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MESENTERIC FIBROMATOSIS WITH INTESTINAL INVOLVEMENT MIMICKING A GASTROINTESTINAL STROMAL TUMOR

Surgery

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ABSTRACT

Introduction: Mesenteric fibromatosis or intra-abdominal desmoid tumor is a rare proliferative disease affecting the mesentery. It is a locally aggressive tumor that lacks metastatic potential, but the local recurrence is common. Mesenteric fibromatosis with the intestinal involvement can be easily confused with other primary gastrointestinal tumors, especially with that of the mesenchymal origin.

Case report: We report a case of a 30-year-old female who presented with an abdominal mass that radiologically and pathologically mimicked a gastrointestinal stromal tumor.

KEYWORDS

mesenteric fibromatosis, desmoid tumor, gastrointestinal stromal tumor, GIST, differential diagnosis.

Introduction:

Mesenteric fibromatosis (MF) or intra-abdominal desmoid tumor is a rare proliferative disease affecting the mesentery. MF is a locally aggressive tumour that lacks metastatic potential, but the local recurrence is common. It resembles gastrointestinal stromal tumors (GIST) that are mesenchymal neoplasms of the digestive tract and show a varied malignant potential. Although GISTs and mesenteric fibromatosis are distinct entities, they are often confused clinically, radiologically and not uncommonly pathologically as well. Misdiagnosis might result in inappropriate therapeutic decisions and worse prognosis.We report a case of a 30-year-old female who presented with an abdominal mass that initially suggested a gastrointestinal stromal tumor.

Case report:

A 30 -year-old female was admitted to our department because of dull aching abdominal pain for 3 days and on self-examination she noticed lump in abdomen since 3 days. Her past medical history was not significant.

The physical examination revealed a mass on palpation in the mid-abdomen that was easily movable. The physical examination was otherwise normal. Laboratory findings were unremarkable. The level of CEA was within normal limits. A transabdominal ultrasound (US) showed an ovoid well-delineated homogenously hypoechoic mass that was 10.1 × 6.0 × 7.2 cm in size. There was a hyperechoic area in the central part of the tumor with posterior acoustic shadowing that corresponded to intraluminal air. The tumor was circumferentially attached to the wall of the small bowel. An abdominal computed tomography (CT) revealed a 8.2 × 7.2 × 7.4 cm mass infiltrating the small bowel. The tumor attenuation was enhanced poorly and homogenously with an intravenous contrast (Figure 1).The above preoperative imaging studies suggested mesenchymal origin most likely a GIST involving the small bowel.

Figure 1

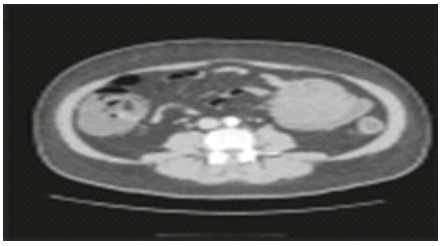


Figure 2

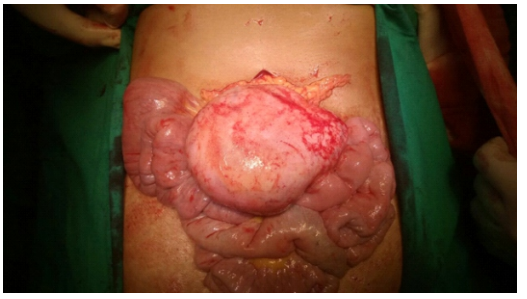
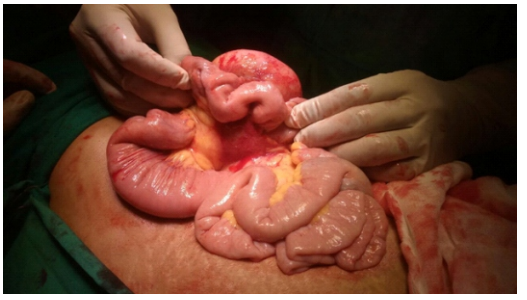


Figure 3



CT scan shows the desmoid tumor of the mesentery infiltrating the small bowel: a well-defined hypodense and homogenous mass diffusely attached to the bowel wall.

The patient underwent an elective laparotomy. Intraoperatively, there was an approximately 10 cm well-circumscribed mass in the mesentery that infiltrated the wall of the small bowel along with duodenum at DJ JUNCTION which narrowed its lumen(Figure 3). This gross appearance suggested a gastrointestinal stromal tumour without peritoneal dissemination. The resection was avoided at that moment because to its infiltration at DJ junction, and proceeding with resection may lead to high morbity .So biopsy was taken to rule out GIST, followed by definitive treatment. The postoperative course was uneventful and the patient was discharged in a good health condition. A follow-up biopsy realved Desmoid fibromatosis.

The primary pathological diagnosis in this particular case was a CD 117-negative gastrointestinal stromal tumor. The microscopic

examination of the biopsy specimen identified a fibromatosis in the mesentery. Histologically it was desmoid tumor that composed of spindle cells with elongated coma-shaped nuclei and the immunohistochemistry was negative for both CD 117 and for CD 34. Beta-catenin overexpression was present on immunohistochemistry. No mitoses were found in 50 high power fields and no sarcomatous transformation.

Discussion:

Mesenteric fibromatosis is a type of fibroblastic proliferation affecting the mesentery that develops usually as a consequence of surgical trauma, but it may occur spontaneously. Patients with familial adenomatous polyposis (FAP, Gardner's syndrome) are especially predisposed to the development of mesenteric fibromatosis.1 Desmoids develop in approximately 10% of FAP patients and most are intra-abdominal. Similarily, fibromatoses associated with FAP follow a more aggressive course and the recurrence after the resection is common.2

The pathogenesis of fibromatoses has been unclear for many years. Currently, these tumors are regarded as a clonal proliferation of myofibroblasts that show APC (adenomatous polyposis coli) gene mutations. These mutations lead to the over expression of beta-catenin.1,3,4.

The clinical behaviour and the natural course of mesenteric fibromatoses are unpredictable. Some desmoid tumors remain stable for years and several cases of the spontaneous tumor regression without any treatment have been reported.5 Nevertheless, a progressive and invading tumor can result in a diffuse infiltration of the mesentery and bowel leading to intestinal ischemia or to the obstruction. The treatment modality in mesenteric fibromatoses is still controversial. The results of the treatment might be biased due to the unpredictable course of this disease with some tumors regressing or remaining stable without any treatment. The management of desmoids should be individualized and multimodal. The surgical resection should be performed only in localized tumors that do not invade the root of the mesentery. Intra-abdominal desmoids can be resected in 53–67% of cases.6 The aggressive surgical treatment of mesenteric desmoids may result in short bowel syndrome or multiple enterocutaneous fistulas requiring a long-term parenteral nutrition. Fibromatoses are locally aggressive tumors that tend to recur when incompletely resected. A high rate of recurrence after the surgical resection results from the incomplete resection, multicentric disease or surgical trauma as a new precipitating factor. Recent studies report comparable recurrence rates after R0 and R1 resection in extra-abdominal desmoids.7,8 Nevertheless, there are no data to support a similar influence of microscopically positive margins on recurrence in intra-abdominal desmoids.

Advanced and unresectable tumors or when the resection will result in short bowel syndrome are best treated within a clinical trial of cytotoxic chemotherapy or other experimental therapies. Radiotherapy is rarely used in intra-abdominal desmoids because of a high risk of radiation enteritis. There are no established chemotherapy regimens used in fibromatoses. Most chemotherapeutic protocols use doxorubicin.9 Recently, a successful therapy of a desmoid tumor resistant to traditional chemotherapeutic regimens was reported with imatinib, a tyrosine kinase inhibitor that is successfully used in advanced gastrointestinal stromal tumours.10,11.

In a series reported by Bertagnolli *et al.*9, 96% of patients with mesenteric desmoids had either a radiographically stable disease or no recurrence for a median of 50 months using a multimodal treatment combining watchful waiting, surgery and chemotherapy.

The growth of desmoid tumors is usually limited to the mesentery, but the infiltration of the adjacent bowel is not uncommon. Mesenteric Fibromatosis may infiltrate the muscularis propria or even the submucosa.12 The diagnosis of mesenteric fibromatosis is usually straightforward in the cases without a concomitant intestinal involvement. On the other hand, desmoid tumors encroaching the bowel wall present a diagnostic challenge and might be easily confused with primary intestinal tumors, especially gastrointestinal stromal tumors. Difficulties in differentiating these two tumors of distinct pathogenesis, natural course and prognosis are still not uncommon.12-14.

Desmoid tumors are firm masses. On cross section these tumors are grayish and grossly homogenous. In comparison, GISTs are usually soft and fleshy. On cut surface, these tumors commonly have areas of necrosis and haemorrhage. In contrast to MF, the gross appearance of GISTs is highly dependant on their size, with large tumors being morphologically more heterogeneous. It follows that a large tumor that is firm and homogenous on cross section without obvious haemorrhagic and necrotic areas is highly suggestive of intra-abdominal fibromatosis. Sometimes, differentiating these two distinct tumors is difficult because of a possible clinical, macroscopic and even histological overlap. Nevertheless, the diagnosis of mesenteric fibromatosis is based on the microscopic examination and immunostaining. It is noteworthy that a CD117 antigen, expressed commonly in GISTs, can be positive in up to 75% cases of mesenteric fibromatosis.11-13 Moreover, both MF and GISTs express vimentin and stain variably for smooth muscle actin and desmin. In contrast to GISTs, MF does not express CD34 and S100 protein. Recently, the expression of beta-catenin was revealed in fibromatoses that might prove helpful in the differential diagnosis in doubtful cases.1,15,16

Radiographically, mesenteric fibromatosis may present as a mass-like or infiltrative lesion.17 Infiltrative desmoid tumors image as an ill-defined whorled thickening of the mesentery and are usually easily recognized. Mass-like desmoids are more challenging. These desmoids appear as well-defined tumors and are often confused with other primary neoplasms, especially gastrointestinal stromal tumors. Nevertheless, the distinction between these two tumors is important because of vital prognostic and therapeutic implications. On the other hand, it should be remembered that a recurrent mass after oncological operations may prove to be an intra-abdominal desmoid, and not necessarily a metastasis. Lee *et al.*22 reported a case of intraabdominal fibromatosis that occurred after the resection of a gastric stromal tumor. A similar case of intra-abdominal fibromatosis suspicious of local recurrence was reported after gastrectomy for gastric cancer.23 Failure to differentiate mesenteric fibromatosis from other tumors may lead to an inappropriate treatment and a worse prognosis.

Conclusions:

The diagnosis of mesenteric fibromatosis should always be considered in the case of mesenchymal tumors apparently originating from the bowel wall that diffusely infiltrate the mesentery.

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