Mesenteric Fibromatosis

Radhakrishna V,[1] Patil RS[2]

[1]Department Paediatric Surgery

Indira Gandhi Institute of Child Health,

Bengaluru, India.

[2]Department of General Surgery

Basaveshwar Hospital, MR Medical College,

Gulbarga, India.

Corresponding Author

Veerabhadra Radhakrishna

Department Paediatric Surgery

Indira Gandhi Institute of Child Health,

Bengaluru, India.

E-mail:vbrps2016@gmail.com.

Citation

Radhakrishna V, Patil RS. Mesenteric Fibromatosis. *Kathmandu Univ Med J.* 2017;60(4):344-5.

ABSTRACT

Mesenteric fibromatosis is a rare, locally invasive benign tumor arising from fibroblasts and fibrous tissue of mesentery. Their infiltrative nature can cause fatal visceral involvement. Most of the cases reported have an association with Gardner's Syndrome, prolonged intake of estrogen, and previous trauma, but mesenteric fibromatosis can occur as a primary ailment without an underlying stimulant. A nineteen-year female presented with mass and pain abdomen diagnosed to have subserous fibroid but eventually turned out to be primary mesenteric fibromatosis. She underwent wide excision along with the resection of adjacent bowel. She is on follow-up with no evidence of recurrence.

KEY WORDS

Estradiol receptors, Fibromatosis, Mesenteric mass, Wide surgical excision

INTRODUCTION

Mesenteric fibromatosis is a non-malignant proliferation of mesenteric fibrous tissue which has a propensity to relapse without distant metastasis.[1] The exact etiology of mesenteric fibromatosis is not known. Most of the cases reported have an association with Gardner's Syndrome, prolonged intake of estrogen and previous trauma, but mesenteric fibromatosis can occur as a primary ailment without an underlying stimulant.[2] Here we present a case of primary mesenteric fibromatosis, presented to our institution.

CASE REPORT

A nineteen-year female presented with three months history of mass per abdomen and ten days of pain abdomen. On examination, vitals were stable. A hard mass of 25 X 15 X 15 cm size was felt arising from the pelvis. Blood investigations were within normal limits. USG abdomen showed an iso-echoic lesion noted above uterine fundus suggestive of a subserous fibroid. FNAC revealed it to be a spindle cell tumor.

Per-operatively, a mass measuring 25 X 20 X 10 cm was found arising from the mesentery of ileum which was excised along with involved part of the bowel, and ileo-ascending colon anastomosis was done (fig. 1 and fig. 2). The mass weighed 2.2 kg.

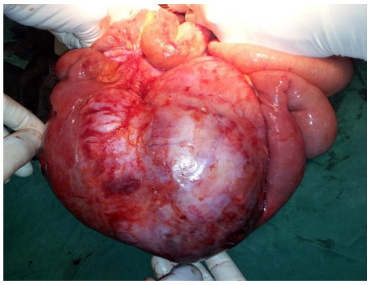

Figure 1. The mesenteric mass intraoperative picture.

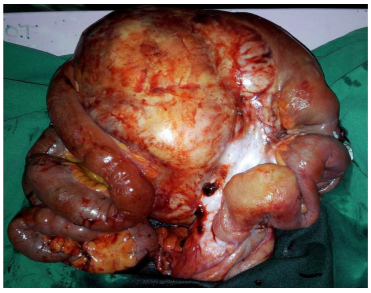

Figure 2. Resected specimen containing mesenteric mass attached to coils of intestine

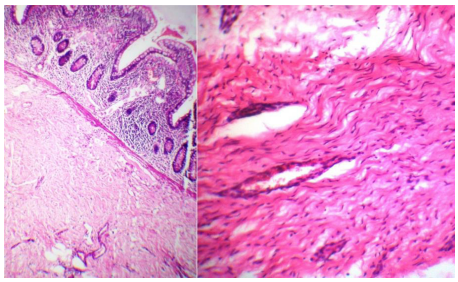

Figure 3. Proliferation of differentiated fibroblasts (myofibroblasts) consisting of large spindle cells with a small dense nucleus, low cytoplasm, and ill-defined borders.

Histopathological examination revealed proliferation of differentiated fibroblasts (myofibroblasts) consisting of large spindle cells with a small dense nucleus, low cytoplasm, and ill-defined borders with no evidence of malignancy; and margins were clear (fig. 3). Post-operative period was uneventful. Patient discharged on the 12th post-operative day.

DISCUSSION

Aggressive fibromatosis or desmoid tumors are rare neoplasms that consisting of 0.03% of all tumors.[3] And Fibromatosis of mesentery is infrequent with only a little number of reports in the literature.

The etiopathology of mesenteric fibromatosis include operative trauma, strong association with Gardner's syndrome suggesting genetic etiology, prolonged intake of estrogen, and infection with HPV. These tumors grow slowly in young girls and reach a peak at menopause, pointing a finger towards estrogen as a growth factor. A study has found higher amounts of estradiol receptors in this tissue than in control tissue.[4]

Progressive fibrous and fibroblastic proliferation within the mesentery leads to the development mesenteric fibromatosis. This can involve local vascular structures. These lesions can compress and obstruct the bowel lumen. Although they are benign lesions histologically, their infiltrative pattern can eventually cause fatal visceral involvement. The presenting complaints can be abdominal pain, discomfort, vomiting, constipation, weight loss, symptoms suggestion of intestinal obstruction and hydronephrosis.

The diagnosis of mesenteric fibromatosis is confirmed only by the histopathological examination. Imaging remains the backbone of preoperative investigation to establish a working diagnosis of mesenteric fibromatosis. Plain radiographs may show a soft tissue mass. Contrast studies can disclose compression and kink of the small bowel loops, and mucosal tethering infrequently.[4] The ultrasound findings of mesenteric fibromatosis are non-specific. The fibroblast and collagen content and vascularity of the lesion define the sonography findings.[4] On computed tomography, mesenteric fibromatosis appears as a soft tissue mass displacing/involving adjacent viscera, usually looks as if as bowel loops are encased.[4] On magnetic resonance imaging, mesenteric fibromatosis appears hypointense on T1-weighted images, because of its predominant fibrous composition. On T2-weighted images, it shows variable signal intensity with a hyperintense signal from those that had demonstrated marked growth on follow-up imaging.[4]

The first-line treatment for mesenteric fibromatosis is the wide surgical excision.[5] As noted in our case, the majority of these lesions require resection of the attached segment of the bowel.6 In cases of recurrent and inoperable lesions, radiotherapy may be used before surgery to shrink the tumor size, reduce vascularity to render it operable. Adjuvant radiotherapy reduces the recurrence rate of mesenteric fibromatosis to 20%-40%, compared to 40%-70% with resection alone.[7] In cases where surgery and radiotherapy don't yield the desired success, systemic therapy with pharmacological agents can be instituted, which include anti-proliferative agents, cytotoxic drugs, estrogen receptor antagonists (tamoxifen), and nonsteroidal anti-inflammatory drugs agents (sulindac). Chemotherapeutic agents such as dactinomycin, vincristine, and cyclophosphamide, singly or in combination, have been tried with varying success.[8]

Mesenteric fibromatosis can present with unusual clinical features and demonstrate a broad spectrum of imaging and histological details. Operating surgeons should have a high index of suspicion while managing a patient with an abdominal mass.

REFERENCES

1. Enzinger FM, Weiss SW. Fibromatosis. In: Enzinger FM, Weiss SW. Soft tissue tumor. St. Louis, MO: C.V. Mosby Co.; 1995: 201-29.
2. Vaswani BA, Shah M, Shah PM, Parikh BJ, Anand AS, Sharma GL. Giant mesenteric fibromatosis in Gardner's syndrome. *Indian Journal of Cancer.* 2011; 48 (1): 140-2.
3. Daryani NE, Momeni AR, Azizi MR, Movafaghi S. Mesenteric Fibromatosis (Desmoid Tumor) presenting as recurrent abdominal abscess: report of a rare case. *Govaresh.* 2008; 13(2): 128–32.
4. Gari MKM, Guraya SY, Hussein AM, Hego MMN. Giant mesenteric fibromatosis: Report of a case and review of the literature. World J Gastrointest Surg. 2012; 4(3): 79-82.
5. Yang CH, Sheen-Chen SM, Lu CC, Ko SF, Eng HL. Computed tomographic presentation of mesenteric fibromatosis. *Dig Dis Sci.* 2005; 50: 348-350.
6. Smith AJ, Lewis JJ, Merchant NB, Leung DH, Woodruff JM, Brennan MF. Surgical management of intra-abdominal desmoid tumours. *Br J Surg.* 2000; 87: 608-13.
7. Khorsand J, Karakousis CP. Desmoid tumors and their management. *Am J Surg.* 1985; 149: 215-8.
8. Yantiss RK, Spiro IJ, Compton CC, Rosenberg AE. Gastrointestinal stromal tumor versus intra-abdominal fibromatosis of the bowel wall: a clinically important differential diagnosis. *Am J Surg Pathol.* 2000; 24: 947-57.