Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Abdominal cocoon syndrome (ACS): a case report of a Chinese male diagnosed idiopathic ACS with inborn short intestine

Fei Li, MD[a,b], Miao Xie, MD[a,*]

Introduction and importance: Abdominal cocoon syndrome (ACS), as a rare cause of mechanical intestinal obstruction, can be divided into primary/idiopathic vs. secondary type. The primary ACS is often asymptomatic and only diagnosed in exploratory laparotomy. The major treatment of surgery can be challenging. Since the gut wall and peritoneum are densely adhered, gut perforation might occur during adhesiolysis. Thus, it is important to have an experienced surgeon to perform the surgery.

Case presentation: The authors present a primary ACS case of a 50-year-old man. The patient demonstrated an unbearable upper abdominal pain upon admission. A computed tomography (CT) scan showed a severe bowel obstruction. An exploratory laparotomy was indicated, leading to the diagnosis of ACS, which was considered idiopathic after ruling out secondary factors. An adhesiolysis was performed successfully. Note that the entire intestine measured was only 2.1 m during the surgery. There was no post-surgical complication. The patient was recovered uneventfully.

Clinical discussion: The aetiology of primary ACS is unknown. The incidence is comparatively low and considered equal between men and women. As a rare cause of gut obstruction, the suspicion of the diagnosis should be strengthened. Surgery including adhesiolysis and bowel resection remains the major treatment. If adhesiolysis fails, bowel resection will be inevitable. The knowledge and experience of surgeon will be tested.

Conclusion: The aetiology of primary ACS should be further explored. And the differential diagnosis of bowel obstruction should cover ACS in order for the surgeon to be prepared before surgery.

Keywords: abdominal cocoon syndrome, ACS diagnosis, ACS treatment, idiopathic, intestinal obstruction

Introduction

Abdominal cocoon syndrome is first depicted by Owtschinnikow in 1907, and Foo in 1978 termed it as "abdominal cocoon"[1], for a thick collagenous cocoon-like membrane completely or partially encases the gut. As a rare cause of gut obstruction, it is often overlooked in those patients and only diagnosed during exploratory laparotomy. The incidence of abdominal cocoon syndrome (ACS) was initially thought to be more common in adolescent females of tropical and subtropical regions, and the reported data showed that it was also abundant among men, children, and premenopausal women[2]. The prevailing view is that it occurs equally between males and females. There are two types of ACS, primary/idiopathic vs. secondary classifications[3]. The primary ACS is often asymptomatic until the onset of a gut obstruction. And this makes the diagnosis difficult.

Most reported abdominal cocoon syndrome cases often have a history of recurrent abdominal pain to track. The secondary type would have noticeable factors such as past abdominal related infection, surgery or injury. However, the primary/idiopathic type is difficult to predict the course of the disease. We hereby present a primary ACS case in a middle-age Chinese male with a complete mechanical bowel obstruction. An adhesiolysis was successfully performed on the patient, who recovered uneventfully and was discharged after the surgery. It is important to strengthen the suspicion of the rare condition after excluding the common causes to a gut obstruction. Since the major treatment of lysis can easily injure the encased intestinal wall and cause bowel

HIGHLIGHTS

- A 50-year-old Chinese man diagnosed with a primary abdominal cocoon syndrome.
- The patient has an inborn short intestines being about 2.1 m.
- Being asymptomatic makes the diagnosis before surgery difficult in most cases.
- A rare cause of bowel obstruction.
- Performing adhesiolysis can be a real challenge and requires an experienced surgeon.

[a]Departments of Gastrointestinal Surgery and [b]Hepatobiliary and Pancreatic Surgery, PUREN Hospital Affiliated to WUHAN University of Science and Technology, Wuhan, China

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

*Corresponding author. Address: Department of Gastrointestinal Surgery, PUREN Hospital Affiliated to WUHAN University of Science and Technology, 1 Benxi Street, Qingshan District, Wuhan 430080, China. Tel.: +861 767 707 9803; fax: +86 027 86363721. E-mail: XMprhospital@126.com (M. Xie).

Annals of Medicine & Surgery (2024) 86:1152–1155

Received 12 September 2023; Accepted 10 December 2023

Published online 18 December 2023

http://dx.doi.org/10.1097/MS9.0000000000001639

leakage, the surgery can be a real challenge to require a surgeon with knowledge, expertise and experience.

Lesson: Diagnosis of primary ACS before surgery is still difficult. And the surgeon's knowledge of this rare condition and experience of surgery are needed to properly treat those patients.

Our case is reported in line with the SCARE guidelines 2020[4].

Case presentation

A 50-year-old Chinese male without any known history of disease presented to our hospital with progressive colicky abdominal pain for one day accompanied by vomiting. There was no history of fever, jaundice, hematemesis, or melena at the onset of the abdominal pain. The vital signs were stable at admission. Physical examination revealed lower abdominal tenderness with local rebound pain, rigidity and increased bowel sounds. No palpable abdominal swelling was detected. Digital rectal examination found empty rectum. He first showed to our hospital Emergency Department where a contrast-enhanced abdominal computed tomography (CT) scan was prescribed and revealed small bowel dilation loops filled with air-fluid level on images, suggesting a bowel obstruction (Fig. 1). Blood test reports demonstrated an elevated white blood cell, WBC 12.40×10^9/l with 90.00% NEU, and a decreased haematocrit plus low serum sodium, chloride, potassium, and bicarbonate, together indicating an abdominal infection and dehydration.

After the patient was admitted and transferred to our department of gastrointestinal surgery, we further ran the following tests of viral infection, coagulation, liver and kidney function, and EKG, chest X-ray, and started the patient on fluid resuscitation, broad spectrum antibiotics, gastric tube decompression, and ordered nothing by mouth. An acute intestinal obstruction was diagnosed based on the clinical manifestation, the findings of the blood works and the imaging studies. Upon the completion of all the tests, no contraindication of surgery was found.

An emergency exploratory laparotomy was performed. A midline incision was chosen. After the abdominal cavity was entered and exposed, the entire small and large bowels were found closely encapsulated within a dense collagenous cocoon-like peritoneum, leading to the diagnosis of ACS (Fig. 2). Since there was no known history of precondition, the secondary ACS could be ruled out and diagnosis was considered a primary type of ACS.

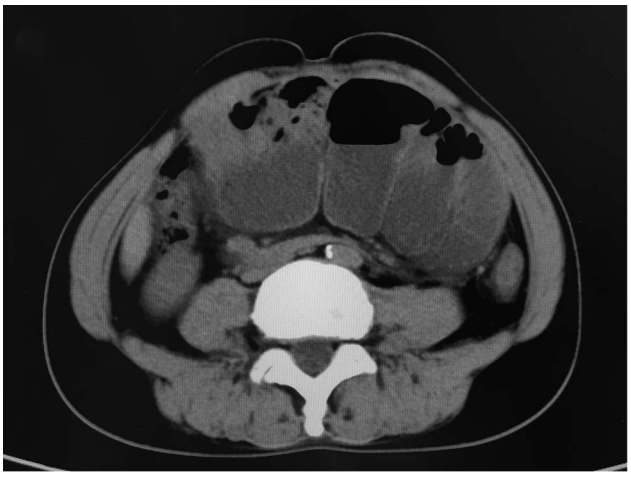

Figure 1. Contrast-enhanced abdominal computed tomography scan. Dilated intestinal loops with multiple air-fluid level.

In order to avoid injury to the intestinal wall, the highest rank of surgeon in our department with abundant experiences performed the surgery. An adhesiolysis was carefully operated to excise the thick sac to loosen the small and large bowel, and the thinned-out and friable gut wall was carefully repaired. The resected peritoneum specimen was sent for pathological examination. Note that after loosening the intestine we estimated the length being only 2.1 m. The patient had an inborn short intestine.

After the surgery, no post-surgical complication was found. The patient was continuously ordered nothing by mouth and treated conservatively with antibiotics, parenteral nutrition, octreotide suppressing bowel secretion, steroid for anti-inflammation and anti-oedema. He recovered uneventfully, and on the sixth postoperative day, another abdominal CT scan showed significant improvement of obstruction and the blood test results returned to normal. After he was put on fluid and then semi-fluid diet, no complaint of discomfort was found. He was discharged two weeks after the surgery on normal diet. The pathological report came back and showed, microscopically, collagenous fibrous connective with hyaline degeneration and fibronoid necrosis, growth of small vessel, inflammatory infiltrate with multinuclear giant cell formation, with no evidence of tuberculosis infection (Fig. 3). A 2-month follow-up was uneventful with the patient being asymtomatic and cured of the gut obstruction.

Clinical discussion

Classification and aetiology

ACS is currently classified into primary vs. secondary types. There are known factors that can lead to secondary ACS, and the most common cause is reported to be continuous ambulatory peritoneal dialysis[3]. Other causes include abdominal TB infection, recurrent peritonitis, previous abdominal surgery, peritoneal shunts, abdominal sarcoidosis, SLE, prolonged beta-blocker treatment, and rarely B-cell lymphoma[5]. None of those factors existed in our case, which was then diagnosed a primary/idiopathic ACS.

The aetiology of primary ACS is still unclear. But some believe congenital condition of a retrograde menstruation and the spread of infection through fallopian tubes play a significant role in females[6]. Yet the epidemiological data showed that the primary ACS was happening no less in males and children. A different theory tried to attribute it to embryological abnormality, where greater omental hypoplasia and mesenteric vessel malformations contributed to encapsulated peritoneal sclerosis[7]. We have reasons to believe that a patient with this congenital condition is likely to demonstrate a recurrent abdominal pain from an early age. However, carefully reviewing the history of our case, we found no history of abdominal pain or any other digestive symptoms. Besides, the patient's entire small intestine was only 2.1 m, which was shorter than the normal 5 m in healthy adults. Interestingly, short intestine syndrome did not develop in the patient. It might be explained that the shortened small intestine had compensated the squeezed abdominal space to fit in it so that the gut obstruction could be delayed in the patient. Thus, it is

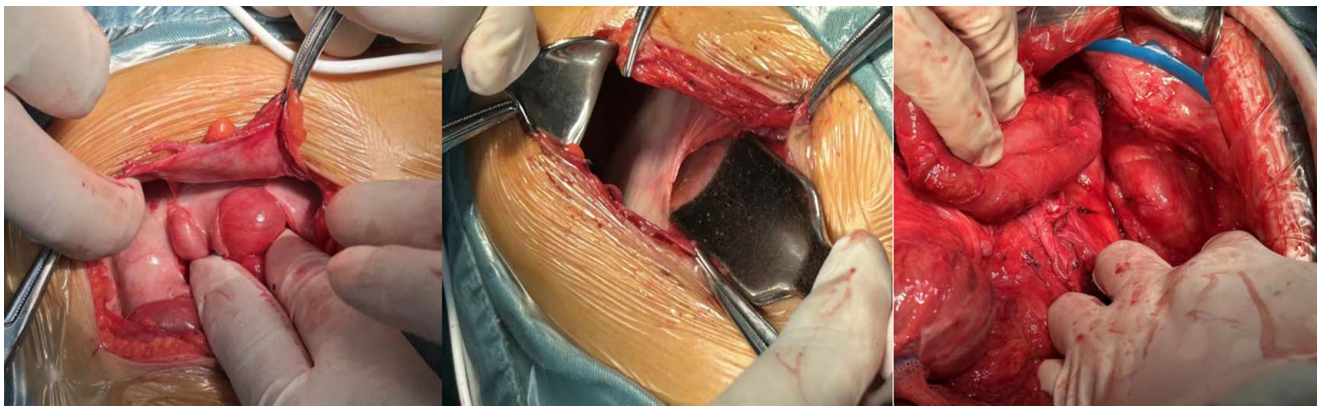

Figure 2. Findings of exploratory laparotomy. A thick membrane is closely encasing the small intestine. the obstructed loops are completely loosed by lysing the membrane.

critical to further investigate the cause of primary ACS in order to deepen our understanding on the progression of this rare disease.

The pathogenesis of cocoon syndrome may associate with epithelial-to-messenchymal cell transition occurring in the peritoneal lining, which then leads to a thick membrane growth. The generally accepted view suggests that any agent irritating mesothelial layer can cause a serositis, or recurrent peritonitis, which then develops a peritoneum fibrogenesis[8]. Microscopically, there might be mesothelial denudation, angiogenesis, interstitial fibrosis, and vascular sclerosis. And the absorption function of peritoneal lining is altered and impaired[9].

Symptoms and diagnosis

Symptoms including colicky abdominal pain, distension, nausea/vomiting and recurrent attacks of subacute intestinal obstruction might manifest in ACS cases. The onset of the symptoms can be acute, subacute, or chronic[10]. Thus, the patients might demonstrate the symptoms from acute small bowel obstruction with abdominal tenderness to chronic abdominal pain associated with nausea, anorexia, and weight loss[11].

Primary or idiopathic ACS as a rare disease can easily be overlooked by doctors. To confirm the diagnosis before surgery is extremely difficult, so it is essential to remain a high index of clinical suspicion in differential diagnosis[12,13]. The early clinical manifestations are unspecific. When the other causes to gut obstruction can be ruled out, this rare condition should be considered in the diagnosis. In our case, we finally established the diagnosis in exploratory laparotomy.

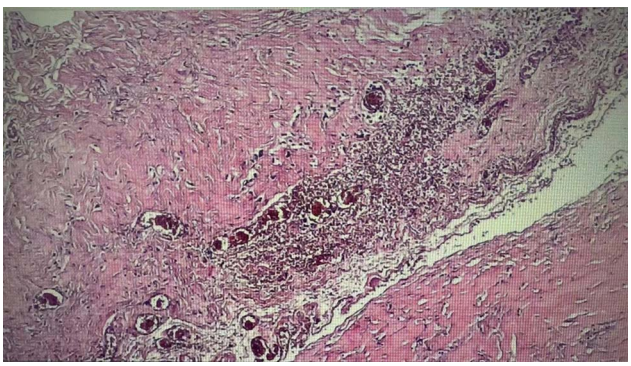

Figure 3. Pathology results. Collageous fibrous connective with hyaline degeneration occurs with fibronoid necrosis, growth of small vessel, and inflammatory infiltrate.

Plain X-rays of the abdomen may reveal a sign of diffused air-fluid levels and clumping of the small intestinal loops, as well as peritoneal or intestinal wall calcifications. Adjunct barium studies to X-rays can present a particular “cauliflower sign” with collected and conglomerated intestinal loops in the centre of the abdomen. Among the available imaging methods, contrast-enhanced computerized tomography is considered to be the gold standard for diagnosis[3]. On CT scan, a thick capsule with a contrast-free periphery may appear around the dilated loops[14].

Laboratory studies are used to monitor the infection, malnutrition, and inflammation of the body, and can help us to assess the condition of the patient both in conservative treatment and post-surgery recovery.

Management

Treatment is mainly devised to deal with gut obstruction, and comprised of conservative vs. surgical approaches. Patients with mild symptoms and partial intestinal obstruction may be treated conservatively on fluid resuscitation, antibiotics and tube decompression, and nutritional support. It is critical to improve the patients’ nutritional status which is the pitfall for an effective conservative treatment and reducing post-surgery complications of infection and fistulae[15]. Surgery is recommended in gut obstruction patients after failure of conservative treatment or with complete bowel blockages. The nature of this condition dictates the surgical approach of being ablation of fibrous tissue, lysis of adhesion, resection of the perforated or ischaemic strangulated bowel plus anastomosis[15,16]. Since the bowel wall is weakly outlined by the capsules and inaccurate enterolysis can easily perforate it, great care should be taken to avoid surgical complications. The chance of recurrence is pretty low if the encasing gut can be completely loosened.

In severe ACS cases, bowel resection can be inevitable if adhesiolysis fails to release the encapsulated gut. The outcome of bowel dissection depends on the length, location, and health of the remaining intestine. Normally, the bowel absorption has a strong adaptive capacity and reservation for resection. To avoid absorption and digestion problem caused by short bowel, the

location of dissection should be taken into consideration. Up to 70% small intestine dissection can be tolerated when ileocecal valve is preserved; in general, the proximal bowel dissection is more adaptable than the distal bowel[17]. In our case, the patient has an inborn short bowel. It raises the question of whether or not our patient can tolerate 70% intestinal dissection without developing short bowel syndrome post-operation. Thus, more data are needed to investigate this issue of ACS in case of having inborn short intestine.

Conclusion

Diagnosis of ACS before surgery is still a challenge, and laparoscopic exploration can provide the ground for it. The aetiology of primary ACS is uncertain and needs further research. In the severe case of bowel obstruction, an adhesiolysis or a bowel resection might ensue following a high index of suspicion of ACS. The surgery is challenging and requires a surgeon with experience and knowledge to avoid surgical complications. The prognosis is promising if the encasing bowel can be completely excised from the capsule and, in this case, the chance of recurrence is quite low.

Ethical approval

Not applicable. We submitted a case report which was a retrospective review and intended for quality improvement, thus it should not be considered research. Besides, we have our patient's signed consent to publish this report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

None.

Author contribution

L.F. is the first author and responsible for the manuscript writing and data collection; X.M. is the corresponding author and conceptualizes this paper.

Conflicts of interest disclosure

There are no conflicts of interest.

Research registration unique identifying number (UIN)

1. Name of the registry: not applicable.
2. Unique identifying number or registration ID: not applicable.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not.
4. applicable.

Guarantor

XIE Miao.

Data availability statement

We state that all the data submitted are available for public access.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Acknowledgements

The authors are thankful to our patient who consented to publish this case.

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] Habeeb MA, Vishwakarma SK, Abdul WM, *et al*. Abdominal cocoon: an enigma. Trop Gastroenterol 2015;36:130–1.
[3] Karona P, Blevrakis E, Kastanaki P, *et al*. Abdominal cocoon syndrome: an extremely rare cause of small bowel obstruction. Cureus J Med Sci 2021;13:e14351.
[4] Agha RA, Franchi T, Sohrabi C, *et al*. The scare 2020 guideline: updating consensus surgical case report (scare) guidelines. Int J Surg 2020;84:226–30.
[5] Machado NO. Sclerosing encapsulating peritonitis: review. Sultan Qaboos Univ Med J 2016;16:e142–51.
[6] Awe JA. Abdominal cocoon syndrome (idiopathic sclerosing encapsulating peritonitis): how easy is its diagnosis preoperatively? A case report. Case Rep Surg 2013;2013:604061.
[7] Alsadery HA, Busbait S, Alblowi A, *et al*. Abdominal cocoon syndrome (idiopathic sclerosing encapsulating peritonitis): an extremely rare cause of small bowel obstruction-two case reports and a review of literature. Front Med 2022;9:1003775.
[8] Moinuddin Z, Summers A, Van Dellen D, *et al*. Encapsulating peritoneal sclerosis-a rare but devastating peritoneal disease. Front Physiol 2014;5:470.
[9] Jarrad B, Ayasa LA, Abboushi MB, *et al*. Intestinal obstruction unraveled: a rare case of primary sclerosing encapsulating peritonitis. Cureus J Med Sci 2023;15:e42289.
[10] Arif SH, Mohammed AA. Abdomen cocoon causing chronic abdominal pain and intestinal obstruction; a case series. Ann Med Surg 2019;48:7–10.
[11] Aziz W, Malik Y, Haseeb S, *et al*. Abdominal cocoon syndrome: a laparoscopic approach. Cureus J Med Sci 2021;13:e16787.
[12] Mousavi MA, Shaghaghi A, Sabouri M, *et al*. Abdominal cocoon syndrome as rare cause of intestinal obstruction: a case report. Int J Surg Case Rep 2022;98:107547.
[13] Ulusoy C, Nikolovski A, Ozturk NN. Difficult to diagnose the cause of intestinal obstruction due to abdominal cocoon syndrome. Eur J Case Rep Intern Med 2021;8:2588.
[14] Solak A, Solak I. Abdominal cocoon syndrome: preoperative diagnostic criteria, good clinical outcome with medical treatment and review of the literature. Turk J Gastroenterol 2012;23:776–9.
[15] Aloraini A, Aljomah N, Almogbel G, *et al*. An incidental intra-operative finding of sclerosing encapsulating peritonitis, "abdominal cocoon" in an emergency presentation of small bowel obstruction, a case report. Int J Surg Case Rep 2023;107:108201.
[16] Chorti A, Panidis S, Konstantinidis D, *et al*. Abdominal cocoon syndrome: rare cause of intestinal obstruction-case report and systematic review of literature. Medicine (Baltimore) 2022;101:e29837.
[17] Townsend CM, Sabiston textbook of surgery: the biological basis of modern surgical practice, 21st edn. ed., Chapter 50. Small Intestine, Imprint: Elsevier, 2022.