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Huge Intra-Abdominal Desmoid Tumor Presenting 1 Year After Elective One-Anastomosis Gastric Bypass (Mini-Gastric Bypass): A Case Report

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ABSTRACT

Desmoid tumors are uncommon neoplasms of the gastrointestinal tract that are also termed aggressive fibromatosis. Manifestations of intra-abdominal desmoid tumors are variable, from an asymptomatic course of severe pain in the abdomen, the presence of a palpable mass, abdominal distension. We present the case of a 27-year-old woman, presented 13 months after a one-anastomosis gastric bypass (mini-gastric bypass) with vague abdominal pain and early satiety, abdominal distention in the last 3 months. A CT scan of the abdomen revealed a giant intra-abdominal tumor, occupying most of the abdominal cavity with displacement of the internal organs laterally and upward. A midline laparotomy was performed with a resection of the desmoid tumor uterus origin, involving the abdominal wall. The patient's postoperative course was uneventful, and she was discharged on the fourth day post-operation. She did not show any complications or recurrence during her follow-up.

KEYWORDS: Desmoid tumors; Gastrointestinal tract; Mini-Gastric Bypass

INTRODUCTION

World Health Organization data based: Worldwide obesity has nearly tripled since 1975; 39% of adults aged 18 years and over were overweight in 2016, and 13% were obese; 38 million children under the age of 5 were overweight or obese in 2019; most of the world's population lives in countries where overweight and obesity kills more people than underweight. Israel has a prevalence of adult obesity of 26.10%, at number 45 in the world. Obesity is preventable. One-anastomosis gastric bypass is one of the most frequently performed bariatric operations in Israel.

Desmoid tumors are locally aggressive tumors with no known potential for metastasis or dedifferentiation. The estimated incidence in the general population is two to four per million population per year. The etiology of desmoid tumors is unknown. Most desmoids arise sporadically, frequently associated with FAP, antecedent trauma, operation, high estrogen level, pregnancy. Here is a medical case of a patient in whom a giant-sized intra-abdominal desmoid tumor was detected 1 year after bariatric surgery (OAGB) for morbid obesity.

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CASE REPORT

A 27-year-old woman with an initial BMI of 43.5, 13 months after elective surgery, Laparoscopic one-anastomosis gastric bypass. Entered in the surgical department after an outpatient examination for surgical removal of a giant intra-abdominal tumor. Upon admission to department complaints of nausea, rapid satiety, dull intermittent pain does not radiating, constipation, abdominal distention in the last 3 months. She described lack of appetite, persistent fatigue for the last several months, denied fever or significant weight loss. No family members were previously diagnosed with gastrointestinal cancer The patient explains the late request for medical help as possible pregnancy. In the history healthy woman, without comorbid conditions, she does not take medicine, the history of two births by Caesarean section 2015 and 2017. Denies a history of familial gastrointestinal cancer or FAP, does not take oral contraceptives.

Upon abdominal examination, a giant tumor-like mass is determined, extending from the pelvic cavity to the hypochondria, of dense solid structure, non-pulsatile, immobile, and tender to deep palpation. A digital rectal examination was not remarkable.

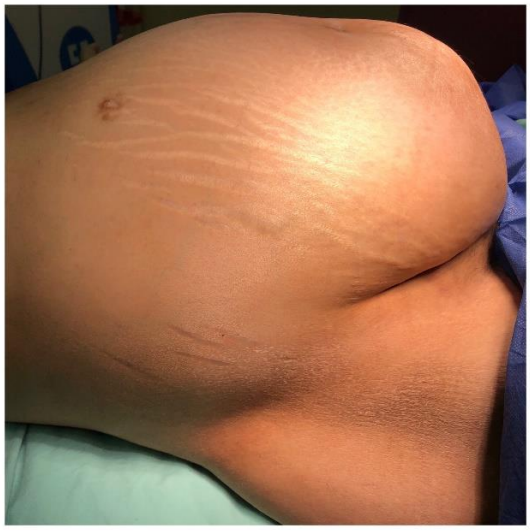

Figure 1: Side view of the abdomen upon admission the patient to the surgical department.

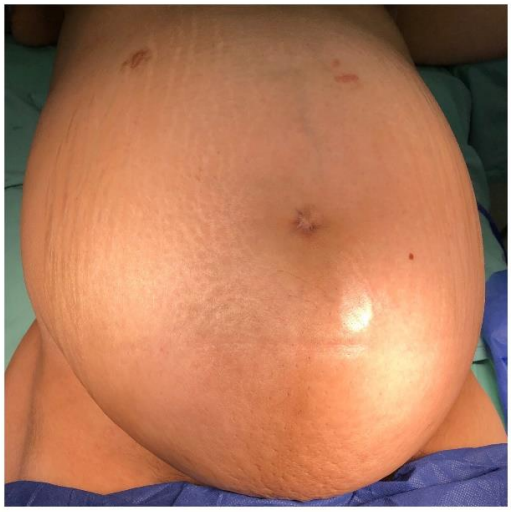

Figure 2: Front view of the abdomen upon admission the patient to the surgical department.

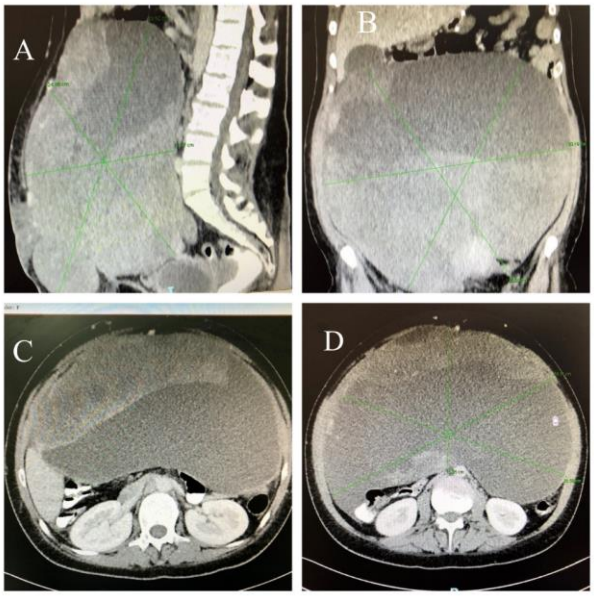

Figure 3: Computed tomography (CT) scans: sagittal (A), frontal (B), axial (C, D) showed a heterogeneous tumor formation, occupying a large part of the abdominal cavity, with displacement of intestinal loops and internal organs laterally and upward.

The results of routine blood investigations were within normal limits. Tumor markers showed an increase in the level of CA-125-102.6. Computerized tomography scan of the abdomen and pelvis were obtained. The abdominal scans demonstrated space occupying lesion 17×28×30 cm, extending from the mid-pelvis and above the umbilicus, heterogeneous structure, partially solid, partially fluid necrotic, tender blood vessels inside the mass is

determined, lateral intestinal displacement without symptoms of obstruction of adjacent structures. The likely source is the ovary.

Due to the huge size of the tumor, a team of surgeons and oncogynecologists performed a midline laparotomy. A large tumor-like mass uterine fundus origin was found, penetrating into the posterior wall of the left rectus muscle and rectus sheath, purple-gray color, heterogeneous structure, in the lower part - solid, in the upper part - liquid cystic; with a widely developed tender vascular network; without signs of distant or peritoneal metastases; with a small amount of serous fluid in the abdominal cavity.

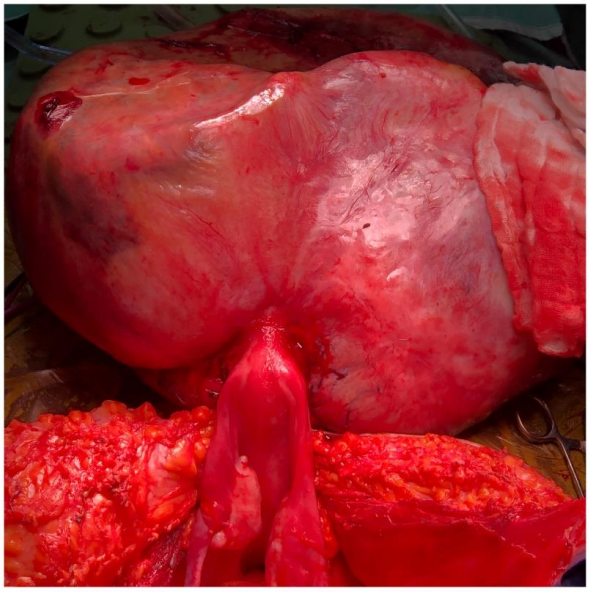

Figure 4: Giant tumor-like mass uterine fundus origin, after mobilization and resection of the affected area of the abdominal wall (part of left rectus muscle and rectus sheath).

The tumor was resected en-block with resection of the wall of the uterus and the affected area of the anterior abdominal wall within healthy tissues. The edges of the resection with the tumor site were sent to frozen sections of intraoperative express biopsy, the answer of the frozen section - benign mesenchymal tumor, the edges of the resection are free of the tumor.

Given the patient's young fertile age, it was decided not to resect the uterus and ovaries until a final pathology answer will be obtained. The patient's post-operative course was uneventful, without complications, the patient was discharged on the 4 postoperative day.

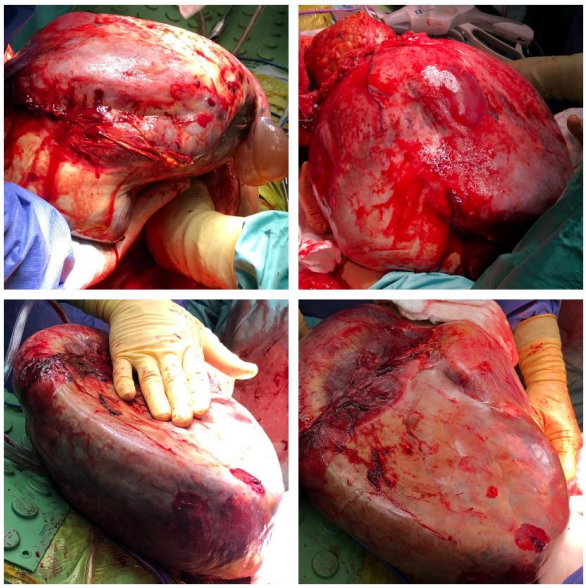

Figure 5: A series of images of a resected giant tumor, oval form, heterogeneous structure, with a smooth surface, tumor size 35×30×20 cm

From a pathology report

Macroscopic description of the specimen: resected tumor mass 35x30x28 sm with a smooth outer surface, a grayish color on section, with areas of hemorrhagic necrosis.

Pathology answer

The tumor cells are diffusely and strongly positive for beta-cathenin, CD99 and Vimentin and focally positive for smooth muscle actin, negative for PLAP, Caldesmon, FOX-L2, Inhibin, Calretinin, S100 protein, STAT-6, CD34, Bc12, Desmin and estrogen receptor. The immunostains results and microscopy support the diagnosis - Intraabdominal Desmoid Fibromatosis.

DISCUSSION

Desmoid tumors (also called aggressive fibromatosis) are benign, slowly growing neoplasms, arise from myofibroblasts, with no metastatic potential, but a

propensity for local recurrence, even after complete surgical resection. Despite being histologically benign, they are local infiltrative and can cause death through destruction of adjacent vital structures and organs. The term "desmoid" originates from the Greek word "desmos," meaning a band or tendon-like, and was first applied in the 1800s to describe tumors with a tendon-like consistency. MacFarlane (1832) first described desmoid-type fibromatosis in 1832, but the term desmoid tumor was not used until 1838 by Mueller (1838). Desmoid tumors are rare; they account for approximately 0.03 % of all neoplasms and fewer than 3 % of all soft tissue tumors. Individuals between the ages of 15 and 60 are most commonly affected; desmoids are rare in the young and in older adults. They are slightly more common in women than in men (about 2:1), and there is no significant racial or ethnic predilection. Common mortality about 11%. Most desmoids arise sporadically, although between 5 and 15 percent are associated with familial adenomatous polyposis (FAP). The prevalence of FAP among patients who develop a desmoid tumor is low 7.5 %. Additional risk factors include pregnancy, exogenous estrogen and antecedent surgical trauma.

Desmoid tumors can develop at virtually any body site, but three main anatomic sites are described: trunk/extremity, abdominal wall, and intra-abdominal (most frequently bowel and mesentery localization). In patients with FAP, intra-abdominal desmoids predominate. Most desmoid tumors present as a deeply seated painless or minimally painful mass with a history of slow growth. Intra-abdominal desmoids can present with intestinal obstruction, bowel ischemia.

Diagnosis is based on imaging (CT scan, MRI, US), histological examination (fibroblastic proliferation appearing as small bundles of spindle cells in an abundant fibrous stroma) and immunohistochemistry (usually positive for vimentin and smooth muscle actin and nuclear beta-catenin but is generally negative for desmin, cytokeratins, and S-100).

Management and treatment of patients with desmoid tumors is complex and variable due to the variety of clinical manifestations and behavior of the tumor, some of which have progressive growth (about 50%), others - spontaneous regression (about 10%).

There are several approaches in the treatment of desmoid tumors, including observation (watch-and-wait), radiation therapy, NSAIDs, tamoxifen, targeted therapy (sorafenib and pazopanib - VEGF inhibitors), cytotoxic therapy (doxorubicin, decarbazine, methotrexate, vinblastine). Surgical resection with negative margins is still considered the treatment of choice for operable desmoid tumors, for tumors larger than 7 cm in greatest dimension and it can decrease the recurrence rate by 25-50%. Guidelines from the National Comprehensive Cancer Network suggest that monitoring is a reasonable strategy for patients with tumors that are not causing significant impairment. History and physical examination with appropriate imaging is recommended every 3-6 months for 2-3 years, then annually.

In the case of our patient, surgical treatment was performed due to the giant size of the tumor with clinical symptoms, impaired bowel function. Intra-abdominal desmoid tumor is an unusual finding after a bariatric surgery. Based on the literature review, four cases of desmoid tumors after bariatric surgery were described (one case after sleeve gastrectomy, one case after one-anastomosis gastric bypass, two cases after Roux-en-Y gastric bypass).

CONCLUSION

In conclusion, there has recently been a significant increase in patients with morbid obesity, which has led to

an increase in the frequency of bariatric surgery. Despite the fact, that antecedent surgical trauma is one of the main risk factors for desmoid tumors, hormonal and metabolic effects of bariatric surgery, changes in gastrointestinal anatomy in patients with metabolic syndrome require further study.

CONFLICT OF INTEREST STATEMENT

None declared.

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