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Case Report **OPEN ACCESS**

Voluminous mesenteric desmoid tumor with superior mesenteric vein reconstruction: A case report

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ABSTRACT

Mesenteric tumor or mesenteric fibromatosis is a pathological entity of the mesentery of the small intestine produced by the proliferation of fibroblasts. It has a frequency of 8% of all desmoid tumors and 0.03% of all neoplasms. It has a benign character, but locally it sometimes invades neighboring structures and can recur after ablation. Clinically, this type of tumor presents as asymptomatic abdominal masses, abdominal discomfort or pain, intestinal obstruction, intestinal perforation. A 49-year-old patient presented to the surgical service for palpable abdominal tumor formation and abdominal pain. Following the exploratory laparotomy, an 18 × 12 cm tumor mass in the ileal mesentery was identified and removed. The histopathological examination confirmed the diagnosis. Emphasizing the rarity of these tumors, the diagnostic and therapeutic difficulties, it is justified to report this case.

Keywords: desmoid tumor, mesenteric fibromatosis, abdominal tumor, rare tumor

INTRODUCTION

The term "desmoid", derived from the Greek "desmos", which translates tendon, was stated in 1838 by Muller [1]. These tumors are define as being rare non-metastatic neoplasms, which derive from the musculo-aponeurotic or fascial structures of the body and which have an incidence of 2-4 million/year in general population [2], approximately 0.03% of total neoplasms and 3.5% of total fibrous tissue neoplasm [3]. The study in [4] reports an incidence of 0.73% of all abdominal tumors. Histopathologically, desmoid tumors show a benign morphology characterized by the absence of necrosis, nuclear pleomorphism and atypical mitoses, but due to the high rate of local recurrence, even after radical ablation, they are classified as intermediate malignant neoplasms [5-7].

The pathogenesis of these tumors is not fully elucidated and although some cases have been associated with trisomy of chromosome 8 or 20 (sporadic type) [4], history of abdominal trauma, abdominal surgery (25%) [5, 8, 9], or with conditions ofhyperestrogenemia, sometimes de novo cases were also mentioned [10]. The involvement of estrogenic hormonal disorders in the pathogenesis could explain the increased incidence in women (female-male ratio 3:1), frequently in young pregnant or postpartum women aged between 25 and 35 [5]. In 10-18% of cases, the existence of mesenteric tumors can be associated with the familial adenomatous polyposis mutation (Gardner syndrome) [3, 5]. Clinically, this type of tumor can reveal asymptomatic abdominal masses, abdominal discomfort or pain, intestinal obstruction and intestinal perforation [1, 4 ,11, 12].

CASE REPORT

We present the case of a 49 years old female patient, from an urban area, with no previous pathological history, admitted in the First Surgery Clinic of the "Sf. Apostol Andrei" County Emergency Clinical Hospital, Galati, for palpable abdominal tumor formation and diffuse abdominal pain, nausea, vomiting and general weakness. The clinical examination describes a patient with a body mass index of 22, what it represents a healthy weight for most adults, an abdomen that moves with respiration, slightly distended, difficult to depress, with mild diffuse muscular guarding and tympanism on percussion. She does not declare chronic medication use or a genetic history of similar pathologies. The paraclinical laboratory tests as well as oncological markers did not reveal any pathological changes. Surgery was performed and a voluminous mesenteric tumor formation (**Figure 1**) with invasion of the enteric loop was

Figure 1. Intraoperative aspect of the mesenteric tumor with the invasion of the enteral loop (Source: Emergency Clinical Hospital "Sf Apostol Andrei" Galaţi, First Surgical Clinic. The patient gave her consent to use her personal data and photos by signing the hospitalization form)

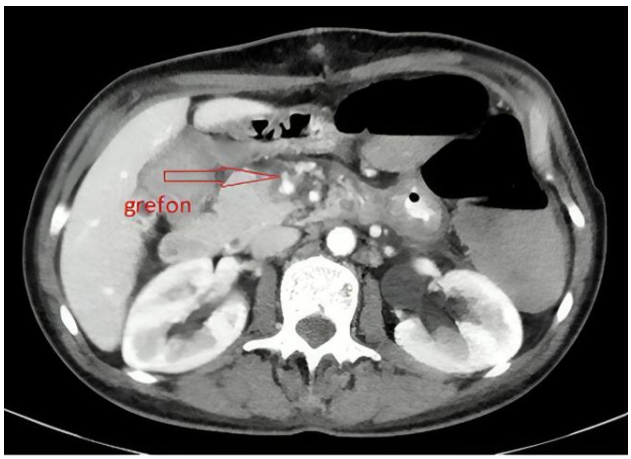

Figure 2. Postoperative CT appearance after superior mesenteric vein reconstruction with venous graft (Source: Emergency Clinical Hospital "Sf Apostol Andrei" Galaţi, First Surgical Clinic. The patient gave her consent to use her personal data and photos by signing the hospitalization form)

found (a tumor formation of 18 × 12 cm was identified approximately 20 centimeters from the ileocecal valve), for which a segmental enterectomy was performed (approximately 25-30 centimeters of ileal loop) with resection of the mesenteric tumor and end-to-end entero-enteroanastomosis.

After 24 hours post-surgery, the patient presented diffuse abdominal pain with generalized muscle spasm, pale skin, cyanosis in the periphery, hypotension, which is why second surgery was performed as an emergency, raising the suspicion of iatrogenic venous intestinal infarction through ligation of the superior mesenteric vein. We consider the ligation of the superior mesenteric vein to have been an intraoperative accident. Laboratory tests reported a hemoglobin value of 6.8 g/dL and a hematocrit of 19.5%. Intraoperatively, the diagnosis was confirmed, and a segmental ileocolectomy with ileotransverso-lateral anastomosis in a double plan, superior mesenteric vein reconstruction with an inverted vein graft from the great saphenous vein was performed (**Figure 2**). The Saphenous vein was chosen due to its high availability, significant length, and ease of harvest.

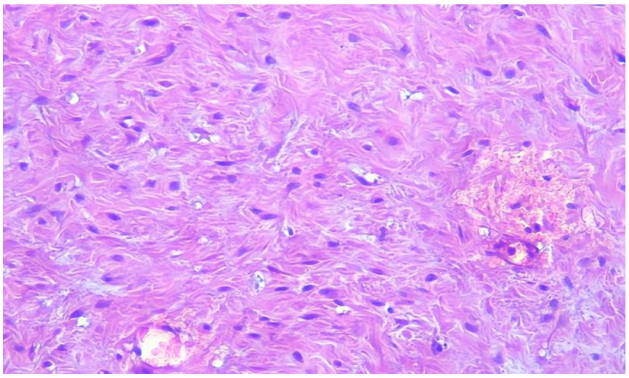

Figure 3. Mesenteric tumor with conventional growth pattern and microhemorrhage (200×, hematoxylin and eosin) (Source: Emergency Clinical Hospital "Sf Apostol Andrei" Galaţi, First Surgical Clinic. The patient gave her consent to use her personal data and photos by signing the hospitalization form)

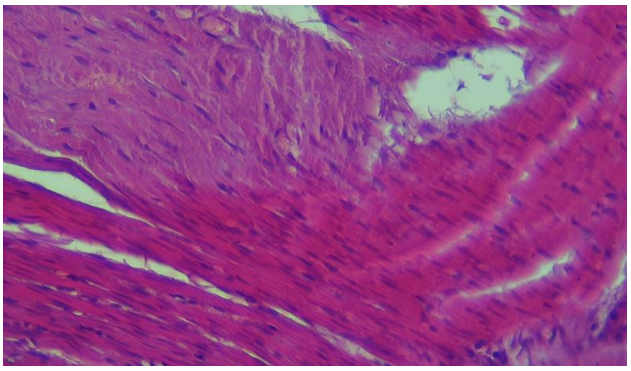

Figure 4. Elongated, narrow, uniform-appearing spindle cells with pale cytoplasm infiltrating the smooth muscle fibers of the tunica propria of the small intestine (200×, hematoxylin and eosin) (Source: Emergency Clinical Hospital "Sf Apostol Andrei" Galaţi, First Surgical Clinic. The patient gave her consent to use her personal data and photos by signing the hospitalization form)

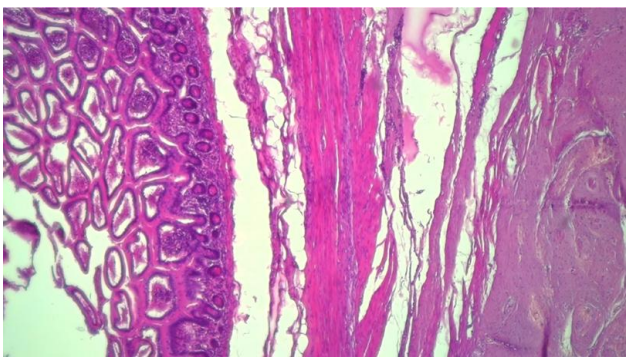

Figure 5. Desmoid tumor infiltrating the wall of the small intestine (40×, hematoxylin and eosin) (Source: Emergency Clinical Hospital "Sf Apostol Andrei" Galaţi, First Surgical Clinic. The patient gave her consent to use her personal data and photos by signing the hospitalization form)

The postoperative evolution under antibiotic, analgesic, antispasmodic, and anticoagulant treatment (clexane 0.4 every 12 hours) was favorable, the patient being discharged on the 16th postoperative day. The histopathological examination established the diagnosis of mesenteric desmoid tumor with intestinal invasion (**Figure 3**, **Figure 4**, and **Figure 5**). Long-

term follow-up was planned with periodic imaging examinations.

DISCUSSION

According to the anatomical location, fibromatosis can be classified into

(1) extra-abdominal fibromatosis (60%),

(2) fibromatosis of the abdominal wall (25%), and

(3) intra-abdominal fibromatosis (8-15%) [5-7, 13-15].

Intra-abdominal fibromatosis is a rare mesenchymal tumor, which develops in the mesentery or retroperitoneum and which is the most aggressive from a biological point of view due to its ability to infiltrate both pelvic and abdominal organs [5]. Mesenteric tumors represents 8% of all desmoid tumors and usually develops in the ileal mesentery, rarely in the mesocolon or the gastro-hepatic ligament [6]. Clinically, the tumor translates into an asymptomatic or painful abdominal mass, intestinal or ureteral obstruction, mucosal ischemia, intestinal perforation, fistula, pyrexia of unknown cause, or functional impairment of the ileal anastomosis after colectomy in cases of familial adenomatous polyposis [4-6]. The differential diagnosis includes GIST (gastrointestinal stromal tumors), lymphoma, carcinoid tumor, fibrosarcoma, or inflammatory fibroma polyp [1].

Computed tomography (CT) scan and magnetic resonance imaging show resectability and degree of adherence to neighboring structures. The histological examination is the only one able to confirm the diagnosis [4].

The therapeutic approach in mesenteric tumors requires a multidisciplinary management that varies depending on the location and extension of the tumor, the intrinsic aggressivity, but also the symptoms and the patient's options.

The most representative factors that predict the recurrence of mesenteric tumors after surgical ablation are the size, the site of the tumor and the age of the patient [16-21].

The long-term recurrence rate of primary small bowel desmoids remains uncertain. Elective surgery is recommended when the patient's condition permits, with the aim of achieving an R0 resection. A multidisciplinary care team is likely beneficial for managing treatment and ongoing monitoring. This case underscores the rarity and treatability of the disease, as well as the challenges involved in managing patients with small bowel desmoids.

Other therapeutic options include systemic treatment in the form of anti-estrogenic therapy and NSAIDs (nonsteroidal anti-inflammatory drugs), tamoxifen and sulindac [22]. Chemotherapy and radiotherapy can be considered for aggressive tumors that have critical invasion of structures, although due to the side effects, they are fewer common options [23-28]. We conducted a literature search to identify comparable cases and present them for comparison, thereby broadening the existing body of knowledge on this rare pathology.

The study in [29] describes a similar case of a 59-year-old patient who was presented to the doctor with an abdominal pain that gradually increased over 3 months. The patient denied experiencing weight loss, nausea, vomiting, or changes in bowel habits. His medical history was notable for arthritis, hypertension, hypercholesterolemia, an appendectomy, and a colonoscopy with polypectomy in 2009. He reported no personal or family history of cancer or inflammatory bowel disease. The procedure was conducted via a standard midline laparotomy. The mass seemed to arise from the mid-jejunum region of the bowel. A segmental enterectomy was carried out, followed by a primary stapled anastomosis. The final pathology confirmed a desmoid tumor originating from the small bowel, with clear surgical margins [29]. The cases were similar; however, the invasion into the depth of the mesentery was probably more pronounced in our case. We suspect that this is why the superior mesenteric vein injury occurred.

The study in [30] considers that dermoid tumors on the abdominal wall are easier to remove, and the recurrence rate is lower compared to those located mesenterically or retroperitoneally. In a percentage of 25%-50%, these tumors relapse despite radical surgical ablation and adjuvant radiotherapy [1]. Neoadjuvant radiotherapy may represent an option [3]. The study in [31] on the role of radiotherapy in mesenteric tumors emphasized its importance with a local control rate of 79% with long-term follow-up and presented it as an alternative to mutilating surgery. The study in [32] mentioned the imaging techniques of CT scanning and/or ultrasound in the follow-up for the early diagnosis of relapses.

Over the past 30 years, isolated case reports have documented adults with mesenteric fibromatosis who presented with additional risk factors for desmoid development alongside Crohn's disease [33-36]. Furthermore, a case has been reported involving a man with Crohn's disease but no other identifiable risk factors for mesenteric desmoids [37]. Similar associations have been noted in adolescents, where no clear additional risk factors were identified [38]. The role of Crohn's disease in the pathogenesis of mesenteric desmoids remains unclear.

CONCLUSION

Mesenteric tumor, although rare, should be considered for differential diagnosis in middle age. Early recognition is quite difficult due to weak and nonspecific symptoms. A multidisciplinary approach with long-term follow-up is useful for detecting tumor recurrence. Surgical treatment, sometimes even in teams with multiple specialists, with resection within the oncological margins, is the elective choice, offering clear benefits. Radiotherapy can represent also a therapeutic modality in mesenteric tumors.

Limitations

The present case report is subject to several limitations, including its single-patient design, which restricts the generalizability and strength of the conclusions.

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