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Case Report

Mesenteric Fibromatosis Simulating a Gastrointestinal Stromal Tumor: A Rare Cause of Intestinal Obstruction

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Abstract

Mesenteric fibromatosis also known as mesenteric desmoids is a rare benign mesenchymal lesion that can occur throughout the gastrointestinal tract, especially small bowel. Because of its biological behavior, mesenteric fibromatosis is often confused with submucosal tumor or malignant neoplasm of gastrointestinal tract, mainlygastrointestinalstromaltumors (GIST). Often asymptomatic or by incidental finding, these tumors can be revealed by signs and symptoms related to the small bowel such as abdominal pain or a palpable abdominal mass, or clinical complications like gastrointestinal bleeding, small bowel obstruction, fistula formation, or bowel perforation. The authors report a rare case of small bowel obstruction secondary to sporadic synchronous intra-abdominal fibromatosis in a 61-years old man who complained for a palpable mass at left lower quadrant of his abdomen and which was initially misinterpreted as a GIST on the CT. A laparotomy was planed, but the patient complained for abdominal pain with a stop of flatus and stools which required emergency hospitalization. En-bloc resection of the tumour with the adjacent jejunum was performed followed by end-to-end anastomosis. The operative course was unremarkable. The diagnosis was revised as mesenteric fibromatosis according to pathologic examination. Bowel obstruction is a rarely revealing complication of mesenteric fibromatosis. Clinical manifestations at a complicated stage require emergency surgery in suitable candidates. In this case, emergency tumor excision made it possible to avoid complications from intestinal obstruction and to correct the first diagnosis, which was initially a GIST.

Keywords: Anastomosis, Surgical; Benign Neoplasm; Fibromatosis, Abdominal; Intestinal Obstruction; Margins of Excision; Mesentery

Introduction

Mesenteric fibromatosis is a rare sporadic mesenchymal neoplasm of the small bowel that arises from myofibroblasts. It is a histologically benign disease and lacks the capacity to metastasis [1,2]. It can involve any site of gastrointestinal tract, but the most common site is the mesentery of the small bowel. Its biological behavior is intermediate between benign fibrous tissue proliferation and fibrosarcoma [3].They are often found incidentally but can be symptomatic or even revealed by digestive, vascular, and urological complications [4]. We report the case of a 61-year-old man, scheduled for the surgical removal of a palpable mass of the left lower quadrant of his abdomen, behaving like a GIST on CT, but who in the meantime has presented an acute bowel obstruction. Emergency laparotomy was performed. Postoperative course was uneventful. Pathologic diagnosis was mesenteric fibromatosis. Although GIST and mesenteric fibromatosis are completely different entities, their clinical, radiological and histological characteristics have many similarities which can confuse clinicians [5,6]. Through the description of this case, the important role of surgery in confirming the diagnosis and preventing complications will be emphasized.

Case presentation

A 61-year-old man was admitted for constipation associated with abdominal pain in the lower quadrant evolving for a week. He had no specific family history and no previous surgical intervention. Physical examination revealed hyperactive bowel sound and palpable mass at left lower quadrant of abdomen. Initial complete blood cell counts and blood chemistry results were unremarkable. Abdominal CT scan had found a mass developing at the expense of the small intestine measuring approximately 15cm x 10cm whose morphological characteristics evoked a stromal tumor (Figure 1).

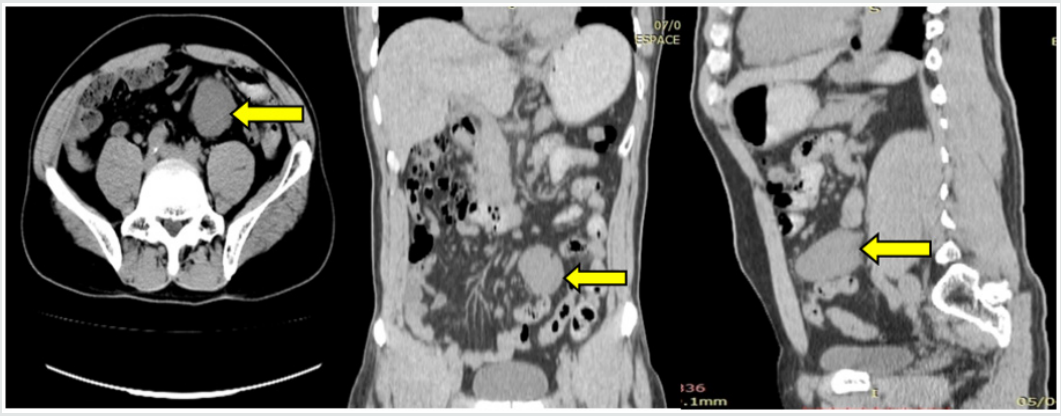

Figure 1: Axial and coronal, sagittal section showing the mesenteric tumor.

A surgical approach was planned in order to confirm the pathological diagnostic bu tbefore the scheduled date, the patient complained of diffuse abdominal pain with a stop of his bowel function. An emergency laparotomy was performed. The lesion was close to superior mesenteric vessels. Resection of the small intestine involved by the tumor was performed followed by manual end to end anastomosis (Figure 2).

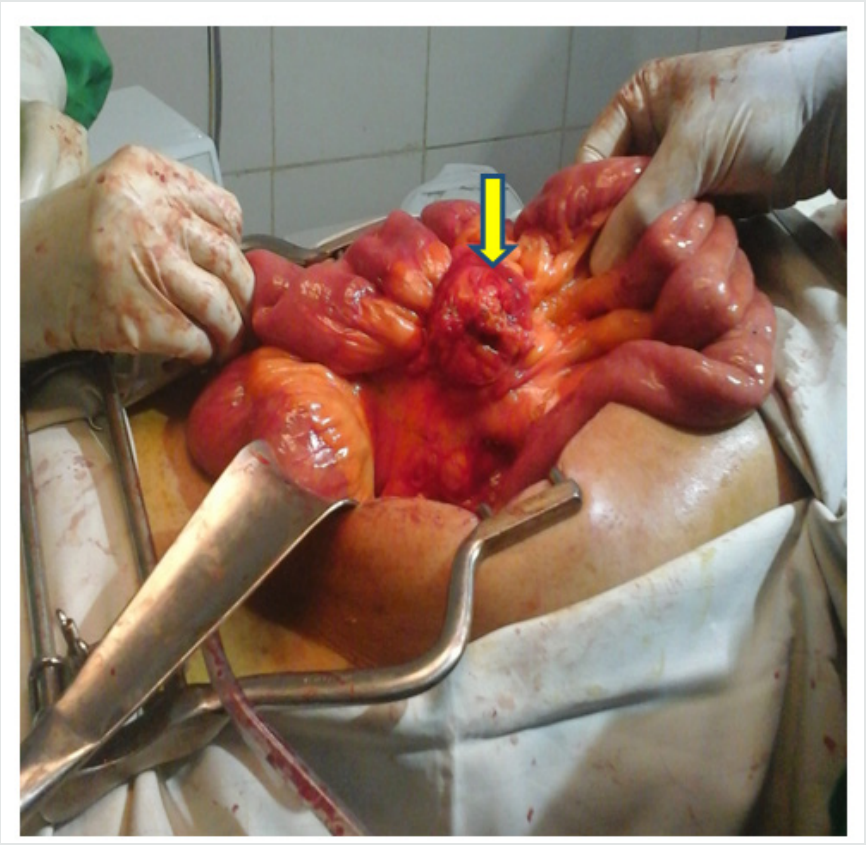

Figure 2: Operative view: The small intestine adjacent to the tumor was invaded on its antimesenteric side explaining the acute intestinal obstruction.

The surgical specimen consisted of an angulated segment of the small intestine, which was 35 cm long, and had a hard ovoid nodule, measuring 10 x 15cm, in the mesentery (Figure 3). Microscopic examination revealed a homogeneous proliferation of mesenchymal spindle cells arranged in bundles, with low mitotic index, no evidence of necrosis, and few hypercellular areas. There was no invasion of surgical margins. The microscopic study shows orderly arrangement of uniform fibroblasts associated with moderate amounts of collagen (Figure 4) compatible with a mesenteric fibromatosis. The postoperative course was uneventful, and the patient was allowed to go home on the seventh postoperative day. No adjuvant therapy has been performed. After 12 months of follow-up, the patient was in very good general condition with no sign of recurrence on clinical examination and on imaging.

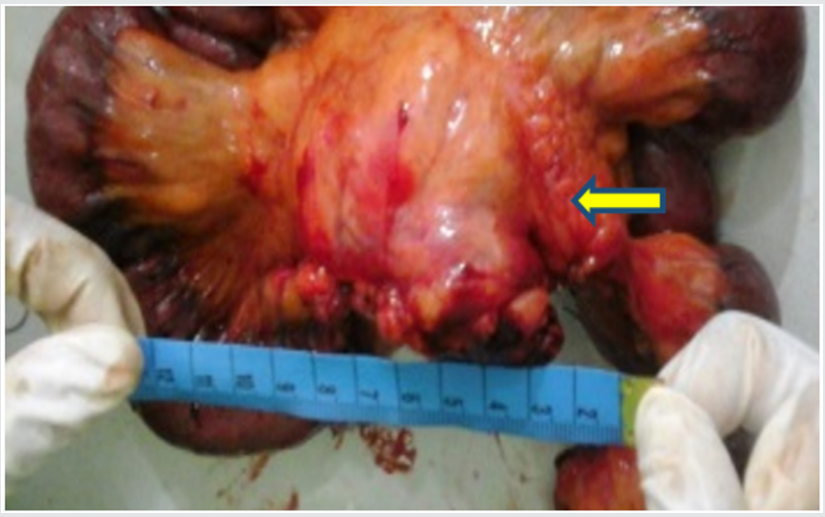

Figure 3: Mesenteric tumor after resection.

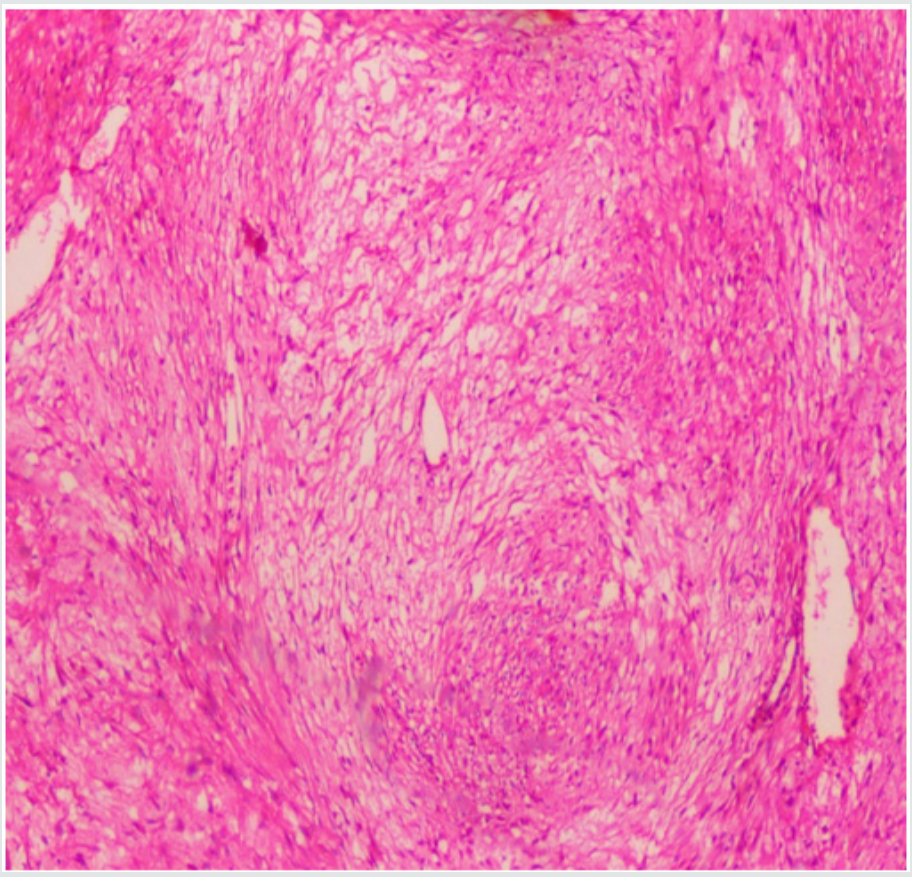

Figure 4: Microscopic appearance suggesting a myxofibroma.

Discussion

Mesenteric fibromatosis is a rare, benign fibrous lesion found in the bowel mesentery or the retroperitoneum. The mesentery of the small bowel is the most common site [3] like in this case. But despite its benign character, mesenteric fibromatosis tend to be invasive and to recurlocally, but do not metastasize [3,7]. Their discovery circumstances are variable. Some cases were incidental finding [4], others have been revealed by symptoms of abdominal discomfort or by complications requiring emergency management [3, 5]. Mesenteric fibromatosis can occur spontaneously or after surgical trauma and also is associated with hormonal therapy, familiar polyposis, or Gardner's syndrome [8,9]. In this case, there was no particular illness or medication in the past history. When the mesenteric fibromatosis is symptomatic, the common signs are: abdominal pain like in this case,constipation, palpable mass on physical examination. These tumors can also be revealed by complications like gastrointestinal bleeding, small bowel obstruction, fistula formation, or gastrointestinal perforation in rare cases [7-9]. The first case of peritonitis secondary to mesenteric fibromatosis reported in the literature was by Gorlin R Jin 1960 and it was initially misinterpreted as a ruptured GIST [6].

Similarly, Ng Eugene PL reported a case of a man who presented signs of peritonitis in clinical examination. Radiological features were consistent with a GIST but in laparotomy there was a tumor diagnosed as mesenteric fibromatosis after the microscopic examination [5]. Mesenteric fibromatosis can also be revealed by occlusive syndrome such as in the case reported by Ricardo PC [10] in which the patient complained for abdominal pain, and CT scan evidenced a heterogeneous nodular lesion in the mesentery, involving the fourth duodenal portion and the initial jejunal segment. Sung HJ & Coskun P [3,11] also reported a case revealed by intestinal obstruction following chronic abdominal pain and constipation. In the case described here, the tumor was discovered following a CT-scan done for chronic constipation with palpable but painless mass. The occurrence of intestinal obstruction while waiting for scheduled surgery has precipitated emergency management. Abdominal fibromatosis being part of desmoid tumors, Sala A. has reported an atypical presentation of small bowel obstruction and perforation secondary to sporadic synchronous intra-abdominal desmoids tumor [12]. Other complications including gynecological and renal complications were reported as possible circumstances of findings [3,10].

Diagnosis of mesenteric fibromatosis is based on clinical suspicion, which depends on the location or local effect of the tumor. The role of imaging is to define the degree of its extension to local and neurovascular structures [13,14]. But, in most cases of mesenteric site, the diagnosis of fibromatosis can be made difficult by the similarity of the symptomatology with the other mainly malignant tumors of the mesentery. Their location and their imaging behavior are often confused with GIST in CT scan [11-14]. In the case reported here, like in more others, the mesenteric fibromatosis was initially misinterpreted as a GIST. Physical examination revealed palpable mass at the left lower quadrant of abdomen and abdominal injected CT scan had found a mass developing in the small intestine whose morphological characteristics evoked a stromal tumor. To distinguish mesenteric fibromatosis from GIST on CT, Zhu H [1] suggested a number of differentiating features in favour of mesenteric fibromatosis including extra-gastrointestinal location, ovoid or irregular contour, homogeneous enhancement, absence of intralesional necrosis and lower degree of enhancement.

Magnetic resonance imaging of mesenteric fibromatosis typically demonstrates low-signal intensity relative to muscle on the T1-weighted image and variable signal intensity on the T2-weighted image. Otherwise, GIST typically has high-signal intensity on T2-weighted images [2]. Surgical excision is essential and is the most effective method for resectable masses [15]. For this patient, excision of the tumor with the small intestine and the corresponding mesentery was performed followed by hand sown end to end anastomosis. There by, in our case, surgery not only allowed resection of the mesenteric mass, but also to treat intestinal obstruction. The patient recovered well postoperatively. For several authors, the preferred treatment of mesenteric fibromatosis is wide local surgical excision with tumor-free margins [8,9,11]. The discrimination between a GIST and mesenteric fibromatosis is difficult since a biopsy cannot be performed, which make surgery an essential step of the management. After surgery, further treatment will depend on the histological nature of the tumor, which will be different for GIST and mesenteric fibromatosis. If in case of GIST, adjuvant treatment with Imatinib is mandatory [16-18], for mesenteric fibromatosis, the need or not for adjuvant therapy is controversial. Surgery with negative surgical margins is sufficient for some authors [8,9] as in our case where as for other authors, surgery only is not advised because of the risk of recurrence. Indeed like GIST, mesenteric fibromatosis is locally aggressive with a high recurrence rate after surgical resection [5]. Studies have shown that the local recurrence rate is 25%–50% at five years after complete resection [10]. For cases with incomplete resection and recurrent cases, radiotherapy may be effective, although this is rarely used for mesenteric fibromatosis because of the severe side effects. Khors and J reported that radiotherapy reduced the recurrence of mesenteric fibromatosis to 20%-40% compared to 40%-70% reduction achieved with surgery only [16]. In addition, chemotherapy (vincristine, cyclophosphamide, and dactinomycineither alone or in combination), non-steroidal anti-inflammatory drugs, hormonal manipulation (tamoxifen, aromatase inhibitors,gonadotropin), and molecular target therapy (imatinib, sorafenib, and sunitinib) may be alternative choices [17]. Pathologic confirmation should be made by microscopic examination and immunohistochemistry [2]. The absence of CD34 and S100 expression supports the fibromatous nature of the lesion and may be helpful in discriminating mesenteric fibromatosis from

GIST [19]. In our case, due to the patient's financial problems, immune histochemical examination could not be performed, the cost of this additional examination being the responsibility of the patient in ou rstudy site. Despite this absence of complementary immunohistochemistry, the pathology examination was sufficient to confirm the diagnosis of fibromatosis by showing an arrangement of uniform fibroblasts associated with collagen. No adjuvant treatment was initiated. No recurrence was observed after 1 year of follow-up and the patient is still alive.

Conclusion

Distinction of mesenteric fibromatosis from GIST is clinically important, as they are different entities with a different clinical course, treatment options, and prognosis. Although, watchful waiting may be offered for asymptomatic mesenteric fibromatosis, surgical resection with microscopic examination and immunohistochemistry remains the best way to confirm the diagnosis, and to avoid complications.

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