Congenital Peritoneal Encapsulation: Radiological Imaging Findings of A Rare Clinical Entity - A Case Report

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ABSTRACT

Congenital peritoneal encapsulations are an extremely uncommon pathology that is characterized by encapsulation of the small bowel loops by an accessory peritoneal membrane. It is commonly asymptomatic and often an incidental diagnosis during surgery or autopsy. This condition may be difficult to detect by radiological imaging and consequently remains largely underdiagnosed. We present the case of a 14 year old male patient who presented to the Department of Radiology with a history of recurrent abdominal pain and his abdominal computed tomography scan revealed features of congenital peritoneal encapsulation. This study aims to highlight the radiological features of congenital peritoneal encapsulation and to emphasize the need for radiologists and clinicians to include this entity in their list of differential diagnosis when assessing small bowel obstruction.

CASE REPORT

INTRODUCTION

Congenital peritoneal encapsulation (CPE) is an extremely uncommon pathology that is characterized by encapsulation of the small bowel loops by an accessory peritoneal membrane which has been attributed to abnormal gut rotation during embryological development [1]. The membrane partially or completely envelopes the small bowel [2,3]. It occurs more frequently in young male patients [2,4] and commonly asymptomatic but may seldom present with recurrent abdominal pain and small bowel obstruction [3]. Other infrequent conditions that can cause small bowel encapsulation include abdominal cocoon(AC) and encapsulating peritoneal sclerosis(EPS).

Given the fact that CPE is asymptomatic, it is commonly an incidental diagnosis during surgery or autopsy [2,3]. This condition may be difficult to detect by radiological imaging hence, it remains largely underdiagnosed [1]. Plain radiograph is often normal or may show evidence of a small bowel obstruction. Computed tomography (CT) appears to be a promising tool for preoperative diagnosis and making a differential diagnosis [5].

The CPE is a rare clinical entity and is often not considered in the differential diagnosis of recurrent abdominal pain and small bowel obstruction. Accurate diagnosis and subsequent patients' treatment with prompt surgical intervention has shown good postoperative outcome. This study aims to highlight the radiological features of CPE and to emphasize the need for radiologists and clinicians to include this entity in their list of differential diagnosis when assessing small bowel obstruction.

Case presentation

A 14 year old male patient referred from emergency department(ED) to the department of radiology for abdominal computed tomography (CT) with a history of recurrent abdominal pain and abdominal bloating. He has a history of repeated hospital visits on account of the same complaint. Laboratory investigations were unremarkable. No history of previous surgery or any known chronic medical condition.

The initial plain abdominal radiograph of the patient revealed evidence of small bowel obstruction with a dilated small bowel loops and multiple air-fluid levels predominantly in the upper abdomen (Figure 1). Post intravenous (IV) contrast CT scan of the abdomen also revealed features of small bowel obstruction with a thin membrane surrounding a cluster of dilated small bowel loops in the left abdominal quadrants as well as small amount of fluid within the peritoneal sac. The peritoneal sac, only partially envelope the small bowel in the left abdominal quadrants, but in the right quadrants there is normal caliber ileum outside the sac. Also, there is posterior displacement of mesenteric vessels behind the sac. No evidence

of bowel ischaemia or perforation (Figures 2-4). Based on these findings a diagnosis of internal hernia was made by the reporting Radiologists.

Consequently, the patient was taken to the operating theater for diagnostic laparoscopy, followed by an open surgery which confirms the presence of a thin membrane encasing aggregated small bowel loops in the left abdominal quadrant. The patient recovered promptly following the surgery and was discharged to follow-up on out-patient bases.

Histopathologist report

Nature of Specimen: Intraperitoneal sac surrounding the small bowel.

Gross Description: Sac-like tissue with attached fibrofatty tissue 6cm in aggregate, representative of section submitted in one block.

Microscopic Description: Section shows a proliferation of fibro-connective tissue composed of sheets of dense collagenous tissue with neutrophils and mononuclear inflammatory infiltrates.

Given the clinical presentation, age of the patient, radiological imaging and surgical findings as well as histopathological report a final diagnosis of congenital peritoneal encapsulation was established.

DISCUSSION

Congenital peritoneal encapsulation(CPE) is an infrequent congenital malformation in which an accessory peritoneal membrane derived from the peritoneum of the yolk sac partially or completely envelope small bowel loops [2]. This malformation is believed to occur around the 12th week during embryogenesis and is blamed on an abnormal return of the physiological umbilical hernia, containing the midgut, to the peritoneal cavity [2,6]. Cleland was the first to describe this pathological entity in 1868 as a rare congenital malformation resulting from development of an accessory peritoneal membrane partially or completely encasing the small intestine [7].

The actual incidence and prevalence of CPE is difficult to quantify given the rarity of this condition. It is also difficult to differentiate it from other entities associated with peritoneal encapsulation such as AC and EPS [4]. Nonetheless CPE has a very low incidence and is often discovered incidentally, either intra-operatively or during autopsy. CPE does not have any preference towards any particular ethnicity [2]. It occurs more frequently in young male patients [2,4]. Most cases are asymptomatic but may seldom present with repeated episodes of nonspecific abdominal pain and recurrent small bowel obstruction [3]. The index case was that of a young male who presented to the emergency department on many occasions with a history of recurrent abdominal pain and CT revealed features of localized aggregated small bowel obstruction within a thin peritoneal membrane.

Preoperative diagnosis is challenging as is rarely complicated by intestinal obstruction. However, radiological imaging findings may point to the diagnosis. Plain radiograph is usually normal or nonspecific unless complicated by small bowel obstruction. When there is small bowel obstruction, plain radiographs may be able to detect the dilated small bowel loops or multiple air-fluid levels. CT may reveal aggregation of small bowel loops encased by thin peritoneal membrane and signs of intestinal obstruction [2,3]. The spiral pattern of the distended small bowel loops in CPE has been described by some authors as a helix sign [2, 8]. CT is the imaging modality of choice for the diagnosis of CPE. In the case under review, CT demonstrated an accessory peritoneal membranous sac that encapsulates dilated and clustered small bowel loops which was confirmed during surgery.

The differential diagnosis of CPE includes abdominal cocoon (AC), sclerosing encapsulating peritonitis (SEP) and Internal Hernia (IH). All these conditions may lead to small bowel obstruction and establishing the definitive diagnosis may be challenging. The term CPE, AC, and SEP have been erroneously used exchangeably but they are actually different pathological disease entities [9]. SEP is characterized by complete or incomplete encasement of the small bowel within a thick fibrocollagenous membrane and is a rare benign cause of acute or subacute bowel obstruction. The etiology is unclear and maybe grouped into primary (idiopathic) or secondary. The primary form of SEP is also known as abdominal cocoon syndrome. The secondary form of SEP is due to any condition that causes peritoneal inflammation and induces intra-abdominal fibrosis. This type is also known as encapsulating peritoneal sclerosis (EPS).

Abdominal cocoon (primary sclerosing encapsulating peritonitis) is a rare idiopathic acquired condition associated with abnormal thick membrane encapsulating the intestine and presenting with clinical signs of bowel obstruction [3]. Abdominal cocoon is common in young girls in the tropical regions and usually presents with acute or chronic bowel obstruction [10].

Encapsulating peritoneal sclerosis (secondary form of sclerosing encapsulating peritonitis) is a complication of abdominal tuberculosis, prolonged use of ambulatory peritoneal dialysis, peritoneovenous or ventriculoperitoneal shunts, previous abdominal surgery, retrograde menstruation, systemic lupus erythematosis, sarcoidosis, familial Mediterranean fever, fibrogenic foreign materials and beta-blockers (eg. practolol) [11,12]. The peritoneal membrane in this case is thickened with associated bowel wall thickening as well as peritoneal or mural calcification. The peritoneum shows enhancement with contrast [13]. This condition is usually asymptomatic but may occasionally present with acute or chronic symptoms of intestinal obstruction. Contrast enhanced CT findings of cluster of dilated small bowel loops in a narrow base surrounded by

a thick membrane seen in patients with EPS or AC has been christened the "cauliflower sign" [14].

CPE on the other hand, is an exceedingly developmental abnormality arising during embryogenesis by the 12th week of gestation and is characterized by the development of a thin accessory peritoneal membrane that partially or completely surrounds the small bowel loops [2,3]. This condition is generally asymptomatic but may rarely be associated with small bowel obstruction.

Congenital peritoneal encapsulation also needs to be differentiated from internal hernias. Internal hernias are protrusions of small bowel through anatomic defects, such as foramina or recesses within the mesentery or peritoneum but remaining within the abdominal cavity. In CPE, the sac displaces and splays the mesenteric vessels posteriorly while internal hernia displaces mesenteric vessels anteriorly. The membranous structure encasing the intestine is not present. However, preoperative diagnosis can be difficult and definitive diagnosis can only be made during surgery [15].

CONCLUSION

CPE is a rare congenital anomaly which remains poorly understood, and underdiagnosed. It is a_salient entity that also needs to be considered in patients with enduring, recurrent abdominal pain and small bowel obstruction. CT remains the imaging modality of choice for the diagnosis of CPE. Knowledge of CPE is essential for surgeons as part of the differential diagnosis of intestinal obstruction to avoid intraoperative dilemma especially when other causes have been excluded. Radiologists should be familiar with the clinical and related imaging features of CPE and know when to look for them in order to facilitate timely and proper patient management.

TEACHING POINT

This study highlights the radiological features of congenital peritoneal encapsulation and emphasizes the need for radiologists and clinicians to include this entity in their list of differential diagnosis when assessing small bowel obstruction.

AUTHORS' CONTRIBUTIONS

Khalid Abdalla and Awajimijan Mbaba were responsible for writing the original draft.

Awajimijan Mbaba and Hamza Ahmed were involved in writing review and editing,

Abdullah Alqarni and Yassir Musa were involved in conceiving the article.

All authors reviewed the results and approved the final version of the manuscript.\

DISCLOSURE

The authors disclosed that there is no competing interest whatsoever.

CONSENT

Written informed consent was obtained from the patients guardian to publish this case report and any accompanying images.

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FIGURES

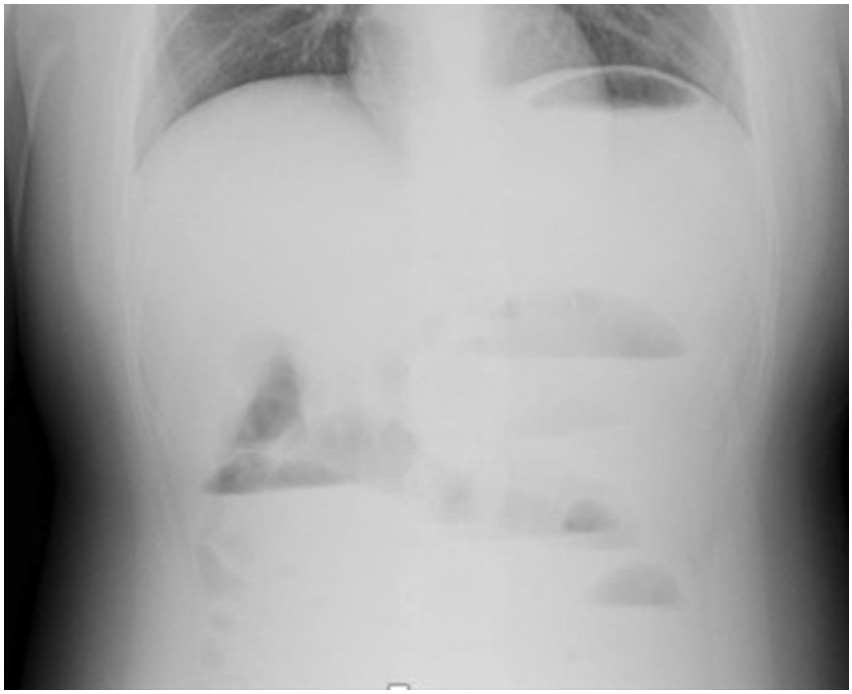

Figure 1: Plain radiograph of the abdomen showing dilated loops of bowels with air-fluid levels in the upper abdomen, predominantly on the left.

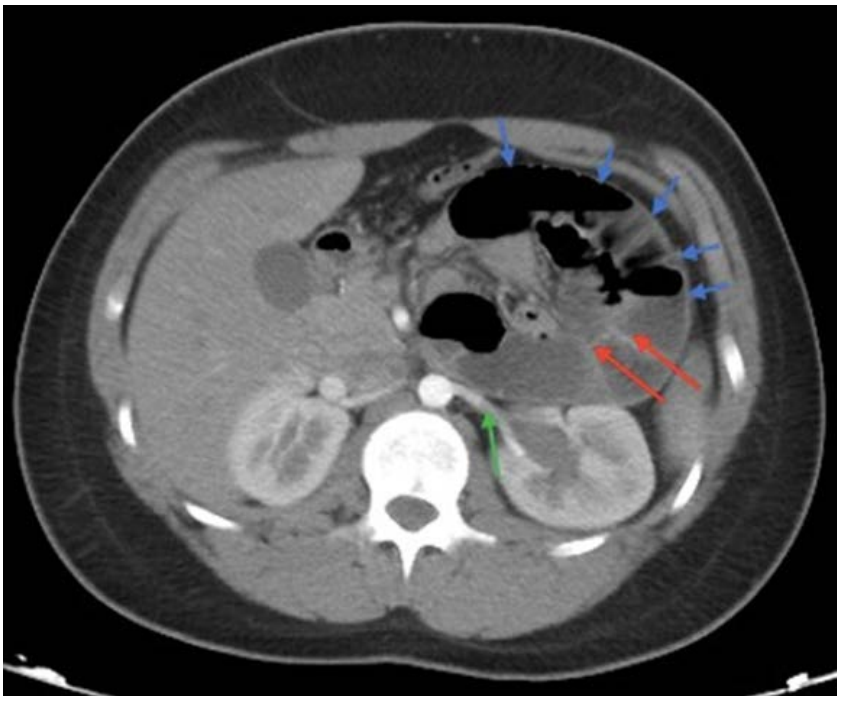

Figure 2: Axial contrast enhanced CT scan of the abdomen showing dilated loops of bowel (red arrows) enclosed within a thin peritoneal membrane (blue arrows) in left abdominal quadrants. Green arrow shows left renal artery posterior to the encapsulated loops of bowel.

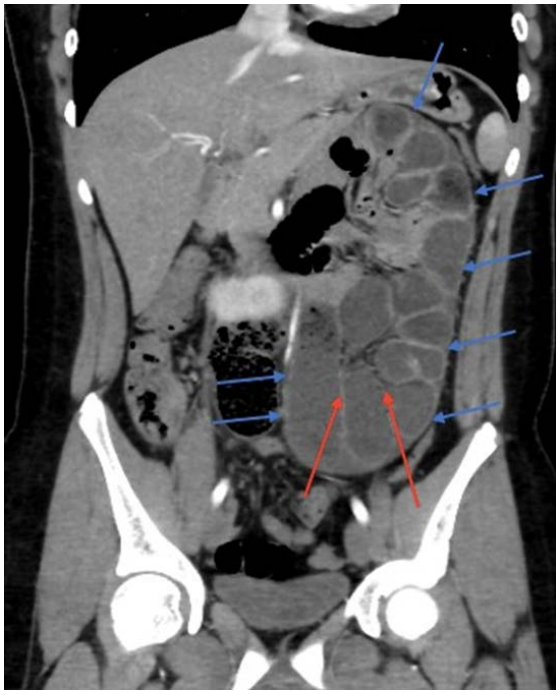

Figure 3: Coronal reformatted contrast enhanced CT of the abdomen showing thin peritoneal membrane (blue arrow) encapsulating clusters of dilated small bowel loops (red arrow) in the left abdominal quadrants.

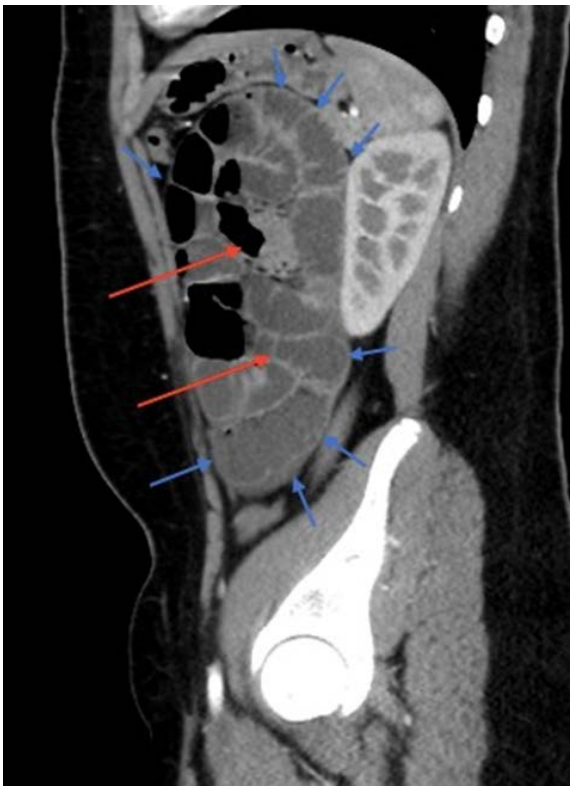

Figure 4: Sagittal reformatted contrast enhanced CT of the abdomen showing thin peritoneal membrane (blue arrow) encapsulating clusters of dilated small bowel loops (red arrow) in the left abdominal quadrants.

KEYWORDS

Congenital Peritoneal Encapsulation, Radiological Imaging Features, Small Bowel Obstruction.

ABBREVIATIONS

CPE = Congenital Peritoneal Encapsulation
AC = Abdominal Cocoon
EPS = Encapsulating Peritoneal Sclerosis
CT = Computed Tomography

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