Case Report

# Mesenteric Fibromatosis Presenting as an Irreducible Inguinal Hernia

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## ABSTRACT

Mesenteric fibromatosis is a rare benign tumor of the abdominal cavity. It can present as a sporadic case or as a part of polyposis syndromes. It often infiltrates surrounding structures but rarely metastasizes. Surgical resection is the treatment of choice. The following is a case report of a mesenteric fibromatosis mass presenting as an irreducible inguinal hernia.

**Key Words:** Desmoid, fibromatosis, hernia

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Mesenteric fibromatosis is usually a rare neoplasm with mostly a benign behavior. Sporadic cases have been reported, but it is usually a part of familial adenomatous polyposis or Gardener's syndrome. It commonly presents as an abdominal mass but can present in many different ways. One rare presentation that we encountered is a scrotal mass that was thought initially to be a hernia in a young male. This presentation has not been reported previously in the literature.

## CASE REPORT

An 18-year-old male with a medical history of right inguinal hernia repair during childhood, presented with a history of left groin swelling for three months. There was a progressive increase in its size. Initially, he had been able to reduce the mass, but now it had become irreducible. His history revealed no symptoms of bowel obstruction.

On examination, he looked comfortable with normal vital signs and no signs of distress. On palpation, a large mass was found in the left inguinal region extending to the left scrotum, about 10 cm in diameter. Cough impulse was present; the transillumination test was negative; and the mass could not be reduced.

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As the presentation was not typical for an incarcerated hernia, further investigations were done. An ultrasound examination of the area revealed normal testes with no hydrocele; it also showed that the mass had increased vascularity most likely representing omentum.

The patient was prepared for surgery and taken to the operating room where the left inguinal canal was explored. The content of the hernia could not be reduced, so the hernia sac was opened and a piece of omentum was seen extending from the internal ring to the scrotum. The incision was then extended into the scrotum and a large encapsulated mass was encountered attached to the omentum [Figure 1]. The mass was easily delivered out of the scrotum as it was not attached to the scrotum or the testes and it was completely excised along with a small piece of omentum to get a clear margin. The hernia was then repaired using a prolene mesh. The patient's postoperative course was uneventful and he was discharged from hospital after one day with no complications.

Grossly, the mass measured 19 × 9 × 7 cm and was well encapsulated. The cut surface had a yellowish myxoid appearance [Figure 2].

The histopathological examination revealed that the tumor consisted of bland spindle-shaped and satellite cells evenly distributed in a collagenized and myxoid stroma [Figure 3a]. A Keloid type collagen was also present. Occasional mitotic

figures were seen. The tumor tested positive for Vimentin, but negative for CD117 and CD34 [Figure 3b]. All were consistent with mesenteric fibromatosis. The patient was called back to perform endoscopy and CT scan of the abdomen to rule out polyposis syndromes or other abdominal lesions, but he did not return for follow-up.

## DISCUSSION

This is the first description of mesenteric fibromatosis presenting as an irreducible inguinal hernia.

Mesenteric fibromatosis is a rare abdominal tumor. It is also known as deep fibromatosis, aggressive fibromatosis, and desmoids tumors.

It usually has a benign behavior; however, it can present as a huge mass or infiltrate surrounding structures but rarely metastasizes.[1]

It can present as a sporadic case or as a part of familial adenomatous polyposis or Gardener's Syndrome.[2,3] Association with Crohn's disease has been reported.[4] It can present after trauma, surgical procedure, pregnancy, or prolonged estrogen use. The usual age of presentation is in the fourth decade of life, but it can present at any age, even in neonates. It is more common in females than males.

The origin is usually the small bowel mesentery, but can originate from any mesenteric or omental surface. The clinical presentation varies according to the size and location of the mass but it is usually insidious. Mesenteric fibromatosis usually presents as an abdominal mass, pain, or discomfort but can present in several other ways as well, including acute abdomen, bowel obstruction, bowel perforation, fistula, or even infiltration of porta hepatis with obstructive jaundice.[5,6]

The lesion is usually well circumscribed on gross appearance with varying sizes and is usually composed of spindle-

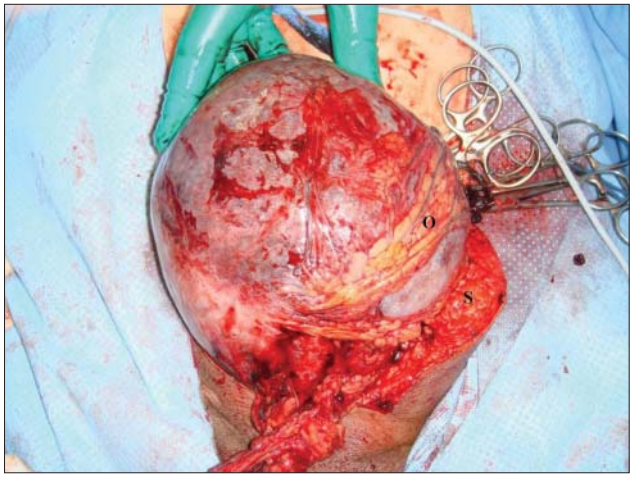


**Figure 1:** The mass exposed after opening the scrotum (S) seen attached with a piece of omentum (O)

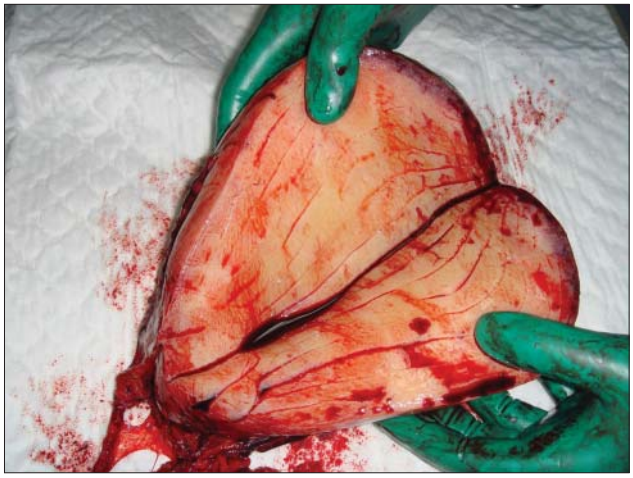

**Figure 2:** The cut surface showing a yellowish myxoid appearance

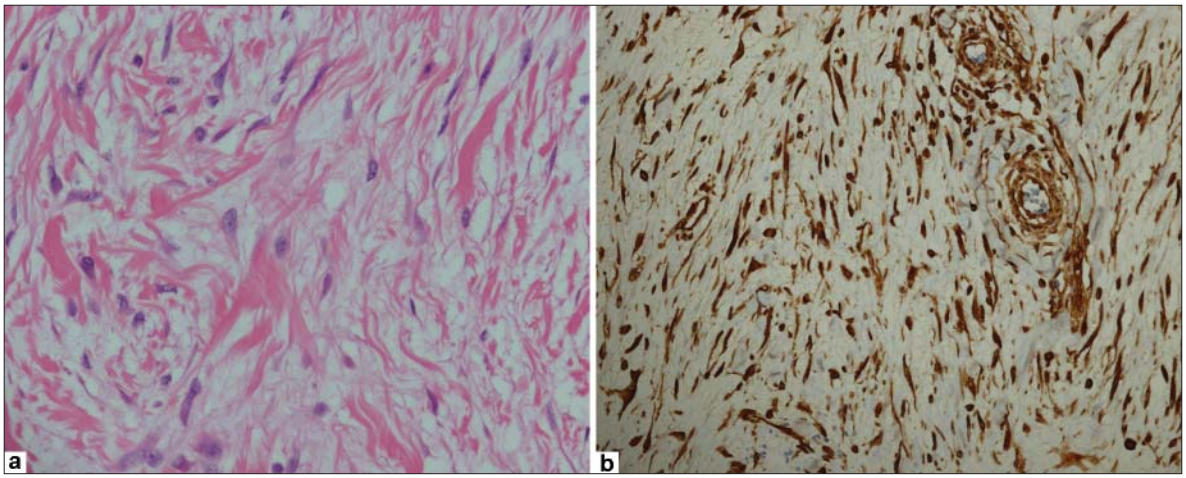


**Figure 3:** (a) Microscopic examination of the mass showing spindle-shaped cells distributed in a collagenized and myxoid stroma (b) positive Vimentin stain

shaped cells with keloid type collagen deposition. Necrosis, hemorrhage, and significant mitosis are usually absent. Mesenteric fibromatosis may stain positive for CD117 but is usually negative for CD34.[3,7]

The differential diagnosis usually includes gastrointestinal stromal tumor, carcinoid, lymphoma, and sarcoma.

If the tumor can be resected safely, then surgical resection is usually the treatment of choice and often requires resection of a segment of bowel. Surgical resection is usually curative and recurrence is rare. In patients with Garden's syndrome, the recurrence is higher than in sporadic cases and the recurrence is usually local.[1,8] Recurrence can still be resected but with increased morbidity and mortality.

If the tumor is unresectable or could only be resected partially due to size or infiltration, then some pharmacological agents can be used alone or in conjunction with surgery to achieve more tumor size reduction. Cyclooxygenase 2 inhibitors were tried and successfully reduced the size of the tumor.[9] Sulindac alone did not show a good response, but the combination of sulindac with the antiestrogen agent toremifen is helpful in reducing the size of the tumor and relieving symptoms, thus improving quality of life.[10] Other chemotherapeutic agents or even radiotherapy have been used but their role is still unclear.

In a case like this, it is very important to rule out other abdominal lesions or polyposis syndromes. Unfortunately, our patient did not respond to our request to return to the clinic for further evaluation.

Medline search was performed using the key words fibromatosis, desmoids, and hernia, and no similar case was reported from 1966 till present.

## REFERENCES


1. Al Jadaan SA, Al Rabeeah A. Mesenteric fibromatosis: Case report and literature review. J Pediat Surg 1998;34:1130-2.
2. Forte MD, Brant WE. Spontaneous isolated mesenteric fibromatosis Report of a case. Dis Colon Rectum 1988;31:315-7.
3. Al-Nafussi A, Wong NA. Intra-abdominal spindle cell lesions: A review and practical aids to diagnosis. Histopathology 2001;38:387-402.
4. Salter G, Greenstein AJ. Mesenteric fibromatosis in Crohn's disease. J Clin Gastroenterol 1996;22:147-9.
5. Holubar S, Dwivedi A, O'Connor J. Giant mesenteric fibromatosis presenting as small bowel obstruction. Am Surg 2006;72:427-9.
6. Karagulle E, Gokturk HS, Turk E, Yildirim E, Kiyici H, Karakayali H. Intestinal perforation from primary intra-abdominal fibromatosis. Saudi Med J 2007;28:639-40.
7. Rodriguez JA, Guarda LA, Rosai J. Mesenteric fibromatosis with involvement of the gastrointestinal tract, A GIST simulator: A study of 25 cases. Am J Clin Pathol 2004;121:93-8.
8. Burke AP, Sobin LH, Shekitka KM, Federspiel BH, Helwig EB. Intra-abdominal fibromatosis: A pathological analysis of 130 tumors with comparison of clinical subgroups. Am J Surg Pathol 1990;14:335-41.
9. Ng TY, Yang MD, Chen YF, Chang CH. Resolution of hydronephrosis due to massive mesenteric fibromatosis using cyclo-oxygenase 2 inhibitors. J Urology 2007;70:591.e3-e4.
10. Bus PJ, Verspaget HW, van Krieken JH, de roos A, Keizer HJ, Bemelman WA, *et al.* Treatment of mesenteric desmoids tumor with the anti-oestrogenic agent toremifene: Case histories an overview of the literature. Eur J Gastroenterol Hepatol 1999;11:1179-83.


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