Peritoneal encapsulation syndrome: A case report and literature review

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A B S T R A C T

INTRODUCTION: Peritoneal encapsulation is an infrequently described congenital anomaly that results in formation of an accessory peritoneal membrane. The case presented below is unique in that it illustrates one of the rare complications of this condition. It is important for clinicians to be aware of this condition and its complications in order to limit potential morbidity and mortality.

PRESENTATION OF CASE: We report on an eleven-year-old boy without prior abdominal symptoms who presented with an acute abdomen after an episode of intense physical exertion. At laparotomy, gangrenous small bowel loops were identified extruding from an opening in a peritoneal sac consistent with peritoneal encapsulation syndrome. All gangrenous bowel (mostly ileum) was resected. The sac was excised and a primary jejunum to ascending colon anastomosis was created. The patient did well post operatively and was subsequently discharged.

DISCUSSION: Peritoneal encapsulation is an aberration of peritoneal development that is frequently confused with other visceral encapsulation syndromes of inflammatory origin. Due to its mostly asymptomatic course, its true incidence remains unknown. An appreciation of the condition and its potential complications allows surgeons to take appropriate action in the event of incidental discovery at laparoscopy or laparotomy.

CONCLUSION: Peritoneal encapsulation is a rare, mostly asymptomatic, surgical finding which may predispose patients to an acute abdominal crisis.

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1. Introduction

This case has been reported in line with the Surgical Case Report Guidelines (SCARE) criteria [1]. Peritoneal encapsulation syndrome is a rare consequence of abnormal peritoneal development [2]. Because of how infrequently the condition occurs, and similarity with other similar intra-abdominal pathology, it has been erroneously referred to by several different terms. These include abdominal cocoon syndrome and secondary sclerosing encapsulating peritonitis [3]. However, peritoneal encapsulation syndrome differs from the latter conditions because of the total or partial encapsulation of the small bowel within a mesothelium-lined membrane. As of 2012, less than 60 cases had been reported in literature [4]. Most reports describe complications secondary to this anatomic aberration [4]. It is likely the true incidence of peritoneal encapsulation syndrome is actually higher as asymptomatic cases are unlikely to be reported. The case presented below is unique in that it shows one of the rare complications of such a condition. It is important for clinicians to be aware of this rare condition as well as its complications so as to reduce the potential morbidity that might result from unanticipated discovery.

We report on an eleven-year-old boy without prior abdominal symptoms who presented with an acute abdomen at a public referral hospital. During laparotomy, gangrenous small bowel loops were identified extruding from an opening in a peritoneal sac consistent with peritoneal encapsulation syndrome.

2. Case report

An 11-year-old boy with a one-day history of acute abdominal pain was brought to a public referral teaching hospital by the mother. The pain started abruptly while he was pushing another boy in a wheelchair over uneven ground. The pain was non-colicky and most prominent in the infra-umbilical region; the intensity of the pain worsened after onset. This was associated with intermittent projectile vomiting without pain relief. There was no fever or chills, nor did the patient complain of associated diarrhoea or
bloody stool. The patient had been otherwise healthy, without any contributory medical or surgical history. He had no relevant drug, family, psychosocial or genetic history.

On examination the child appeared septic, lethargic and severely dehydrated. He had a temperature of 37.5°C, and a pulse of 157 bpm, with a blood pressure of 97/41 mmHg. His abdomen was moderately distended with generalised guarding and rebound tenderness. Digital rectal examination and assessment of the other systems was unremarkable.

Serum biochemistry suggested dehydration with a blood urea of 10.3 mmol/l (2–6.7) and serum creatinine of 87 umol (98–131). He had an elevated white cell count of 14,700/mm³ (4–11) and a haemoglobin of 16.8 g/dl (15 ± 1.7).

The patient received fluid resuscitation, was catheterised, and a nasogastric tube was inserted. Oxygen was administered per face mask and patient taken for surgery.

At laparotomy, gangrenous loops of small bowel were encountered. At the base of the non-viable bowel was what was suspected to be a large 8 x 8 cm mesenteric cyst. No normal small bowel was visible within the abdominal cavity. However, on closer inspection, it was evident that the ischemic loops of bowel were originating from an opening in the 'mesenteric cyst' within the vicinity of the ileo-cecal junction. The ‘cyst’ was then opened and viable small bowel loops were discovered within, in association with a clearly gangrenous loop of terminal ileum. (Figs. 1–3)

All gangrenous bowel (which constituted most of the distal small bowel excluding an 80 cm remnant of proximal jejunum, the ileocecal valve, and the cecum) was resected. The sac was noted to extend between the ascending and descending colon laterally, the transverse mesocolon superiorly and to the suprapubic region inferiorly and was completely excised. A primary jejunum to ascending colon end-to-end anastomosis was created. The abdomen was irrigated and closed primarily. The operation was done by two surgical trainees. Intra-operatively, patient became haemodynamically unstable and inotropic support was required. Post intervention concerns were of sepsis, anastomotic leak and short bowel syndrome. Post-operatively, the child was managed in the intensive care unit where inotropic support was weaned off and the child extubated on the 4th post-operative day. Broad spectrum antibiotics were continued. Regular abdominal examination showed no signs of an anastomotic leak. He was discharged on post-operative day 9 with plans for continued monitoring in an outpatient setting. Six months post-operatively, the child was discharged from follow-up care without clinical evidence of short-bowel syndrome or other significant morbidity. The patient and mother were grateful to the surgical team. Histology results were not available at the time of write-up.

3. Discussion

The term peritoneal encapsulation (PE) is often used interchangeably with abdominal/small bowel cocoon syndrome and sclerosing encapsulating peritonitis (SEP). These are however, terms used to describe three distinct entities with differing pathogenesis, as described by Akbulut in 2015 [3, 5].

Peritoneal encapsulation is a rarely described pathology resulting from aberrant fetal development, often only diagnosed incidentally [4]. Because of its rarity, only case reports have been described in literature and the epidemiology is still largely undefined. It has been noted to occur in all age groups, with the oldest patient described in literature being 82 years of age [5]. Cleland in 1868 first described PE as a congenital anomaly that arises in the 12th week of development as bowel loops return to the abdomen after physiological herniation and become enveloped in the dorsal yolk sac mesentery that ordinarily forms the transverse mesocolon, thus forming the characteristic peritoneal sac [2]. The sac is typi-
cally described as encasing part of, or the entire small bowel from the ligament of Treitz to the ileo-colic region. Peritoneal encapsulation is an anatomical anomaly, in contrast to SEP or abdominal cocoon syndrome, which are of inflammatory origin.

Patients with peritoneal encapsulation are typically asymptomatic [4–7]. The anomaly is usually identified in one of several ways. It is either discovered incidentally at laparotomy for other indications or post-mortem, or in patients who present with an acute abdomen for small bowel obstruction or strangulation [4–7]. Naraynsingh et al. noted the presence of fixed asymmetrical abdominal distension and differences in the consistency of the abdominal wall on examination in a patient presenting with small bowel obstruction who was later noted to have PE at laparotomy [7]. Radiological assessment may be normal or may show non-specific features of intestinal obstruction. On CT scan, a helical arrangement of small bowel loops encased within a membrane may be observed, known as the Helix Sign [8].

Features at laparotomy that distinguish peritoneal encapsulation from abdominal cocoon syndrome and secondary sclerosing encapsulating peritonitis include the ability to separate the membranous layer easily from the underlying loops of bowel. The extent of the membrane is also typical, covering all or part of the small bowel loops from the ligament of Treitz to the ileo-cecal region, and lying between the meso-colon and the parietal peritoneum inferiorly. The bowel loops within the capsule are also noted to lie freely without adherence to each other [3]. (see Table 1 below)

Based on our patient’s history and the findings at surgery, we made a diagnosis of peritoneal encapsulation. This is the first reported case in Zimbabwe. The patient’s history of present illness is typical of this condition. Abdominal examination findings were however not as described by Naraynsingh [7]; our patient had overt generalised peritonitis from strangulated small bowel.

The surgical findings suggest that this was a patient who had initial asymptomatic peritoneal encapsulation with all small bowel contained within the membranous sac. It is postulated that an acute increase in intra-abdominal pressure related to the patients physical activity may have resulted in a loop of ileum herniating through an opening at the ileo-cecal border of the sac and subsequently becoming strangulated. This would explain the surgical findings of gangrenous small bowel outside of the sac communicating through a tight aperture to viable bowel within. The lack of adhesions between loops of bowel and the presence of sac underscore this was indeed peritoneal encapsulation and not sclerosing encapsulating peritonitis.

The extent of gangrene necessitated an extensive resection of all ileum leaving a remnant 80 cm of jejunum. The length of resected bowel and the loss of the ileo-cecal valve put the patient at risk of short bowel syndrome. However, the patient’s age and the normal remnant small and large bowel were favourable for avoiding short bowel syndrome. The patient developed diarrhoea initially which eventually resolved. Because of the loss of terminal ileum, it was important to consider the likelihood of vitamin B12 deficiency, fat malabsorption and anaemia in the follow up management of this patient.

4. Conclusion

Peritoneal encapsulation is a rare mostly asymptomatic surgical finding which may predispose patients to an acute abdominal crisis. In the event of complications, application of basic surgical principles allows the unanticipating surgeon to take appropriate corrective surgical action. Future development of clinical and imaging diagnostic criteria may one day allow for an accurate pre-operative diagnosis to become a possibility.

Conflicts of interest

There is no conflict of interest.

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Ethical approval

Consent obtained from the parent.

Consent

Signed consent obtained from the parent.

Author contribution

Chenesa Mbanje - case report design, subject research, consent and writing.
Dennis Mazingi - case report design and writing.
Joseph Forrester- case report design, writing and editing.
Simbarashe Gift Mungazi - case report design, writing and editing.

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