

## Fibromatosis of Small Intestine in Children

Mansoor Mahmood, Nadeem Akhtar, Abdul Hameed, Israr Zahir,  
M. Zaheer Abbasi

Department of Pediatric Surgery Children Hospital, Pakistan Institute of Medical Sciences Islamabad Pakistan

**Abstract:** Fibromatosis is a rare spindle cell lesion which can at times present as tumor which is characterized by the remarkable proliferation and invasive growth of fibrous tissue. It often arises from the abdominal wall or the extremities and rarely from the mesentery and abdominal organs. The patient was 9 year old child with major complaints were on & off abdominal pain and mass in lower abdomen. She underwent exploratory laprotomy, with excision of mass and end to end anastomosis.

**Key words:** abdominal fibromatosis, GI Tract.

### Introduction

Fibromatosis are a heterogeneous group of lesions, some of which are potentially lethal entities requiring wide excision and careful follow-up for cure. Fibromatosis is a benign tumor that occurs most commonly in the abdominal wall or extremities. Intra-abdominal fibromatosis is the most common primary mesenteric tumor with spindle cell morphology. It may occur at any age, can be solitary or multiple, and may involve the small bowel, omentum, mesocolon and retroperitoneum<sup>1,2</sup>. It can sometime mimic the clinical characteristics, radiological imaging and histopathological appearance of a gastrointestinal stromal tumor (GIST), thus leading to an incorrect diagnosis.

**Case Report:** A 9 years old female patient presented with history of abdominal pain on & off from last 3 months. Pain was colicky, non radiating in nature, and not associated with any aggravating or relieving factor. Her past medical or surgical history and family history was not significant. On examination pale looking child and abdominal palpation there was around 10 cm mobile mass, rounded in shape, firm in consistency, non tender in the umbilical and hypogastric region. Her baseline investigations were normal and hepatitis profile was negative for both hepatitis B & C. urine R/E was normal.

The ultrasound abdomen showed heterogeneous mass in pelvic 8\*7.3\*10 cm in size. It was separated from uterus and genito-urinary tract. The mass showed echogenic margins and echodense areas within it. Rest of the mass showed in homogenous texture with echogenic foci. No vascularity was seen in the mass. It is non compressible and did not show any peristaltic activity. The CT scan showed fat containing mass lesion in lower abdomen measuring approximately 6\*5\*7 cm. It is visualized in mid lower abdomen anterior to aortic bifurcation. However it was mobile and changing its position.

### Gross Examination:

An exploratory laprotomy was performed. A mass measuring 10cm to 7 cm was found in the proximal jejunum. Mass was resected with clear small intestinal margins fig 1, 2.

### Microscopic Examination:

The sections revealed well circumscribed benign tumor composed of benign mature collagen fibers arranged in thick clusters. There are very few fibroblasts with nuclei. At places the collagen has become hyalinised (Fig 3&4)

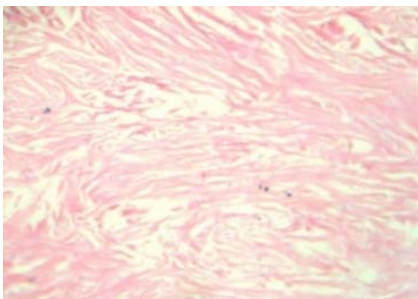


From Department of Pediatric Surgery, Children Hospital, PIMS, Islamabad. Correspondence may be directed to Dr. Mansoor Mahmood

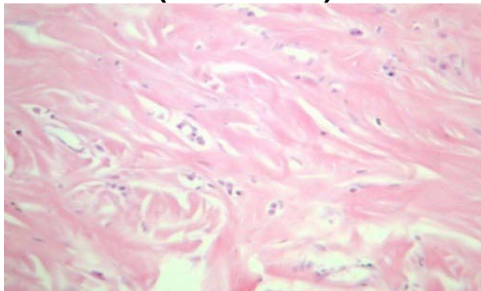
**Figure 1: Mobile firm mass, the proximal**



**Figure 2: Cut surface of the mass**



**Figure 3: Dense mature collagen fibers (H&E x 100)**



**Figure 4: Dense mature hyaline collagen fibers at higher magnification (H&E x 400)**

### Discussion

Fibromatosis is a benign tumor that occurs most commonly in the abdominal wall or extremities. Very rarely, it can occur from the mesentery. In many cases the tumors are related to Gardner's syndrome<sup>3</sup>. Intra-abdominal fibromatosis (IAF) is the most common primary mesenteric tumor<sup>4</sup>. Its biological behavior is intermediate between benign fibrous tissue proliferation and fibrosarcoma. Fibromatosis characteristically is locally invasive and tends to recur, but does not metastasize. Although mesenteric desmoids tumors tend to be aggressive, there is considerable variability in

their growth rate during the course of the disease. In fact, the biology of intra-abdominal desmoid may be characterized by initial rapid growth, followed by stability or even regression. However, mesenteric desmoids, by virtue of its relationship to vital structures and its ability to infiltrate adjacent organs, may cause important complications, including intestinal obstruction, ischemia and perforation, hydronephrosis, and even aortic rupture<sup>5, 6</sup>. Despite these complications, the overall ten-year-survival for patients with intra-abdominal desmoids can be as high as 60 to 70%<sup>7</sup>. The ideal treatment is complete excision<sup>8</sup>. When the lesion involves vital structures and resections become difficult bypass procedures have to be undertaken. Prognosis is good after complete excision. Local recurrences are known which are again to be treated by excision. When the tumor is left behind with a bypass procedure, it takes several years before it requires a second exploration. Subsequent surgery is made difficult by extensive intraperitoneal adhesions due to previous surgery.

Our case illustrates another possible cause of acute abdominal pain. Although rare, fibromatosis should form part of the list of differentials for abdominal masses and pain. Once the diagnosis has been made, the only current treatment option is surgical resection.

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