Mesenteric mass in a 28-year-old woman

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A 28-year-old woman presented with a painless lower abdominal mass for six months. There was no history of previous pregnancy or abdominal surgery. On examination there was a mobile lump in the umbilical region which was nontender and firm with a smooth surface. Haematological and biochemical investigations were within normal limits. Ultrasonography revealed a homogenous solid mass without pressure effects. On exploratory laparotomy through a midline incision a whitish 5-cm round tumour was found in the mesentery of the ileum (figure 1). The mass did not extend up to the serosal surface of the bowel and it was possible to completely excise it.

The cut surface was homogenous, grey/white and had a fibrous look. A photomicrograph of a section from the mass is shown in figure 2.

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Figure 1 The excised tumour

Figure 2 Section from tumour (H & E stain, original $\times 500$)

Question

What is this condition called?

Table Reported cases of primary mesenteric fibromatosis (from⁴)

Ref	Age	Sex	Treatment	Follow-up
1	27	M	complete excision	?
2	34	M	incomplete excision	recurrent disease; died after 3.5 years
4	22	M	complete excision	recurrent disease; died after 6 months
Hailemarien, cited in 4	37	M	complete excision	8 months; no recurrence
Pilegard, cited in 4	42	M	incomplete excision	no recurrence
present case	28	F	complete excision	3 years; no recurrence

Answer

Primary mesenteric fibromatosis. Histological examination revealed a spindle cell tumour of moderate cellularity without atypia, composed of broad bundles and sheets of parallel oriented fibroblasts separated by dense fibrillary collagen (figure 2). The margins of resection were clear. There was no family history of polyposis and sigmoidoscopic examination showed no abnormalities. The patient has been under follow-up for three years, with clinical and repeated ultrasonological examinations, without any evidence of recurrence.

Discussion

Mesenteric fibromatosis, also referred to as ‘desmoid tumour’ of the mesentery, is a rare, histologically benign but locally aggressive non-metastasizing tumour. It is notorious for local recurrence if excision is incomplete.¹ It commonly occurs in association with familial adenomatous polyposis, especially if Gardner’s syndrome is present.^{1–3} The other predisposing causes reported are pregnancy and abdominal surgery or trauma.⁴ Mesenteric fibromatosis occurring in the absence of any of the known predisposing factors is rare. David and Khanduri⁴ called it primary mesenteric fibromatosis and reported five cases from the literature (table). All these patients were males. Mesen-

teric fibromatosis, in general, is more common in women of child-bearing age, as hormonal influence is strongly implicated⁴ and yet ours is the first female case of primary mesenteric fibromatosis. It presents as a mass in the abdomen which is usually asymptomatic,² leading to delayed presentation. Thus, in two of the five previously reported patients, the disease had already spread to retroperitoneal structures.^{2,4} Surgery is the mainstay of treatment and complete excision of the tumour is curative. Inadequate excision results in recurrence within a year as observed in two cases. Both radiotherapy and chemotherapy (cytotoxic and non-cytotoxic) have been used for recurrent and inoperable disease in mesenteric fibromatosis with variable results.⁵ Non-cytotoxic drug therapy with nonsteroidal anti-inflammatory drugs (sulindac and indomethacin) and anti-oestrogenic drugs (tamoxifen) has been reported to be effective in obtaining remission in some cases.^{5–8} These drugs may therefore be useful in cases of primary mesenteric fibromatosis with residual disease after excision.

Final diagnosis

Primary mesenteric fibromatosis.

Keywords: mesenteric fibromatosis, desmoid tumour

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