

Case report

A Giant Desmoid tumor of the sigmoid colon mesentery

Abstract

Desmoid tumors of the colon are exceptionally rare and represent an aggressive form of intra-abdominal desmoid tumors. The sigmoid colon mesentery, while generally associated with lower morbidity, can be affected by these tumors. The etiology of mesenteric desmoid tumors includes factors such as trauma, surgery, hormonal influences, and hereditary conditions like familial adenomatous polyposis coli and Gardner's syndrome. Intra-abdominal desmoid tumors are most commonly located in the mesentery in about 80% of cases.

Solitary desmoid tumors in the sigmoid colon mesentery are typically benign and have a favourable prognosis. These tumors are characterized by mesenteric fibroblastic growth, which can occur spontaneously or as a result of surgical trauma. Mesenteric desmoids most frequently develop in the intestinal mesentery, followed by the omentum and mesocolon.

These tumors more commonly originate from the left side of the colon and the sigmoid colon, accounting for 60-70% of cases. They are usually diagnosed using simple ultrasonography and CT scans of the abdomen and pelvis. Complete surgical excision is the preferred treatment.

We report a case of a giant desmoid tumor of the sigmoid colon mesentery in a 37-year-old male patient, diagnosed via CT scans of the abdomen and pelvis. The patient presented with a painless large mass in the pelvis, arising from the mesentery of the sigmoid colon, and was planned for open surgery.

Key words

Desmoid tumor, Familial adenomatous polyposis, Gardner's syndrome, Sigmoid colon

Introduction

Desmoid tumors are monoclonal fibroblastic proliferations arising from connective tissue. They can be categorized into extra-abdominal (60%), anterior abdominal wall (25%), and intra-abdominal (15%) types. Abdominal desmoid tumors occur sporadically or in association with familial adenomatous polyposis and Gardner's syndrome in 10-15% of cases. [1,2,4]

Solitary desmoid tumors of the sigmoid colon mesentery are rare benign neoplasms, with only a few reported cases. A literature review revealed just 15 reported instances of these tumors arising from the mesentery of the colon. Mesenteric masses present a radiological evaluation challenge, utilizing ultrasound, CT, MRI, and PET CT. On ultrasound, desmoid tumors typically appear as hypoechoic masses with internal homogeneity. On CT, they usually present as well-defined solid masses with homogeneous density and no calcification. [3,4,6]

Complete surgical resection of the tumor with negative microscopic margins is the standard treatment goal. Patients with sporadic desmoid tumors generally have a low recurrence rate following resection. [1,2,8]

Case Report

A 37-year-old male patient was admitted to our center on October 12, 2022, presenting with a large, slowly progressive, painless pelvic mass of 12 months duration. He was a truck driver who had sustained an abdominal trauma from a steering wheel injury two year prior. One year after the trauma, he noticed a pelvic mass that progressively increased in size.

On physical examination, the mass was non-tender, firm, and mobile, extending from the left lower abdomen to the umbilicus and up to the epigastric region. The surface of the mass was smooth, with limited mobility. Laboratory reports were within normal limits, and tumor markers such as CEA, CA 19-9, CA-125, and alpha-fetoprotein were also within normal limits.

Ultrasound revealed a large heterogeneous mass in the pelvis extending from the urinary bladder to above the umbilicus. A CT scan of the abdomen and pelvis showed a large, well-circumscribed heterogeneous solid mass, measuring 17x12x10 cm, involving the sigmoid colon mesentery. The mass closely abutted the dome of the bladder and sigmoid colon and reached up to the abdominal wall anteriorly. There was no evidence of peritoneal spread, suggesting a benign tumor.

The patient was scheduled for surgery through a midline explorative laparotomy, which revealed a large, well-circumscribed mass arising from the lateral aspect of the sigmoid mesocolon, measuring 17x12x10 cm. The mass had no attachment to adjacent structures such as the urinary bladder and small intestine, and it was completely resected.

The post-operative course was uneventful, and the patient was discharged on the 10th post-operative day. The benign nature of the tumor, radiation or systemic therapy was not advised. Gross examination of the tumor showed a well-circumscribed mass with firm consistency, measuring 17x12x10 cm and weighing 5 kg. The cut section revealed a glistening white surface with a fascicular appearance. Histologically, the tumor exhibited typical features of desmoid fibromatosis, with spindle cells in a collagenous stroma. All resection margins were negative. Immunohistochemically, the tumor cells were positive for β -catenin and negative for CD117, CD34, and DOG-1. **(Fig 1-7)**



Fig-1CT abdomen and pelvis shows a large solid mass in pelvis

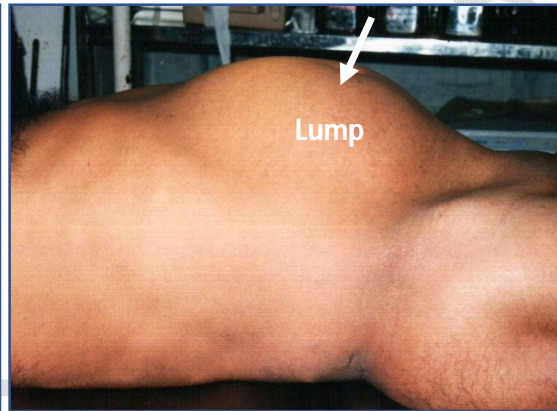


Fig-2Photographs showing lump extending from suprapubic to epigastric region

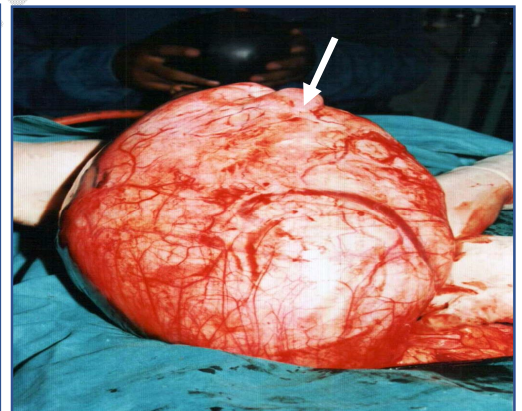
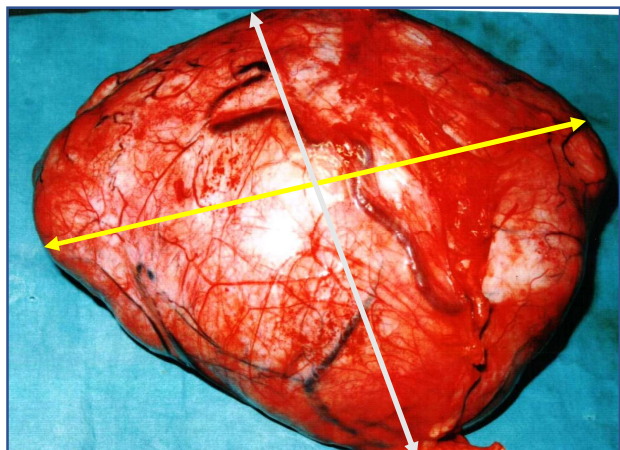
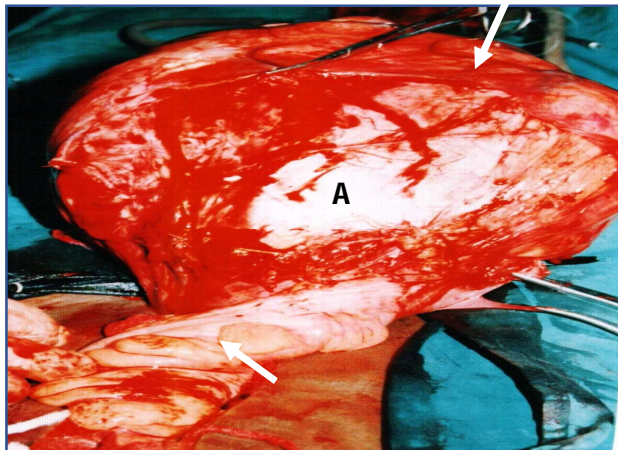


Fig-3 Intraoperative photographs showing a large, solid tumor



B

Fig-4Intraoperative photographs showing a large, A- solid tumor arising from B-sigmoid colon

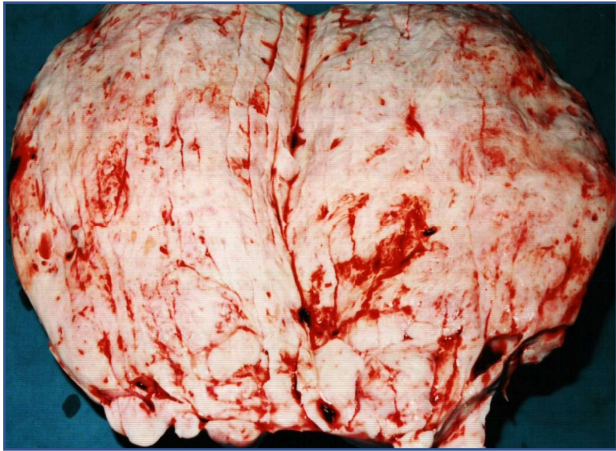


Fig-5Photographs showing a large solid mass measuring 17x12x10 cm. weight 5kg

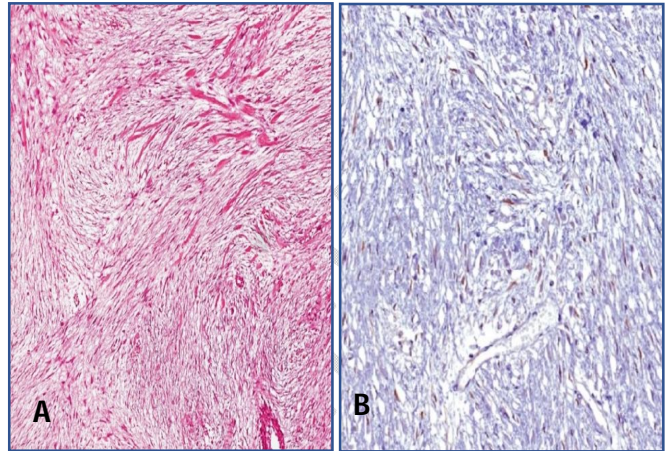


Fig-6The cut section revealed a glistening white surface with a fascicular appearance

Fig-7A-features of desmoid fibromatosis, with spindle cells. **B**-positive for β -catenin

Discussion

Desmoid tumors, also known as aggressive fibromatosis, were first described by Muller in 1838. The annual incidence of desmoid tumors is only 2-4 per million, accounting for 3% of all soft tissue tumors. Intra-abdominal desmoid tumors comprise nearly 8% of all fibromatosis cases. The etiology of mesenteric desmoid tumors includes factors such as trauma, surgery, hormones, and heredity. Mesenteric desmoid tumors of the colon have a low morbidity rate. [1,2,4]

According to the literature, desmoid tumors most commonly involve the sigmoid colon (60-70%), followed by the cecum (25-40%), and the transverse colon (1-4%). Desmoid tumors are classified based on their anatomic location: [2,3]

1. Extra-abdominal (60%)
2. Abdominal wall (25%)
3. Intra-abdominal (8%)

Mesenteric desmoid tumors can occur either spontaneously or as a result of surgical trauma. They most frequently develop in the intestinal mesentery, followed by the omentum and mesocolon. A literature search revealed only 15 reported cases of large bowel mesenteric desmoid tumors. The median age of presentation ranges from 27 to 70 years, with a male preponderance. These tumors more commonly originate from the left side of the colon, rectum, sigmoid colon, and descending colon than the right side. Desmoid tumors of the sigmoid mesocolon are usually benign, have a good prognosis, and do not recur after surgery. [1,2,5,6]

Clinically, most patients are asymptomatic due to the slow growth of the tumor. The clinical presentation varies depending on the tumor size and location, often presenting as an intra-abdominal palpable mass, abdominal pain, constipation, and abdominal discomfort. Radiological evaluation can be challenging; ultrasound, CT, MRI, and PET CT are used. On ultrasonography,

desmoid tumors appear as hypoechoic masses with internal homogeneity. On CT scans of the abdomen and pelvis, desmoid tumors usually appear as solid masses with homogeneous density and regular margins. MRI may be an alternative diagnostic tool for intra-abdominal recurrence cases. [7,8,9]

The differential diagnosis includes GISTs, mesenteric fibromatosis, leiomyomas, leiomyosarcoma, lymphoma, and solitary fibrous tumors. The immunohistochemical characteristics of mesenteric desmoid tumors include positive staining for vimentin and β -catenin, but negative staining for smooth muscle actin, S-100, CD117, and CD34. [2,3,5]

Complete surgical excision with a wide surgical margin of at least 2 cm is the first-line therapy for resectable tumors. In our case, it was an R0 resection with a margin greater than 1 cm, and after one year, there was no sign of recurrence. Complete resection of the tumor with negative margins macroscopically prevents local recurrence. Solitary desmoid tumors of the sigmoid mesocolon are benign and can be surgically resected. They do not usually recur or metastasize, but close follow-up is needed. Histologically, these tumors show spindle cells, with nuclear atypia or mitoses not usually observed. These tumors are typically benign and have a good prognosis. [7,8,10]

Conclusion

Solitary desmoid tumors of the sigmoid mesocolon are usually benign and have a good prognosis, with no recurrence after surgery. Complete surgical excision with adequate margins is the treatment of choice for these tumors.

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