

Quiz

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A 29-year-old female presented with gradually developing, generalized vague abdominal pain since 3 months. There were no associated constitutional symptoms. Her abdominal examination revealed a firm, well-circumscribed, non-tender and mobile mass filling all quadrants of the abdomen. The initial laboratory work-up was normal while the images of the contrast-enhanced CT scan are illustrated in Fig. 1. The patient underwent exploratory laparotomy and a mass was removed. The final histological picture of the mass is shown in Fig. 1.

What is the most probable diagnosis?



Figure 1:

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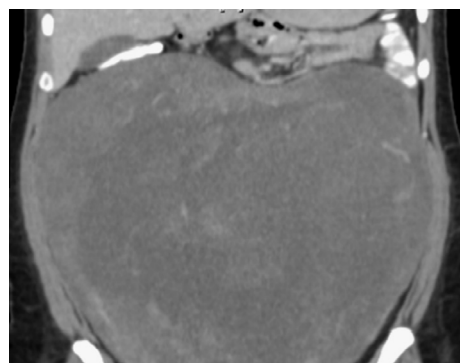


Figure 2:



Figure 3:

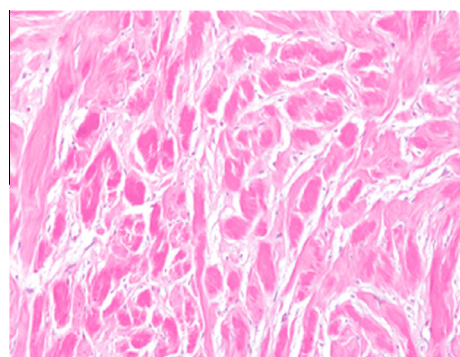


Figure 4:

There are diverse causes of vague abdominal pain including inflammatory, neoplastic, traumatic, and obstructive factors. Out of the neoplastic etiologies, the benign tumors often pose diagnostic challenges to the treating physicians. Mesenteric fibromatosis is one of the uncommon clinical entities which is rarely reported in the literature with a wide range of presentations including pain and discomfort, constipation, vomiting, weight loss and organ compression symptoms, such as small bowel obstruction and hydronephrosis.¹ Various etiopathogenetic theories have been coined but the exact nature of the causative agent still remains unknown. Trauma², infectious etiology due to its association with retroperitoneal fibromatosis³, and genetic predisposition may play a role in the development of this disease. The differential diagnosis often includes gastrointestinal stromal tumors and less commonly fibrosarcomas or inflammatory fibroid polyps.⁴

The microscopic examination of mesenteric fibromatosis shows minimally positive desmin, vimentin positive, S100 minimally focally positive, cd34 negative, SMA focally positive and MSA largely negative, and Cd117 positive. All these tumor markers are pathognomic of mesenteric fibromatosis.⁵ The mitotic count is relatively low with no evidence of necrosis and nuclear dedifferentiation. On CT scan, fibromatosis may present as a solitary or multiple, well-defined or ill-defined mass with attenuation similar to muscles. A whorled appearance, with radiating mesenteric strands, displacement, retraction and/or compression of the bowel loops, has also been described.⁶ Surgical excision by taking wide margins of the normal surrounding tissue remains the mainstay of treatment. Large tumor size (larger than 10 cm), multiplicity, extensive tumor invasion, encasement of small bowel loops and entrapment of the ureter have been reported as poor prognostic factors in familial adenomatous polyposis patients with mesen-

teric fibromatosis.⁷ For extensive infiltrative tumors, various nonsurgical treatments have been tried; estrogen receptor antagonist, tamoxifen; nonsteroidal antiinflammatory agents; aromatase inhibitor therapy; chemotherapy with dactinomycin, vincristine, and cyclophosphamide; and radiotherapy, singly or in combination.⁸

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