CASE REPORT

Idiopathic sclerosing encapsulating peritonitis presenting as a right iliac fossa mass in a teenaged girl

Hafez Mohammad Ammar Abdullah,¹ Muhammad Asim Shahzad,² Sajjad Ullah,³ Waqas Ullah¹

¹Department of Internal Medicine, Khyber Teaching Hospital, Peshawar, Khyber Pakhtunkhwa, Pakistan

²Department of Internal Medicine, Hayatabad Medical Complex, Peshawar, Khyber Pakhtunkhwa, Pakistan

³Department of General Surgery, Khyber Teaching Hospital, Peshawar, Pakistan

Correspondence to

Dr Hafez Mohammad Ammar Abdullah, dr.ammarabdullah@gmail.com

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SUMMARY

Sclerosing encapsulating peritonitis (SEP) is a rare peritoneal condition characterised by a firm and thick membrane that encases a part of or the whole of the small intestine. It can be idiopathic or secondary to a variety of conditions. It usually presents as an acute or subacute small bowel obstruction, however, the presentation can vary. It may be diagnosed by radiology, although definitive diagnosis is by laparoscopy or laparotomy. It is treated by excision of the covering membrane. We report a case of a teenaged girl who presented with abdominal pain and a palpable mass in the right iliac fossa. The patient underwent emergency laparotomy after her work up was inconclusive, and was found to have a case of SEP. The distal ileum and the sigmoid colon appeared to be covered by a thick membrane, which was excised. Follow-up of the patient was unremarkable.

BACKGROUND

The term sclerosing encapsulating peritonitis (SEP) describes a condition in which the small bowel, especially the distal parts, gets covered by a thick fibrous sheath. It can be idiopathic or secondary to a variety of underlying conditions, including previous abdominal surgery, pelvic inflammatory disease, peritoneal dialysis, disseminated tuberculosis and sarcoidosis. It most commonly presents as an acute or subacute intestinal obstruction, usually with a chronic history, but the presentation varies widely. Rarely, it has been known to present as a right iliac fossa mass. As such, it often offers a diagnostic dilemma, as diagnosing it preoperatively can be quite difficult. Radiology can be helpful, with the diagnosis in some cases made by CT scan. However, it is still diagnosed in many cases by laparoscopy or exploratory laparotomy. Regardless of how it is diagnosed, it is treated with surgical excision and adhesiolysis.

CASE PRESENTATION

A 15-year-old girl presented to the gynaecology department of our tertiary care centre, with a history of lower abdominal pain intermittently for the past 5–6 months. The pain was insidious in onset, dull in character, non-radiating and mild in intensity. There were no aggravating factors and pain responded temporarily to over-the-counter pain medications. The patient remained in the gynaecology unit for 3 days. Work up was carried

out to determine the cause of her pain. Her baseline investigations and abdominal sonography were normal. She improved symptomatically and was discharged after she refused further work up. However, she experienced a sudden exacerbation of pain 15 days after being discharged. She was readmitted at a surgical unit, on suspicion of acute abdomen. On admission, the pain was moderately severe and present throughout the lower abdomen; this was associated with nausea, vomiting and retching. Her bowel movements were also normal. She had no significant medical nor surgical history and had regular menstrual flow, with her past menses 15 days prior.

On general physical examination, she was vitally stable with blood pressure of 130/90 mm Hg, pulse of 90 bpm and temperature of 98°F. She appeared dehydrated and in discomfort. She was pale looking, but the rest of the general physical examination was normal.

On abdominal inspection, neither distention nor mass was apparent. Palpation showed a diffusely tender abdomen with marked tenderness in the right iliac fossa. There was a 7×4 cm palpable mass in the right iliac fossa. The mass was soft and doughy in consistency, with regular margins and normal overlying skin. The rest of the patient's systemic examination was unremarkable.

INVESTIGATIONS

The laboratory tests showed normal blood counts, with a haemoglobin level of 11 g/dL, total leucocyte count of 6400/μL and platelets 190 000/μL. Erythrocyte sedimentation rate was 10 mm/h. Serum electrolytes, urea, creatinine and liver function tests were all in the normal range. Erect X-ray of the abdomen was carried out, which showed no apparent abnormality. Abdominal sonography showed mildly distended bowel loops. Neither abdominal CT scan nor MRI could urgently be arranged.

The patient had also been investigated during her first admission. At that time, her complete blood count, renal function tests, liver function tests, serum electrolytes, and urinalysis were normal. Her abdominal ultrasound was also normal.

DIFFERENTIAL DIAGNOSIS

On presentation, the patient was suspected to be suffering from acute appendicitis. When her abdominal examination revealed a right iliac fossa

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mass and tenderness, she was suspected to be suffering from an appendicular abscess or rupture. The other differentials were adnexal torsion, ovarian cyst rupture and tubo-ovarian abscess.

TREATMENT

The patient underwent exploratory laparotomy through a midline incision on the day of her admission. The distal ileum starting from the ileocaecal junction and extending up to 3 feet proximally appeared to be grossly distended and covered by a thick fibrous sheath (figure 1). Adhesiolysis was performed on the numerous adhesions that were seen. After the fibrous sheath was slowly removed, the underlying gut was found to be healthy and viable (figure 2). Biopsy was taken from one of the mesenteric lymph nodes and sent for histopathology. The sigmoid colon was also covered with a thick fibrous sheath and fixed to the posterior abdominal wall, which is unusual for this condition, as it most commonly involves only the small intestine. The sigmoid colon was also freed and made mobile. Haemostasis was secured, a drain placed and the abdomen was closed.

OUTCOME AND FOLLOW-UP

The patient was managed with postoperative care for another 3 days in the hospital. The lymph node sample was sent for biopsy, which turned out to be normal. The patient was sent home on oral medications, including antibiotics and painkillers, and advised follow-up after 1 month.

The patient came for follow-up 1 month after her surgery. She had no specific abdominal symptoms and her incision wound was clean and well healed.

DISCUSSION

SEP was first described in young females by Foo *et al*¹ in 1978. The idiopathic form, also referred to as an abdominal cocoon, is usually found in young females, mostly in tropical and subtropical regions, as was our patient, with very few cases reported from temperate regions.² The incidence of idiopathic SEP is not known due to a paucity of cases. Its aetiology and pathogenesis is not well known but it has been theorised as being associated with retrograde menstruation or gynaecological infections causing subclinical peritonitis.^{1 3} However, it has since been described in males as well and that hypothesis is no longer considered valid for all cases.^{3 4} Secondary SEP, on the other hand, has been associated with a variety of causes, including peritoneal dialysis,^{5 6} previous abdominal surgery, disseminated tuberculosis,⁷ peritoneoventricular shunt, recurrent peritonitis, peritoneovenous shunting for refractory ascites,⁸ sarcoidosis and the use of the β -blocker, practolol.⁹ A secondary cause must be actively sought in each case, as SEP has been

Figure 2 Removal of the fibrous sheath.

known to recur if the cause is not treated. In our case, we could not find any secondary cause.

Idiopathic SEP is a rare condition that most commonly presents as intestinal obstruction, however, the presentation can vary widely and it can be asymptomatic as well. It is known to occasionally present as an abdominal mass, as in our case, which presented as a right iliac fossa mass.^{3 10} This can complicate the diagnosis as it is not a very common cause of an abdominal mass. As such, it must be differentiated from the more common causes of abdominal and right iliac fossa masses such as an appendicular abscess or malignancy, a tubo-ovarian pathology, psoas abscess, tumour of the ascending colon or caecum, ileocaecal tuberculosis or ileocaecal intussusception. It is an important differential to keep in mind when a patient presents with chronic episodes of abdominal pain or features of intermittent intestinal obstruction.

However, preoperative diagnosis of SEP remains a challenge and it is still diagnosed in many cases on exploratory laparotomy. Radiology does not always pick it up, as in our case. Abdominal X-rays may show signs of intestinal obstruction, while an ultrasound may detect fluid collection or any mass produced by the clumping together of the small intestine. In our case, the sinologist could not pick it up as he reported excessive bowel gases obscuring sonographic details. A barium follow through may show a delayed transit of contrast through the bowel and clumping together of the small intestine in the middle of the abdomen.^{3 11} This clumping together may give an appearance of a cauliflower and has been termed the cauliflower sign. It may be diagnosed with a CT scan, which may also show a sheath encompassing and clumping together distended loops of the small intestine in the centre of the peritoneal cavity.¹¹ However, this is not always the case, and CT and MRI are non-specific in many cases. Our patient presented in emergency and a barium follow through or a CT or MRI, could not be arranged

Figure 1 A fibrous sheath encasing the distal ileum.

Learning points

- ▶ Sclerosing encapsulating peritonitis (SEP) is a condition in which part of the small intestine gets covered by a thick fibrous sheath.
- ▶ SEP can be idiopathic or secondary to a wide range of conditions, most commonly peritoneal dialysis.
- ▶ SEP can present with chronic abdominal pain and intermittent intestinal obstruction or, rarely, as a right iliac fossa mass.
- ▶ Greater emphasis must be laid on the preoperative diagnosis of SEP, as it can help reduce complications during surgery.

preoperatively. Definitive diagnosis is by laparoscopy or laparotomy, as in our case.

The treatment of SEP includes adhesiolysis and complete removal of the sheath.^{3 8} Resection of the bowel is associated with many complications and as such is indicated only if a segment of the small bowel becomes non-viable. Our patient had no necrotic bowel segments, and so did not require any bowel resection. Asymptomatic cases do not require any surgical intervention.^{11–13}

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Competing interests None declared.

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REFERENCES

- 1 Foo KT, Ng KC, Rauff A, *et al.* Unusual small intestinal obstruction in adolescent girls: the abdominal cocoon. *Br J Surg* 1978;65:427–30.
- 2 Yoon YW, Chung JP, Park HJ, *et al.* A case of abdominal cocoon. *J Korean Med Sci* 1995;10:220–5.
- 3 Tu JF, Huang XF, Zhu GB, *et al.* Comprehensive analysis of 203 cases with abdominal cocoon. *Zhonghua Wei Chang Wai Ke Za Zhi* 2006;9:133–5.
- 4 Al-Ani AH, Zayani NA, Najmeddine M, *et al.* Idiopathic sclerosing encapsulating peritonitis (abdominal cocoon) in adult male. A case report. *Int J Surg Case Rep* 2014;5:735–8.
- 5 Afthentopoulos IE, Passadakis P, Oreopoulos DG, *et al.* Sclerosing peritonitis in continuous ambulatory peritoneal dialysis patients: one center's experience and review of the literature. *Adv Ren Replace Ther* 1998;5:157–67.
- 6 Holland P. Sclerosing encapsulating peritonitis in chronic ambulatory peritoneal dialysis. *Clin Radiol* 1990;41:19–23.
- 7 Kaushik R, Punia RP, Mohan H, *et al.* Tuberculous abdominal cocoon—a report of 6 cases and review of the literature. *World J Emerg Surg* 2006;1:18.
- 8 Stanley MM, Reyes CV, Greenlee HB, *et al.* Peritoneal fibrosis in cirrhotics treated with peritoneovenous shunting for ascites: an autopsy study with clinical correlations. *Dig Dis Sci* 1996;41:571–7.
- 9 Eltringham WK, Espiner HJ, Windsor CW, *et al.* Sclerosing peritonitis due to praxolol: a report on 9 cases and their surgical management. *Br J Surg* 1977;64:229–35.
- 10 Sahoo SP, Gangopadhyay AN, Gupta DK, *et al.* Abdominal cocoon in children: a report of four cases. *J Pediatr Surg* 1996;31:987–8.
- 11 Hur J, Kim KW, Park MS, *et al.* Abdominal cocoon: preoperative diagnostic clues from radiologic imaging with pathologic correlation. *AJR Am J Roentgenol* 2004;182:639–41.
- 12 Célécourt B, Levard H, Hay J, *et al.* Sclerosing encapsulating peritonitis: early and late results of surgical management in 32 cases. French Associations for Surgical Research. *Dig Surg* 1998;15:697–702.
- 13 Serafimidis C, Katsarolis I, Vernadakis S, *et al.* Idiopathic sclerosing encapsulating peritonitis (or abdominal cocoon). *BMC Surg* 2006;6:3.

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