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A giant mesenteric fibromatosis adherent to the appendix and colonic wall, case report

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ABSTRACT

BACKGROUND: Mesenteric fibromatosis, is a rare neoplasm arising usually from the bowel mesentery, with intermediate behavior and local invasion potential. They can be sporadic or related to multiple factors contributions.

They usually presents as an asymptomatic growth of intraabdominal mass, and can reach a large diameter before symptoms appearance. Surgical excision is the definitive treatment when achievable.

CASE PRESENTATION: In this case we present a case of 34 years old gentleman, presenting for painless abdominal distension, and found to have a giant mesenteric fibromatosis of 23 cm diameter and 4.5 kg arising from the appendix and colonic mesentery. Treated surgically, and was free of recurrence after 1 year follow up.

CONCLUSION: We report this rare case to encourage physicians to keep this etiology in mind as part of the differential diagnosis of unspecific abdominal mass.

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1. Background

Mesenteric fibromatosis is a rare neoplasm taking part of the fibromatosis entities, which rise usually from the retroperitoneum or small bowel mesentery, and can be as big as 10 cm before having symptoms. We present a rare case of giant mesenteric fibromatosis arising from appendix and colon mesentery, and reaching 23 cm in diameter. This case was reported in accordance with the SCARE criteria [1].

2. Case history

A 34 years old gentleman, previously healthy, with a surgical history of left inguinal hernia repair (6 years prior to presentation), presented to out clinics with a complaint of abdominal distention

and hair loss. On physical examination, abdomen was soft, no tenderness, a palpable soft mass noticed in the right abdominal middle and inferior quadrant.

Ultrasound of abdomen and pelvis, showed large intra-peritoneal mass of 24 cm long axis, with heterogeneous echotexture, extending from the supraumbilicus area to the pelvis, and associated with 2 cystic structure at its base, with posterior enhancement, the largest measure 10.6 × 3.8 cm (Fig. 1).

A contrast enhanced CT Scan of abdomen and pelvis done, confirming the presence of the large intraabdominal mass, of tissue density (33HU), measuring 26 × 25 × 12 cm and occupying the region extending from the renal arteries origin to the supra bladder. It presents a well-defined borders, without infiltration of the surrounding adipose tissue, and pushing the intestines to the periphery. Three other structures are identified at the left infero-lateral part of the mass, non-enhancing post contrast injection, and are of cystic appearance. No evidence of invasion of major vascular axes. The liver is homogeneous and free of detectable lesions (Fig. 2).

Patient admitted for elective surgery, done under general anesthesia and in supine position. Midline laparotomy incision realized, extending from sub-xiphoid to supra-pubic region. Identification of a very bulky mass of around 30 cm in diameter fixated in the right iliac fossa, adherent to the great omentum by large vessels, coagulated using vessel sealing electro-surgical cautery device (Fig. 3).

Abbreviations: CT, Computed Tomography; FAP, Familial Adenomatous Polyposis; GIA, Gastro-Intestinal Anastomosis; GIST, Gastro-Intestinal Stromal Tumor; HIFU, High-Intensity Focused Ultrasound; MRI, Magnetic Resonance Imaging.

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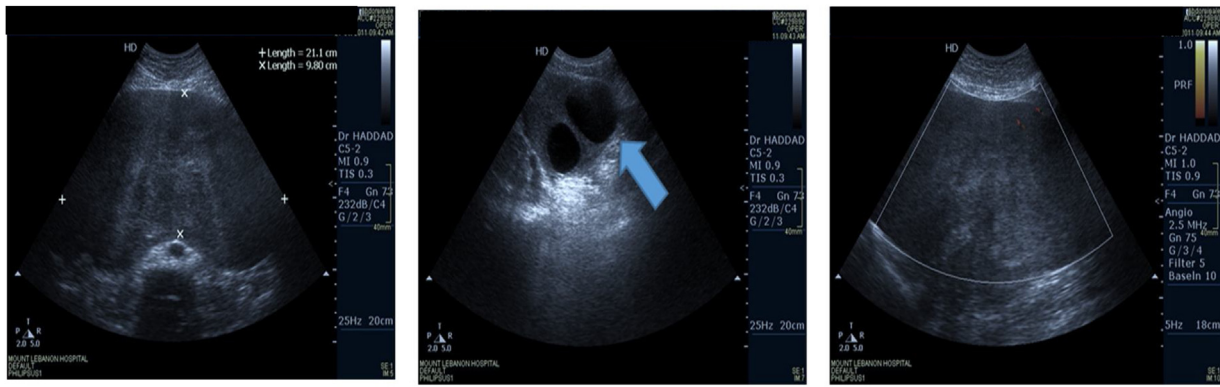


Fig. 1. Ultrasound: Slightly hypoechoic mass with some cystic area (Arrow).

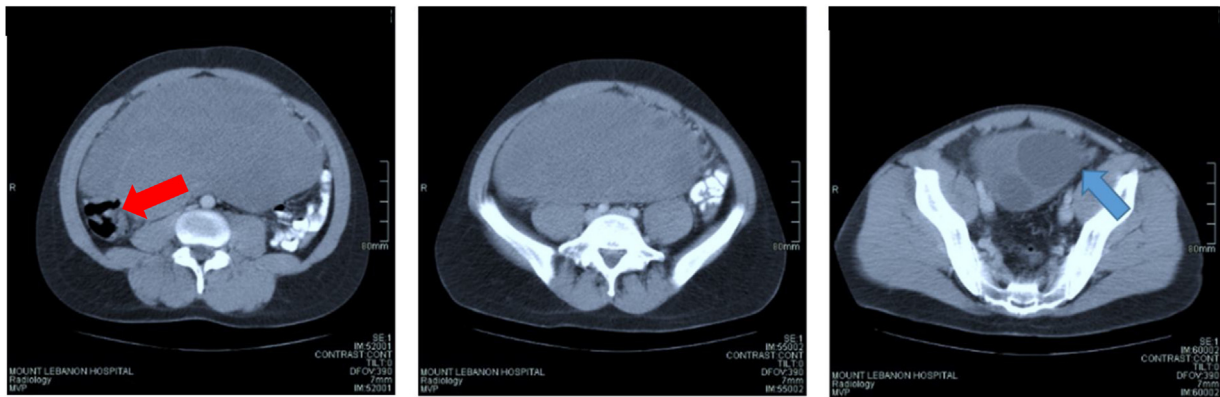


Fig. 2. CT Scan: Slightly isodense mass, adherent to ascending bowel (Red Arrow), with some cystic areas (Blue Arrow).

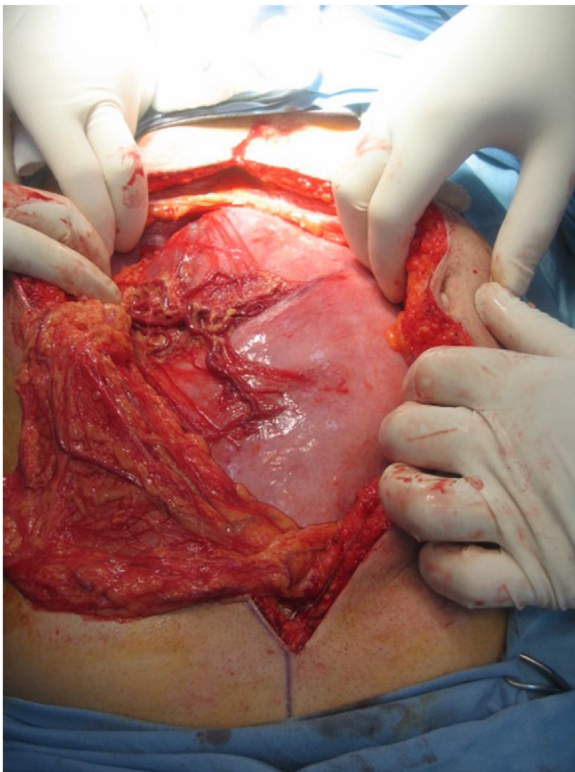


Fig. 3. Intraoperative Image: Bulky mass adherent to the great omentum by large vessels.

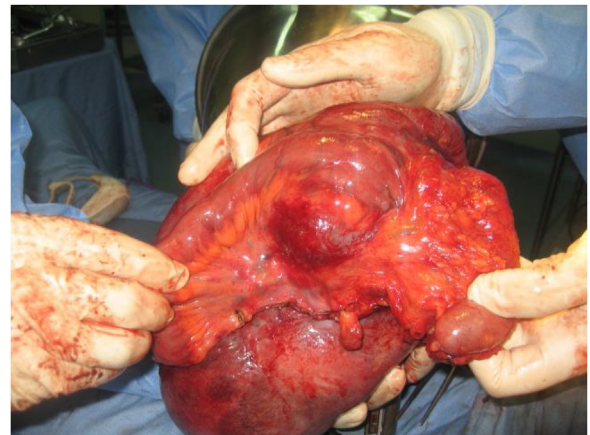


Fig. 4. Intraoperative Image: Removal of the mass 'en bloc' with terminal ileum and colon.

Identification and preservation of the iliac vessels and the right ureter. The mass was adherent to the cecum and the last intestinal loop without clear cleavage plane. Ileocecoappendicular pedicle ligated. Section of the small intestine at 15 cm proximal to the tumoral invasion, and of the colon at the level of the hepatic angle using GIA at both sides. Removal of the mass 'en bloc' (Figs. 4 and 5). Hand sewn ileo-transverse, termino-lateral anastomosis.

The patient tolerated the surgery well and was discharged home on the sixth day following surgery without major complications.

The pathology report revealed that the mass grossly was a nodular tumor of 23 × 23 × 16 cm and 4.5Kg, was found attached to the colonic wall. The tumor had a white, fasciculated, largely

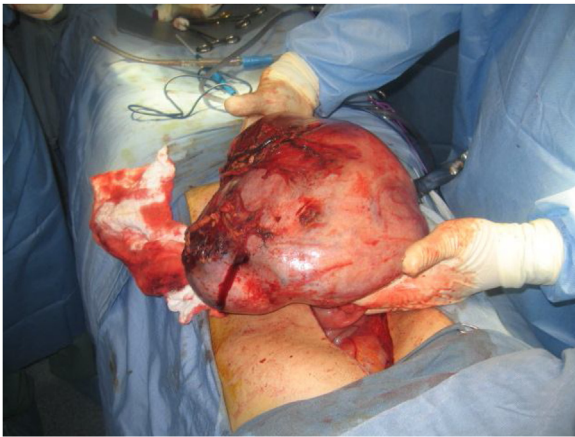


Fig. 5. Intraoperative Image: Removal of the mass 'en bloc'.

myxoid and focally cystic cut surface. Microscopic examination showed fascicles of spindle cells adherent to the wall of the cecum and appendix without mucosal invasion (Fig. 6 and 7). Rare mitotic figures were found. The stroma is myxoid and highly vascularized (Fig. 8). No lymph node invasion. Immunohistochemical study revealed negativity for anti-CD117, anti-CD34 and anti-S100 excluding gastro-intestinal stromal tumor (GIST), solitary fibrous tumor and nerve sheath tumors. Ki-67 stain showed a low mitotic index (<2%). All these findings were compatible with mesenteric fibromatosis adherent to colon and appendix wall.

After a 1 year follow up with an enhanced Thoraco-Abdomino-Pelvis CT scan, the patient haven't developed a recurring tumor and no intra-abdominal lesion (Fig. 9).

3. Discussion

Mesenteric fibromatosis (MF), is a rare tumor, derived from the mesenchymal tissue [2], they are considered a proliferation of fibroblasts and myofibroblasts [3] with uniformly bland nuclear features [4]. It have an intermediate behavior between fibrosarcomas and benign fibrous neoplasm [3], This behavior is characterized

by local aggressive invasion, but without metastasizing potential [3,2].

MF accounts for 0.03% of all neoplasms and 3% of soft tissue tumours [5] and 8% of fibromatosis [2], which rarely arise from the mesentery and abdominal organs, but instead they often arises from the extremities or the abdominal wall [4]. MF is considered as the most common primary tumor of the mesentery arises most frequently in the retroperitoneum or in the small bowel mesentery, but they rarely involve the omentum and mesocolon [3]. They occur especially in the 3rd and 4th decades, but they can arise in all ages [3]. It can be sporadic or hereditary as in the case of Gardner's syndrome [2], that why patients diagnosed with intra-abdominal fibromatosis should be screened for familial adenomatous polyposis (FAP) endoscopically [6].

Multiple factor contribute to the development of these tumors, such as trauma, especially surgical trauma (the surgical resection of MF itself can be this kind of trauma leading to another formation and recurrence), genetic predisposition, autoimmune, infectious and hormones mainly estrogen [3] (multiple studies showed the positivity of estrogen receptor in 90% of cases [5]).

Patients usually presents with painless abdominal mass [2] and they are commonly large around 10 cm at presentation [5]. Symptoms are caused by the compression and infiltration of the adjacent organs as the tumor develop [3] and according to their size and location, they can progress to an ischemia, bowel obstruction [2], small bowel fistula formation, gastrointestinal bleeding and abscess formation [6].

It is essential to set the proper diagnosis using multimodalities imaging, helping in the identification of the location of the mass and its extent to the surrounding structures [3] and eliminating the differential diagnosis which may consist of lymphoma, fibrosarcoma, GIST, carcinoid tumor [2]. Ultrasound findings are non-specific, with variable vascularity, variable solid appearance and posterior acoustic enhancement in 75% of cases [5]. On CT lesions are mostly isodense to skeletal muscle and is usually seen as a mass in the mesentery of soft-tissue and with projection of radiating strands into the adjacent mesenteric fat [5]. MRI is essential in accurate evaluation of the surrounding structures and their relation to the tumor, and can be used as well as CT for follow up [5].

An ultrasound or CT guided Needle aspiration cytology is the only preoperative qualitative technique, but it less used due to

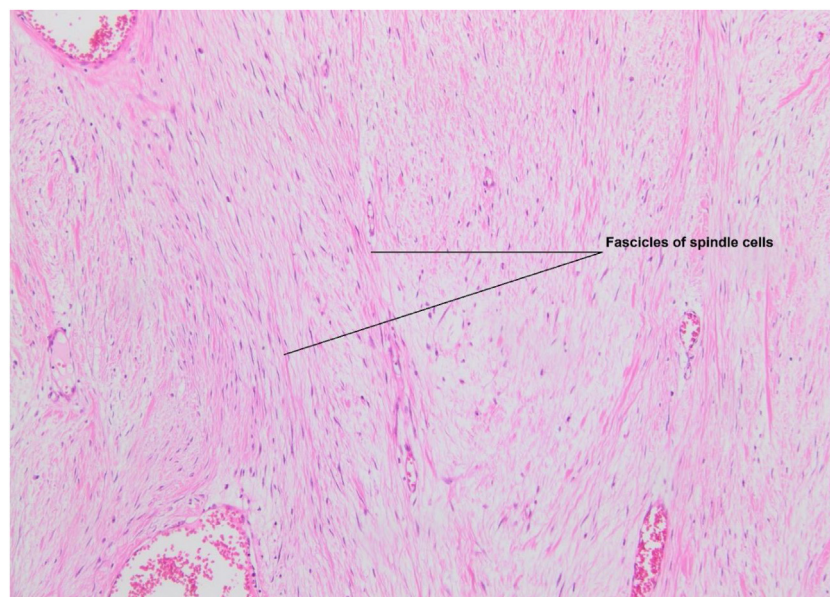


Fig. 6. Fibromatosis showing fascicles of spindle cells. ($\times 1000$).

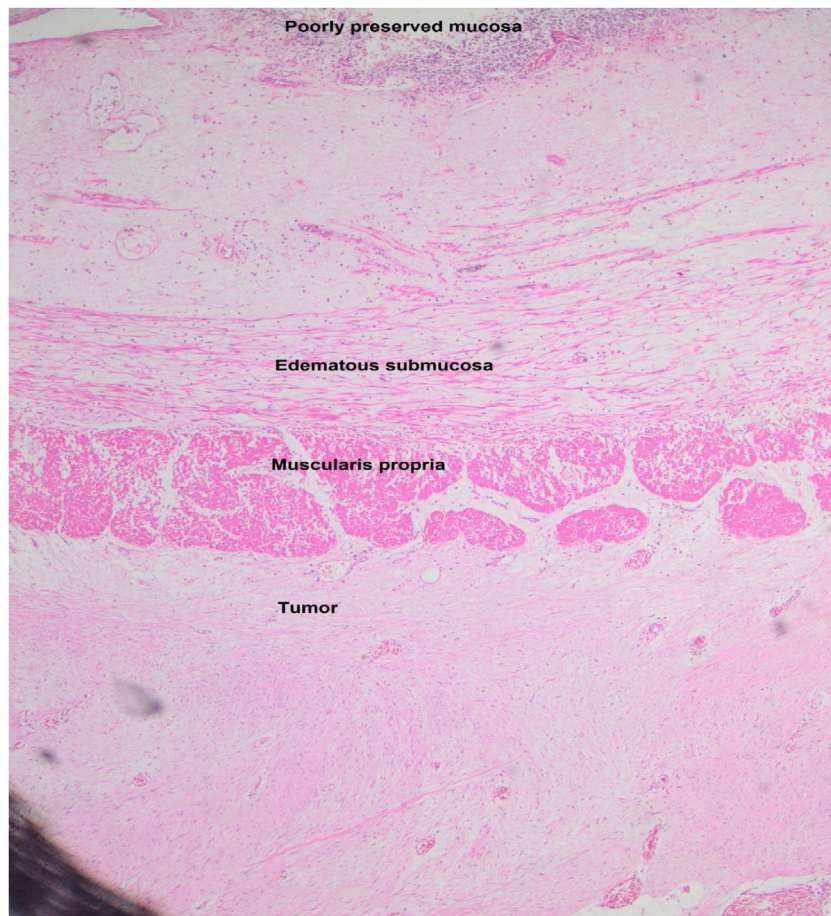


Fig. 7. Fibromatosis adherent to the appendicular wall. ($\times 400$).

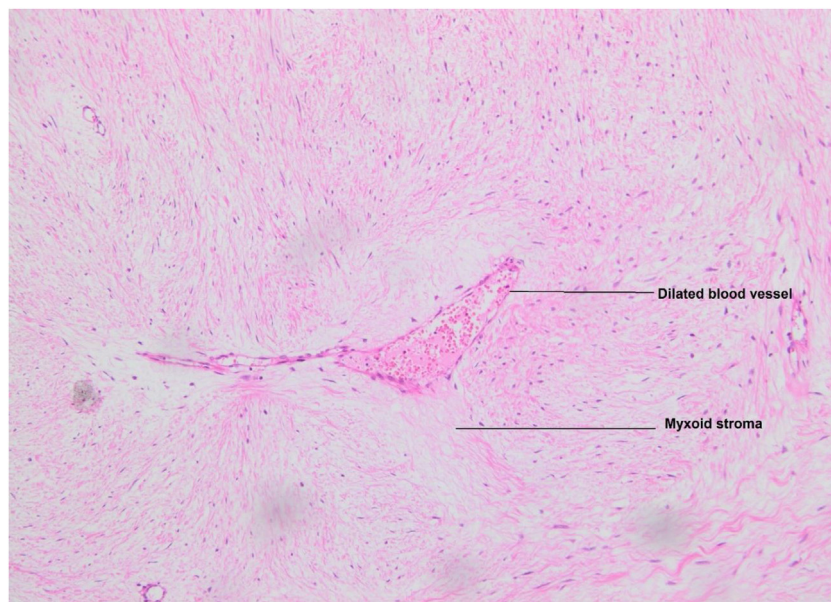


Fig. 8. Fibromatosis, vascularized and myxoid stroma ($\times 1000$).

its corresponding complications, such as bleeding, perforation and seeding [2]. It is always contraindicated to do an excisional biopsy [5]. The gold standard in diagnosis is the final pathology result [3] confirmed by immunohistochemistry examining mainly β -catenin, CD34, β -catenin, KIT⁺ and DOG1 [2]. When the following findings

are present: mass size > 5 cm, mass growing in size, deep mass, and pain, we have high suspicion of sarcoma [5].

The definitive treatment is surgical resection when possible [2]. These tumor grow fast in a asymptomatic fashion, so they are usually late diagnosed and are no more suitable for complete resection



Fig. 9. 1 year CT scanner follow-up shows no intra-abdominal lesion.

[3] they often invade the mesenteric vessels and the root of the mesentery, menacing the bowels vascularization, thus complete resection of the tumor may lead to small bowel syndrome [3]. When complete surgical excision or recurrence exists, radiotherapy may be implicated [2]. Other alternative choices may include molecular target therapy, Chemotherapy, hormonal manipulation and Nonsteroidal inflammatory drugs [2], and ultrasound-guided high-intensity focused ultrasound (HIFU) ablation [3].

But unfortunately they usually recur, Recurrence rate is affected by the location of the tumor, age of patient, and the negative margins achievement during resection [3]. In contrast, in cases where we have association with FAP, we may have higher recurrence rate after radical surgical resection [3].

New approach “wait-and-see”, is getting more popular, since some tumors may unexpectedly decrease or disappear, and may have sustained free of pain disease [5].

To our knowledge, this is the first documented case of mesenteric fibromatosis, adherent to the wall and arising from the mesentery of the ascending colon and appendix.

Declaration of Competing Interest

This article has no conflict of interest with any parties.

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Ethical approval

The study type is exempt from ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Writing the paper: Etienne El-Helou, Jessica Naccour, Mariana Zaarour.

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