Self-assessment corner

Intestinal obstruction in an adolescent female

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A 16-year-old girl was admitted with a history of abdominal pain, recurrent vomiting and abdominal distension for the last three years. On examination there was a diffuse lump occupying the central abdomen. Sigmoidoscopy showed normal mucosa till 20 cm. Apart from a polymorphonuclear leucocytosis (total white cell count of $25 \times 10^9/l$ with 80% polymorphs), other laboratory investigations were within normal limits. A barium enema revealed incomplete filling of the colon proximal to the rectosigmoid junction. A laparotomy, with a provisional diagnosis of subacute intestinal obstruction, was undertaken. The entire small intestine, caecum and colon up to the rectosigmoid junction were found to be encased in a thick fibrous membrane. This membrane was completely excised (figure 1) and lysis of the inter-loop adhesions was performed. The postoperative recovery was uneventful.

A photomicrograph of a H & E-stained section from the excised membrane is shown in figure 2.

Figure 1

Figure 2

Question

What is this condition called?

Answer

Abdominal cocoon. Histological examination of the excised cocoon wall, showed characteristic neurofibromatous components seen as twisted wire-like neurofibrils (figure 2). Interspersed in between were variable amounts of collagen bundles.

Abdominal cocoon, or small bowel obstruction due to a membranous encasement, is a rare clinical entity. It has been described predominantly in adolescent females residing in tropical and subtropical areas.^{1,2} The aetiology of this condition remains obscure, although an infective pathology is suspected,¹ due to retrograde peritonitis through the fallopian tube. Histological examination of the excised membrane usually reveals fibrous tissue only, with no evidence of inflammation or granuloma formation.^{1,3}

Clinically, the present case had many similarities to previously described cases of abdominal cocoon.^{1,2} The patient was an adolescent female from a tropical area. However in the previously described cases, only the small bowel was encased, completely or partly,¹ whereas, in the present case the entire small bowel and colon up to the rectosigmoid junction were involved.

Known causes of sclerosing peritonitis, resulting in a membranous encasement of the bowel are the use of β -blockers, especially practolol,⁴ chronic ambulatory peritoneal dialysis,⁵ use of a LeVeen shunt,⁶ talc granuloma from a previous laparotomy⁷ and liver cirrhosis³ (box).

A similar clinical presentation is seen in the congenital condition of ‘peritoneal encapsula-

Differential diagnosis of membranous encasement of bowel

- abdominal cocoon
- congenital peritoneal encapsulation
- sclerosing peritonitis: due to β -blockers, use of LeVeen shunt, talc granuloma from previous laparotomy, chronic ambulatory peritoneal dialysis, or in liver cirrhosis
- intra-abdominal plexiform neurofibroma (present case)

tion’.² There is an accessory membrane, derived from the peritoneum of the yolk sac,¹ present in front of the small bowel, which can be easily removed. However, this condition is completely asymptomatic and is found incidentally at laparotomy or autopsy.

Neurofibromas, though rare, are the most common tumours involving nerves. They may arise in any nerve containing Schwann cells. Intra-abdominal neurofibromas are characteristically retroperitoneal, although plexiform neurofibromas involving the intestinal mesentery and the liver have been described.⁸ However, to our knowledge, this is the first reported case of an intra-abdominal neurofibroma presenting as an abdominal cocoon.

Final diagnosis

Intra-abdominal neurofibroma presenting as an abdominal cocoon.

Keywords: abdominal cocoon, neurofibroma

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