1A–D) that showed an approximately 10.6 × 9.0 × 10.08 cm (anteroposterior (AP) × transverse (TR) × craniocaudal (CC)) well-marginated, heterogeneous mass with mild enhancement in the infracolic compartment contiguous with the mid-transverse colon with a segment of colon entrapped within the lesion seen as a linear contrast filled area in the periphery. Streaky densities were noted extending to small bowel mesentery. There was no evidence of local infiltration to other adjacent structures. A

diagnosis of GIST was considered. The patient underwent transverse colectomy.

Peroperatively, the mass was arising from small bowel mesentery and was infiltrating a segment of mid-transverse colon. Gross section showed a nodular mass attached to serosal aspect of colon and jejunum. Histopathology and immunohistochemistry (IHC) confirmed mesenteric fibromatosis. Histopathology showed that the lesion was composed of diffuse sheets of spindle shaped cells with variable cellularity intervened by collagenous stroma and was seen involving the muscularis propria and serosa of the colon in one area. No significant mitotic activity was seen. Ki index was less than 1%. Follow-up done up to 6 years did not show any evidence of recurrence.

Case 2

A 27-year-old male patient presented with a palpable mass in the right iliac fossa. Ultrasound showed a homogenous density right iliac fossa mass and CECT revealed (► Fig. 2A–C) large lobulated homogeneously mildly enhancing solid lesion involving the right iliac fossa, closely related to the distal ileal loops and ileocecal junction, measuring approximately 9.6 × 11 × 11 cm (AP × TR × CC). Significant luminal narrowing was noted in the distal ileum likely due to mass effect. A segment of cecum was seen entrapped within the mass seen as a linear contrast-filled streak extending into the lateral aspect of the mass from the cecal base. Right hemicolectomy was done.

Gross section showed a mass attached to the ileocolic mesentery. Histopathology showed a neoplasm infiltrating the muscularis propria and perimuscular fatty tissue of the

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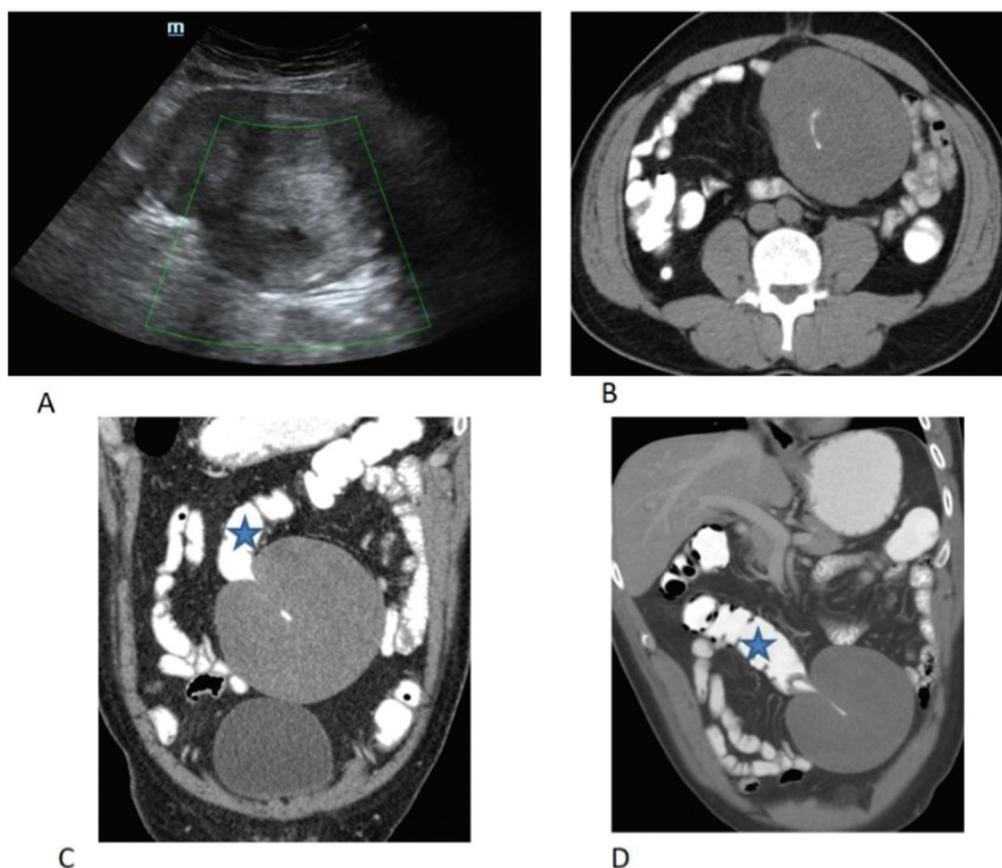


Fig. 1 Ultrasound (A) image showing heterogeneous mass in the epigastrium. Plain axial (B) showing well-margined mass in the abdomen at the level of umbilicus. Postcontrast coronal and oblique (C, D) images showing well-margined fairly homogenous mass contiguous with the transverse colon (asterisk). Tethered long segment of transverse colon seen entrapped within the lesion in Fig. D.

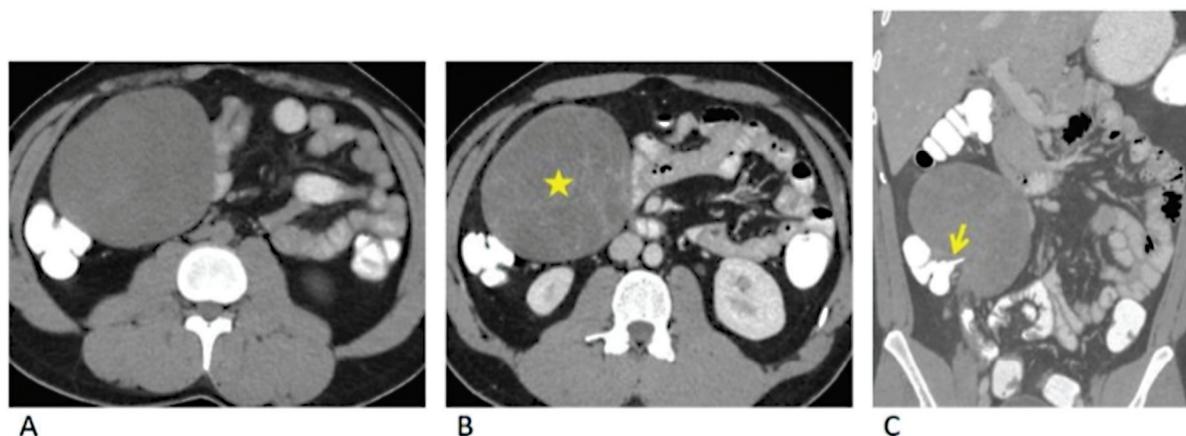


Fig. 2 Plain (A), axial contrast-enhanced computed tomography (CECT) (B), and coronal CECT (C) images showing moderate sized well-margined mass in the right iliac fossa region. Postcontrast axial CT showing mild heterogeneous enhancement (asterisk). Fig. (C) showing a tethered segment of cecum within the lesion (arrows).

cecum. The tumor cells were arranged in fascicles and bundles. The stroma showed myxoid components. Focal areas of increased mitosis 6 to 8/10 high power field were seen. Findings were suggestive of spindle cell neoplasm with focal increase in mitotic activity. IHC confirmed mesenteric fibromatosis (–Fig. 3A, B). IHC showed positivity for H-caldesmon with focal positivity for β -catenin and CD 10. Ki 67 index was 6 to 8%.

The term “desmoid” was coined by Muller in 1838 and is derived from the Greek work “desmos” meaning band or tendon. Desmoid tumors are rare mesenchymal neoplasms and constitute approximately 3.5% of all fibrous tissue tumors.³ Desmoid is the most common primary tumor of the mesentery.

Desmoid tumors usually affect individuals in the 15 to 60 age group with peak incidence at 35 to 40 age group.^{4,5} Most

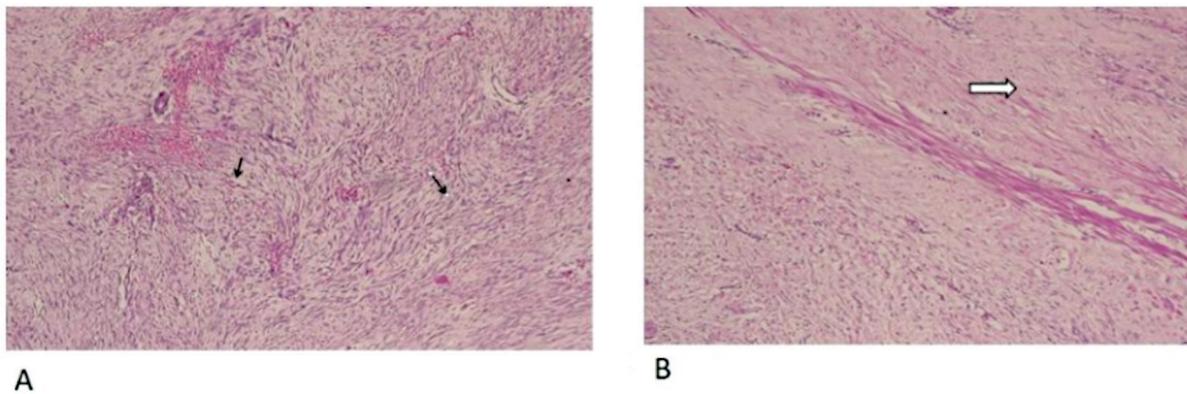


Fig. 3 (A) Hematoxylin and eosin low power stain with cellular areas showing spindle cells (arrows) arranged in fascicles and bundles. (B) Tumor with hypocellular areas (arrow).

of them appear in women in their reproductive age group mostly during pregnancy.³ They are usually solitary and large at presentation; in 10 to 15% of the cases they are multiple.⁶ Estrogen receptors are present in 33% of desmoid tumors.^{3,7}

According to anatomic location, mesenteric fibromatosis is classified into intra-abdominal (mesenteric, pelvic, and retroperitoneal fibromatosis), abdominal wall (abdominal fibromatosis), and extra-abdominal fibromatosis. Among the intra-abdominal sites, the small bowel mesentery is the most common site of origin. Besides small bowel mesentery, mesenteric fibromatosis can also involve omentum, ileocolic mesentery, transverse, sigmoid mesocolon, and ligamentum teres.⁸

Mesenteric fibromatosis could be primary and secondary. Primary is rare, lesions are sporadic, and are mostly extra-abdominal. Only 5% of sporadic desmoid tumors are intra-abdominal in location.⁶ Secondary desmoids occur due to trauma, hormonal stimulation, previous abdominal surgery or is associated with familial adenomatous polyposis (FAP) or Gardner's syndrome.³ Those associated with FAP have APC gene mutation that leads to overexpression of nuclear β -catenin.² Mesenteric fibromatosis occurs in 9 to 18% of patients with FAP.³ In patients with FAP, the lesions are smaller and multiple.^{9,10}

The clinical behavior of mesenteric fibromatosis is variable with some indolent tumors demonstrating spontaneous regression. Usual presentation is with large palpable tumor causing abdominal discomfort or pain, GI bleeding, and perforation. Morbidity results from local complications like small bowel obstruction, ischemia, perforation,¹¹ and fistula formation. Hydronephrosis can occur due to periureteric infiltration.¹² Rates of local recurrence following resection can be high.¹³ Distant metastasis is rare.

Pathology

Desmoid fibromatosis is a locally aggressive, nonmetastasizing myofibroblastic neoplasm with an infiltrative growth pattern with whorled cut surface in gross specimen. Microscopically it shows long sweeping fascicles of slender spindle cells with hypocellular areas showing myxoid background, abundant keloidal fibers, or zones of hyalinization. Vascularity is variably prominent. Extensive soft tissue infiltration

can be seen. With immunohistochemistry, the cells are positive for smooth muscle actin and occasionally desmin. Nuclear expression of β -catenin is observed in 80% cases.¹⁴

Imaging

On ultrasound, desmoid tumors have variable echogenicity and Doppler characteristics. Ultrasound has a limited role for characterization of mesenteric masses.

CT is considered as the primary imaging modality for diagnosis of intra-abdominal desmoid tumor. CT scans show mesenteric desmoid tumors often appearing as soft tissue masses with ill-defined margins and radiating spicules extending into the adjacent mesenteric fat or as a well-demarcated lesion. Mass-like and infiltrative pattern can coexist. The lesion shows variable enhancement in postcontrast study. Myxoid stroma appears more hypodense, whereas highly collagenous stroma demonstrates soft tissue density enhancement. CT can identify the location, the multiplicity, and relationship with mesenteric vessels and intra-abdominal organs as well as associated complications. Entrapment of bowel loops is described in mesenteric fibromatosis⁹ as in our cases.

Magnetic resonance imaging (MRI) is considered if CT is contraindicated. MRI often shows heterogeneous pattern, with iso- to hypodense signal on T2-weighted sequence. Low signal intensity linear bands may be seen corresponding to dense collagenous stroma. Necrosis is very rare even in large tumors. The signal characteristics of desmoids on MRI depend on the amount of collagenous, myxoid stroma, and extracellular matrix.^{12,15}

Desmoid tumors show variable uptake pattern in positron emission tomography-computed tomography ranging from low-to-moderate grade.

Metastatic carcinoid tumor and sclerosing mesenteritis are differentials for mesenteric fibromatosis with infiltrative margins.

Mesenteric desmoids fibromatosis with well-defined margins have to be distinguished from other mesenteric lesions like GIST, lymphoma, soft tissue sarcomas, neurogenic and inflammatory myofibroblastic tumors. GISTs are usually well-defined lobulated lesions which arise from the bowel and infiltrate the mesentery. They rarely originate in the mesentery. The average

age group for occurrence of GIST is above 50 years. Small tumors show homogenous, intense enhancement. When large, the tumor is often heterogeneous with intralesional hemorrhage and central necrosis.¹⁰ At times, the cystic necrosis would predominate and GIST would appear as cystic lesions with peripheral solid rim enhancement with central fluid attenuation. GIST can occur anywhere along the GI tract, most common in the stomach and small bowel. Lack of necrosis in mesenteric fibromatosis is a differentiating feature from GIST.

In case 1, the site of origin of lesion was from the small bowel mesentery encasing antimesenteric border of mid-transverse colon. In case 2, the lesion was arising from ileocolic mesentery with focal increase in mitotic activity. Although the mitotic rate is not usually high in fibromatosis, scattered mitotic figures may be seen. The presence of diffusely elevated mitotic rate should prompt regular follow-up to rule out recurrence.¹⁶

Both our cases were sporadic without any associated causes. Entrapment of colonic loops was seen in both cases, which would indicate chances of perforation if left untreated.

Moderate-to-large-sized desmoid tumors with well-defined margins are often treated by surgical resection.¹⁴ Mesenteric fibromatosis associated with FAP often has infiltrative margins encasing the mesenteric vessels and hence surgical resection may be incomplete. Other treatment modalities include anti-inflammatory agents, antiestrogen, targeted therapy as well as radiation therapy, though efficacy is unpredictable. In spite of radical surgery and adjuvant radiotherapy, 25 to 50% cases recur.³ Radiotherapy is considered only if the lesion is inoperable or progressive in spite of other treatment modalities. Regarding follow-up after surgery, there is no clear guideline for postoperative surveillance¹⁴ as the recurrence rates are variable.

The diagnosis of mesenteric fibromatosis represents a challenge for the radiologist. Though it is rather easy to make a diagnosis of mesenteric fibromatosis with stellate margin, GIST and other soft tissue sarcomas should be considered in differentials of desmoid with well-defined margins. Lack of necrosis in spite of the large size is a differentiating feature from GIST. Entrapment of bowel wall within the lesion may give a clue to diagnosis. Definitive diagnosis requires histopathology confirmation. Local recurrence rate is high; hence, close follow-up is recommended.

Conflict of Interest

None declared.

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