CASE REPORT: A CASE OF FIBROMATOSIS IN A SIX YEAR OLD WITH COARCTATION OF THE AORTA

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SYNOPSIS

A six year old Malay girl presented with a one week history of abdominal pain. She was found to have coarctation of the aorta. While in the ward she developed an acute abdomen. At the emergency laparotomy, torsion of a rare tumour — a pedunculated mesenteric fibromatosis, with intestinal ischaemia was found. The incidence, pathology, diagnostic investigations and management of mesenteric fibromatosis are reviewed.

INTRODUCTION

Fibromatosis is defined as an infiltrating fibroblastic proliferation; the fibroblasts are well differentiated with little mitotic activity. There is no inflammatory infiltration. They are locally invasive but do not metastasize, a differentiating point from fibrosarcomas.

We describe here a patient with fibromatosis of the mesentery, or mesenteric desmoid. This is an uncommon tumour. Gupta et al (1) found 3 mesenteric tumours in this review of 72 cases of extra-abdominal desmoids.

CASE REPORT

A six year old girl was admitted with abdominal pain for one week. A similar episode occurred one month back. The pain was colicky and occasionally associated with vomiting. Bowel habits and stools were normal.

Examination revealed a thin girl weighing 11 kg. Blood pressure 200/130 mm Hg in the upper limbs and 70 mm Hg systolic in the lower limbs. Femoral pulses were not palpable and there was mild cyanosis of the toes. The apex beat was not displaced. A soft ejection systolic murmur was heard over the pulmonary area.

The abdomen was scaphoid. Except for some voluntary guarding no localised tenderness or abdominal mass was felt. The liver and spleen were not enlarged.

Coarctation of the aorta was confirmed by an arteriogram.

The abdominal pain was attributed to ischaemia of the gut.

On the 16th hospital day, she developed severe abdominal pain and the blood pressure dropped to systolic 80 mm Hg. A vague non-pulsatile mass was felt over the epigastrium 4 cm in diameter.

She was assessed to have an acute abdomen from either a dissection of the aorta or a ruptured aortic aneurysm. At laparotomy, 200 to 300 ml of blood clots were found. The whole of the small bowel extending to the caecum and ascending colon was cyanotic. An infiltrating cystic mass measuring 8 to 10 cm in diameter was found at the root of the mesentery. There was arterial bleeding into the tumour. Enlarged lymph nodes were present at the aortic bifurcation. Because of the infiltrative nature of the tumour, it was deemed inoperable. A biopsy was taken. The child died 2 days later.

POST MORTEM FINDINGS

The body was that of a thin Malay child with a height of 115 cm and weighing 12,700 gms.

Internal examination showed the presence of 200 ml of blood with a few blood clots present within the peritoneal cavity. There was a large haemorrhagic uncircumscribed mass at the root of the mesentery that was adherent to the pancreas and to adjacent loops of small bowel. The mass appeared to infiltrate along the aorta inferiorly to surround the right iliac vessels. The intestines were generally severely congested and contained blood stained fluid within the lumen, features consistent with haemorrhagic infarction.

There was also an area of mild coarctation of the aorta measuring 1.0 cm in circumference at the descending segment of the aortic arch. There was dilatation of the aorta distal to the coarctation with a short length of dissection within the aortic wall. No intimal tear was present.

Histological examination of the mesenteric mass revealed a collagenous tumour showing intense congestion with areas of haemorrhage and infarction (Figure 1). The collagenous tissue had infiltrated into the mesenteric fat and had extended around and into the pancreas (Figure 2). Within the collagenous tissue, there were numerous fibroblasts that were generally benign-looking. No evidence of pleomorphism and no mitotic figures were present. (Figure 3) There was an acute inflammatory response to the infarction.

The cause of death was certified as Mesenteric Fibromatosis with Torsion.

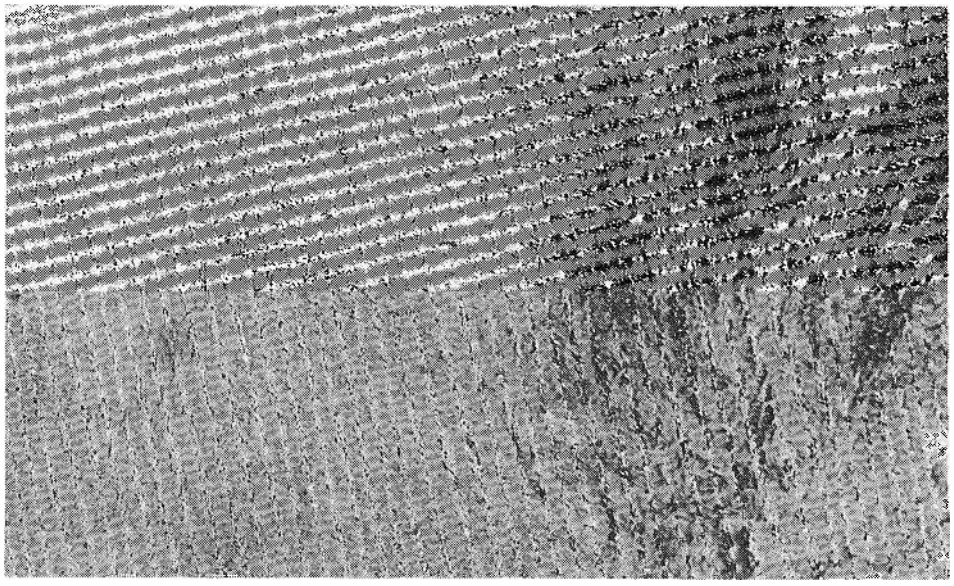

Figure 1: A low power view of the collagenous tumour showing areas of haemorrhage. H&E, ×30

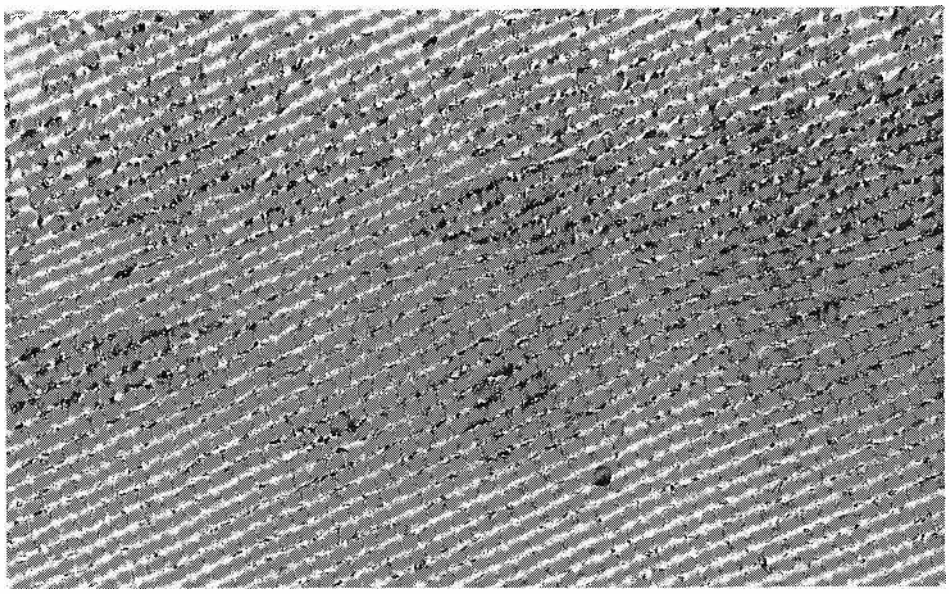

Figure 2: A section taken from the periphery of the pancreas showing the collagenous tissue infiltrating into the pancreas resulting in an entrapped island of pancreatic tissue (left side of the field). H&E, ×75

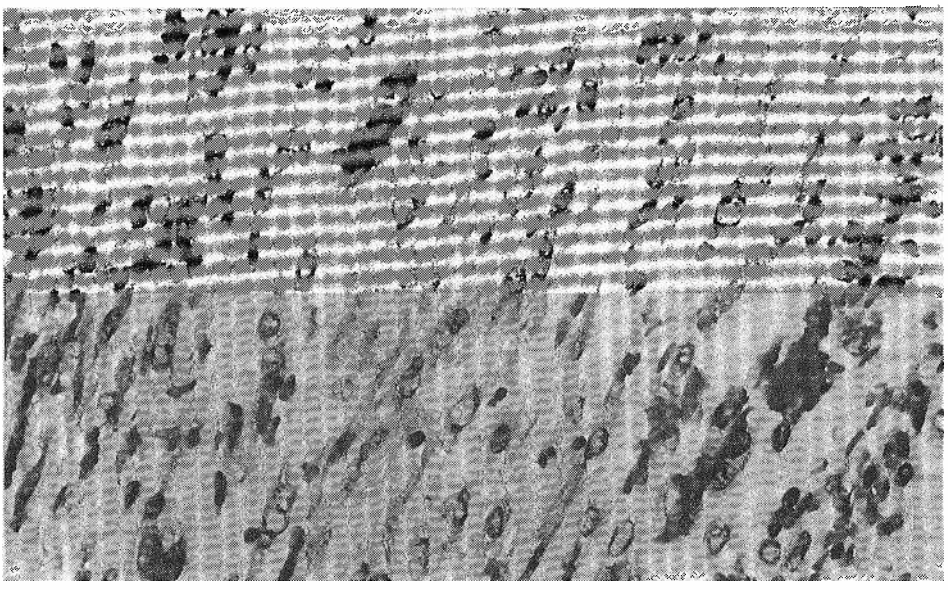

Figure 3: A representative high power field of the tumour showing the fibroblasts and the lack of mitoses. H&E, ×300

DISCUSSION

The fibromatoses can be classified (2) as follows:

I. Superficial (fascial) fibromatoses
   - A. Palmar fibromatosis (Dupuytren's contracture)
   - B. Plantar fibromatosis (Ledderhose's disease)
   - C. Penile fibromatosis (Peyronie's disease)
   - D. Knuckle pads.

II. Deep (musculoaponeurotic) fibromatoses
   - A. Extraabdominal fibromatosis (extra-abdominal desmoid)
   - B. Abdominal fibromatosis (abdominal desmoid)
   - C. Intraabdominal fibromatosis (intraabdominal desmoid)
     1. Pelvic fibromatosis
     2. Mesenteric fibromatosis
     3. Gardner's syndrome

In the classical study of solid tumours of the mesentery by Yannopoulos and Stout, (3) Mesenteric Fibromatosis (MF) was the most frequent histological type: 12 out of the 44 cases reviewed.

The age range varied from 6 days to 73 years; the largest tumour being 50 cm in diameter. They were usually located in the mesentery of the small intestine, occasionally in the mesocolon and gastrohepatic ligament. They did not metastasize but tended to recur following excision.

Apart from isolated tumours, a second group presents in patients with Gardner's Syndrome (4 to 9), a condition characterised by familial multiple polyposis of the colon, osteomas and soft tissue tumours. MF and adenocarcinomatous change of the colon and polyps contribute significantly to the morbidity and mortality of the syndrome.

Mesenteric Fibromatosis can present as a silently growing abdominal mass, colicky abdominal pain, discomfort, altered bowel and urinary habits, weight loss or intestinal obstruction (9,10,11,12). M. Hashomnai (13) described a 14 year old boy with acute torsion of a pedunculated MF which bore some resemblance to our case.

The unusual feature in our patient is the coincidental presence of a coarctation of the aorta which complicated the diagnosis and management.

Investigation such as ultrasound of the abdomen to elicit the cause of the presenting symptoms ascribed to ischaemic pains below the narrowing of the aorta could have led to an earlier diagnosis.

The sudden collapse of the patient resulted from torsion and massive haemorrhage into the tumour. Unfortunately the already compromised blood supply to the gastrointestinal tract (the small and large intestine was cyanotic at laparotomy) made surgical management in an emergency situation extremely difficult.

DIAGNOSTIC INVESTIGATIONS

Plain abdominal X-Rays may reveal sizeable tumours as a radiological mass (14). Compression and angulation of the small bowel loops, sometimes 'pseudo diverticula', may be seen on a barium study. Mesenteric arteriography (14) may reveal displacement, separation and angulation of the major and minor branches of the superior mesenteric artery.

Ultrasound (14,15) is a useful diagnostic tool. The tumour is seen as a hypo-echoic mass containing scattered high amplitude echoes. CAT scans (16) have been used to define the extent, subsequent growth of the tumour post operatively and response to therapy.

TREATMENT

The treatment of choice for mesenteric fibromatosis is complete surgical excisioin (1,16), although this is not always possible. Bypass procedures are done when vital organs are involved. There is a high recurrence rate with incomplete excision and subsequent surgery is associated with increasing difficulty, morbidity and mortality because of fibrous adhesions and local invasion.

Irradiation (14,17) has been used with mixed results. Other treatment modalities include chemotherapy. Drugs that affect the metabolism of cyclic 3', 5' adenosine monophosphate (18): testolactone, theophylline and chlorthiazide appeared useful. Sulindac 5, a nonsteroidal antiflammatory compound which inhibits prostaglandin synthesis is being investigated.

CONCLUSION

Mesenteric fibromatosis is an uncommon tumour which although histologically benign, can have a devastating effect on a patient because of its locally invasive nature, and its asymptomatic growth to a large size, leading to pressure effects. We present a patient with acute abdomen due to torsion of the tumour where diagnosis and management was made difficult because of a coincidental coarctation of the aorta.

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