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Case Report

Sclerosing encapsulating peritonitis: A rare cause of intestinal obstruction in 2 patients ☆

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ABSTRACT

Sclerosing encapsulating peritonitis is a very rare cause of intestinal obstruction. It usually follows peritoneal dialysis. The idiopathic form is also called abdominal cocoon and is more common in tropical and subtropical regions. We hereby present the clinical histories and imaging findings of 2 confirmed cases of sclerosing encapsulating peritonitis who presented with chronic symptoms of bowel obstruction.

Introduction

Sclerosing encapsulating peritonitis (SEP) is a rare inflammatory condition that causes multiple abdominal symptoms and small bowel obstruction of variable acuity and extent. The 2 major forms are idiopathic SEP, for which no definite source is found, and secondary SEP, with multiple causes [1,2]. SEP results in a thick fibrocartilaginous membrane that primarily encases small bowel loops, much like a larval cocoon. Abdominal cocoon is another name for idiopathic SEP and usually occurs in tropical and subtropical countries in young and middle-aged patients [1,3]. Only 7 cases are reported from Africa despite the rich tropical and subtropical climates of the continent [4].

Surgery is the major modality of treatment if conservative options fail. Although preoperative diagnosis is preferable for proper surgical planning and avoidance of unnecessary bowel

Abbreviations: SEP, sclerosing encapsulating peritonitis; CT, computed tomography; MRI, magnetic resonance imaging; TB, tuberculosis.

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resection, it is a challenging task given the unfamiliarity and rarity of the condition, inadequate workup for acute presentation, and nonspecific symptoms [1,3–6]. As a result, patients are usually misdiagnosed with mimics such as TB [7,8]. Abdominal radiography, barium studies, ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) are all used in an attempt to identify the condition. CT is superior in correctly identifying the typical features of SEP and identifying complications [5].

Case presentation

Case 1

A 20-year-old female presented with abdominal distension, vomiting, and weight loss of 3 months duration. Two years earlier, she received treatment for intraabdominal Tuberculosis (TB) after ascetic fluid analysis confirmed TB following her complaints of abdominal distension, fever, and weight loss. She completed an anti-TB treatment with astounding adherence and has been well for about 9 months in between. In the current presentation, the patient had a stone-like, firm, protuberant abdomen that was not movable even with hard compression. Her abdominal CT scan report from an outside setup indicated peritoneal carcinomatosis. But tumor markers and basic blood profiles were negative, for which she had been sent for an image-guided biopsy to our department. Her abdominal CT (Fig. 1) and sonography (Fig. 2) showed a significantly thickened membrane-like encapsulation of the bowel loops, as well as the bilateral ovaries and adnexa, causing near-complete small bowel obstruction and marked thickening of the walls of the small bowel loops. There were also minimal ascites. A diagnostic laparoscopy was recommended, as these constellations of imaging findings were suggestive of an extreme form of abdominal cocoon. However, initially, the surgical team hesitated to perform either diagnostic and/or therapeutic laparoscopy or open surgery. Thus, the patient underwent an ultrasound-guided biopsy of the markedly thickened membrane-like covering, which showed chronic inflammation and no evidence of malignancy. After a multidisciplinary team discussion involving abdominopelvic radiology, gastroenterology, hepatic medicine, gastric, and colorectal surgery, the team advised surgery, but the patient refused and went against medical advice.

Case 2

The second patient, a 25-year-old female, sought medical attention for persistent abdominal discomfort that increased in intensity in the preceding 3 months. Alongside this, she reported troubling symptoms of malaise, nausea, vomiting, low-grade fever, and anorexia. Prolonged constipation further complicated the clinical picture. Notably, the patient had no prior TB and had not undergone any abdominal surgeries. She also had an unremarkable gynecological history. She did not report any inherited autoimmune or connective tissue disorders. A physical examination revealed abdominal distension with mild tenderness during palpation.

An abdominal CT (Fig. 3) scan revealed a thick, enhancing membrane encasing the bowel loops and giving rise to a cauliflower configuration. The patient underwent surgical exploration, upon which a thickened peritoneum with a leathery appearance was discovered, a gross surgical feature consistent with sclerosing encapsulating peritonitis. Her post-operative period was uneventful.

Discussion

Sclerosing encapsulating peritonitis (SEP) is a chronic inflammatory condition that encases the small bowel with a fibrocartilaginous membrane. The disease is one of the rare causes (6%) of small bowel obstruction and presents in an acute, subacute, or chronic manner [2,4,6,7].

A proper definition of SEP requires recognition of its 2 major forms, namely idiopathic, or primary, and secondary. Abdominal cocoon is synonymous with idiopathic SEP [1]. This form is a diagnosis of exclusion after missing any plausible etiologies for SEP on clinical assessment, laboratory, radiological, and surgical work-ups. Secondary SEP, on the other hand, has multiple systemic and local causes. The former includes medications such as long-term use of beta-adrenergic blockers, methotrexate, and toxins like asbestosis. Many local causes are implicated, including peritoneal dialysis, which is the leading cause worldwide. TB, abdominal surgery, abdominal trauma, abdominal malignancies, liver transplant, ventriculoperitoneal and peritoneovenous shunts, liver transplantation, recurrent peritonitis, sarcoidosis, and systemic lupus erythematosus are also well-recognized [1,3,8–10]. Accordingly, the SEP in our first case is likely attributable to her previous abdominal TB. Our second case on the other hand had no identifiable etiologies, thus making his SEP the idiopathic form.

Foo in 1978 [11] was the first to use the term abdominal cocoon after treating it in 10 young female patients with small bowel obstruction. All of them did not have any definite predisposing conditions. Intraoperatively, variable degrees of small bowel encasement by a fibrous membrane separate from the peritoneal membrane, inside which involved bowel segments were coiled in concertina fashion, were found. He hypothesized retrograde menstruation-induced inflammation with superimposed viral infections as the underlying pathogenesis. From the descriptions, his cohort had the primary form of sclerosing encapsulating peritonitis. Obviously, retrograde menstruation is inadequate for explaining the disease in males, children, and perimenopausal women.

Akbulut et al [1] reviewed 193 cases of idiopathic SEP and found men were affected twice as commonly as females. The average age was 34.7 ± 19.2 years. The majority of the reported cases were from tropical or sub-tropical countries, with China, India, and Turkey reporting the largest number of cases. Cases have also been reported from Africa, including Sudan, Nigeria, and Senegal. A study by Li et al [3] from China was included in the same study and reported 65 cases, the largest case series among the reviewed reports, and corroborated the young and middle age and tropical and subtropical preponderance of idiopathic SEP cases, but differed in that the majority of

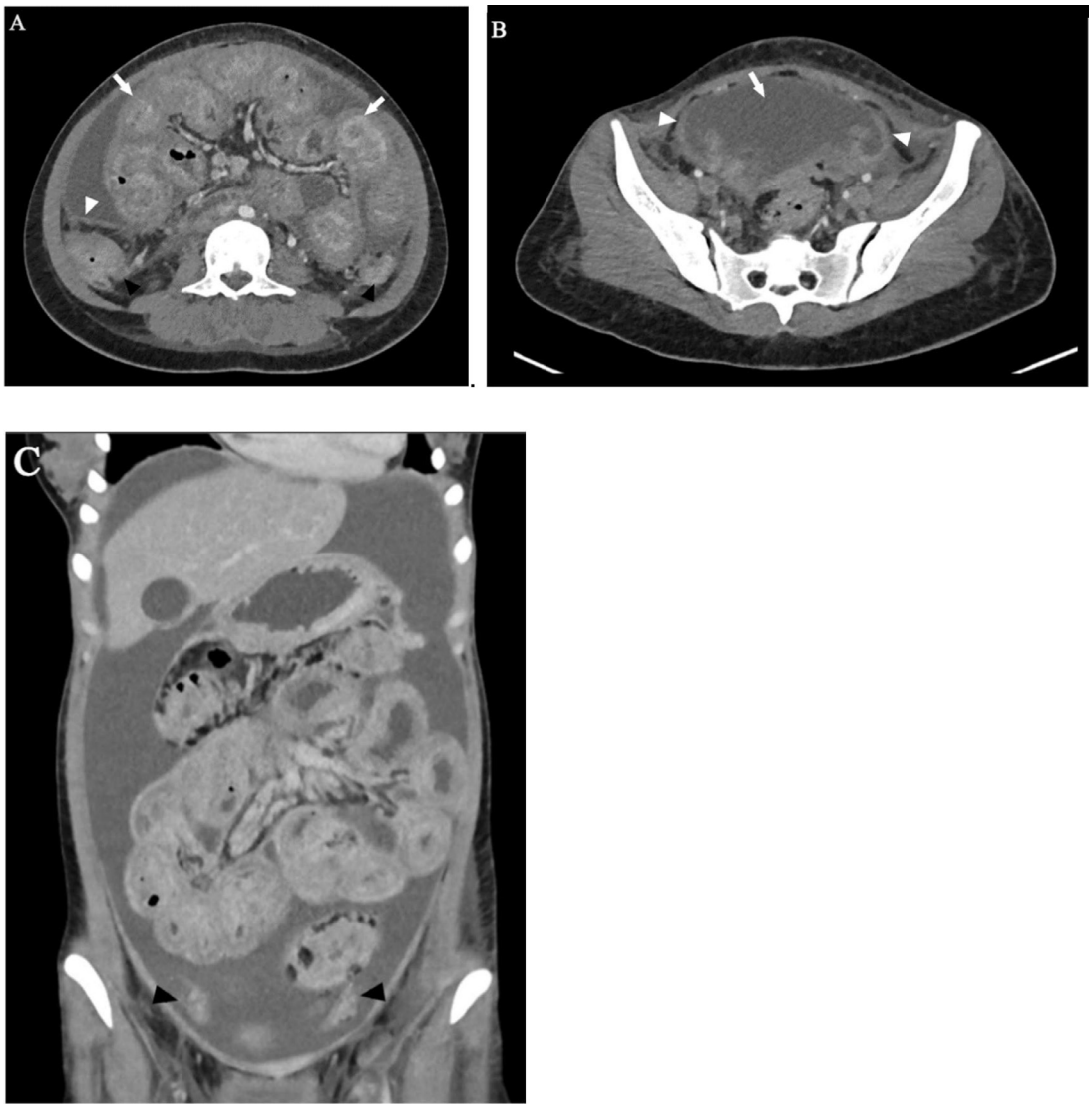

Fig. 1 – Axial (A, B) and coronal post-contrast abdominopelvic CT images: Small bowel loops and loculated fluid are positioned centrally (white arrow in A and B) are encased by a thick enhancing extra-peritoneal membrane that extends down to the pelvis (white arrowheads in A and B). The ascending and descending colon are not involved (black arrowheads in A). The ovaries are within the encasement (black arrowheads in C).

their patients were females. Also, in the study by Wei et al [12], with 24 cases (the second largest case series), 15/24 cases were females. Therefore, sex is not a strong criterion for favoring SEP.

The etiology of idiopathic SEP is not known, but congenital and vascular etiologies are suggested. Wei et al [12] discovered 41.7% of their cases had omental dysplasia and hypothesized intrauterine descent of the membranous greater omentum along the transverse colon and encasement of the intestines as the mechanism. Mesenteric vascular malformations are also seen in some cases of idiopathic SEP [2].

Secondary SEP is more common than the primary form and has multiple causes, as mentioned above. Secondary SEP decreases the ultrafiltration capacity of the peritoneum and increases the risk of small bowel obstruction. Long-standing peritoneal dialysis carries a 0.5%-2.8% risk of SEP and is the leading cause worldwide [1,2,8]. In endemic parts of the world, TB commonly causes SEP and occurs together with other abdominal stigmata such as mesenteric abscesses, caseating lymphadenopathies, and serosal tubercles. In regions where TB is endemic and peritoneal dialysis is not a common practice, it is plausible to consider it the leading cause. In fact, erroneous empiric anti-TB therapy is offered for SEP symptoms in these regions [4,13,14].

Histological examination of the encasing membrane reveals proliferation of fibro connective tissue, mononuclear inflammatory infiltrates, and dilated lymphatic channels. In idiopathic forms, giant cells or foreign body granulomas are absent [1,5].

The majority of SEP patients are symptomatic, with an average length of illness lasting 2-4 years. Acute, subacute, and chronic small bowel obstruction (partial or complete), abdominal pain, abdominal distension, constipation, fever, nausea and vomiting, weight loss, and malnutrition are the common

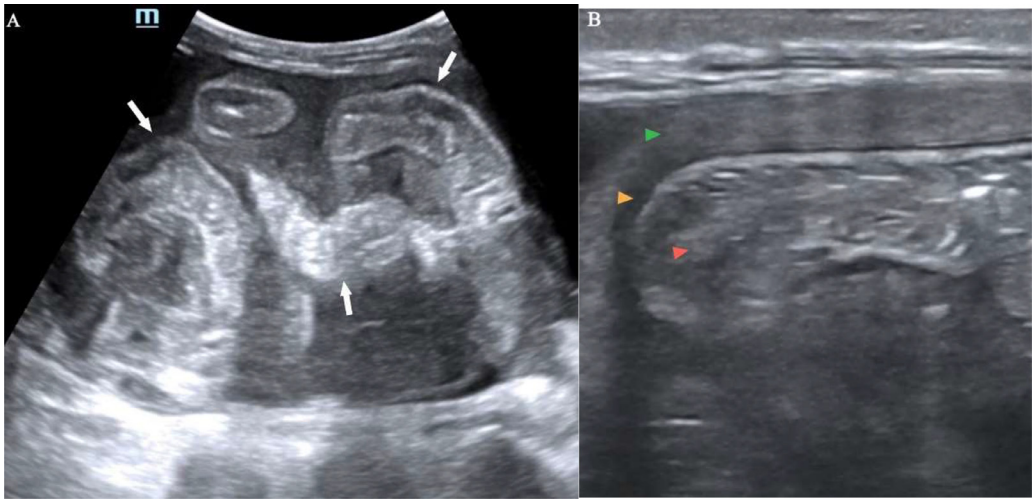

Fig. 2 – B-mode low (A) and high (B) frequency abdominal ultrasound scans: A shows centrally clumped up bowel loops (white arrows in A) and ascites fluid. A markedly thickened membrane (green arrowhead), bowel wall (yellow arrowhead), and bowel interface (red arrowhead) giving a trilaminar appearance are shown in B. Note the barely resolved outline of the membrane, the peritoneum, and the bowel wall, which tend to merge with each other.

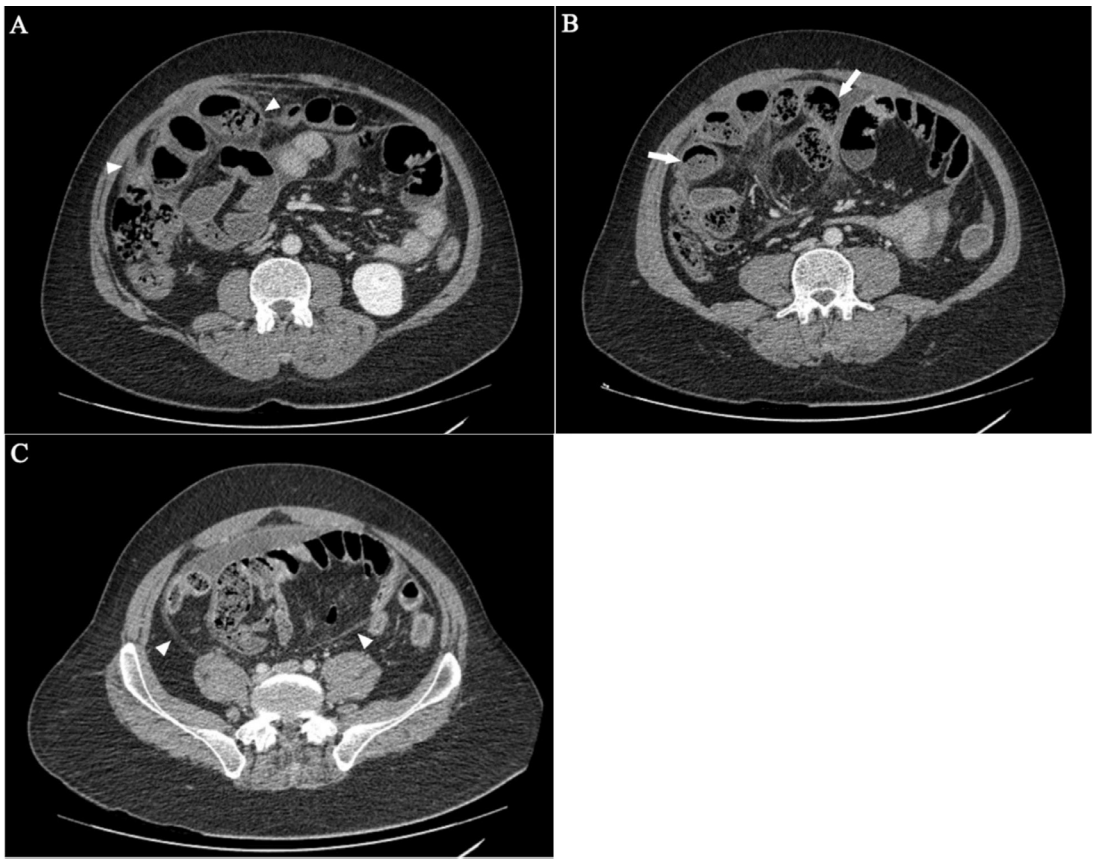

Fig. 3 – Axial post-contrast abdominopelvic CT images: White arrowheads (A and C) show smooth enhancing membrane encasing loops of small bowels which are assuming a cauliflower shape (white arrows in B). Loculated interloop ascites is seen in within the encasement in C.

manifestations [3,12,13]. Perforation is extremely rare but has occurred in both primary and TB-associated SEP [1,15].

Preoperative diagnosis of idiopathic SEP is vital but extremely difficult to achieve and requires a high index of suspicion as the condition is rare, has nonspecific clinical symptoms, and patients who present with acute obstruction are rushed to surgery. Lack of advanced imaging modalities in many emergency departments further contributes to the challenge [1,13]. The importance of settling the diagnosis lies in avoiding an unnecessary bowel resection, which can lead to short small bowel syndrome. Among radiological pursuits, CT is preferred for accurate diagnosis. Nonetheless, many patients are still only diagnosed intraoperatively: 4/24 in Wei et al [12] and 34/65 in Li et al [3]. The presence of one of the predisposing conditions is a clue to secondary SEP.

Based on the extent of the encapsulating mantle, SEP is classified into 3 groups. Type 1 is partial small bowel encasement; Type 2 is complete small bowel encasement; and Type 3 is extra intestinal involvement of other structures such as the colon and ovaries [2,12]. In Wei et al [12] the least common type was type 3, occurring in 4/24 cases.

Radiology plays an important role in diagnosis. Plain abdominal X-rays are nonspecific and go as far as showing features of small bowel obstruction only. If a small bowel obstruction is long-standing, these might not even be seen on X-rays [5]. Small intestinal barium studies are also employed in patients without severe obstruction. They show fixed and conglomerated small bowel loops. Maguire et al [9] reported their experience with 5 cases of secondary SEP following orthotopic liver transplant and found delayed transit time through the nondilated small intestine in 4 patients (one of them also had upper gastrointestinal dilation) in a barium study. A previously described contrast sign, the cauliflower sign [16], was not seen in any of their patients. But the authors admit barium studies are challenging to interpret in the presence of massive ascites after liver transplantation. In another case series, though, the barium contrast study was largely noncontributory [13].

Ultrasound and CT are more accurate imaging tools. On sonography, the typical finding is narrow-based small bowel loops (which might show to-and-fro peristalsis) clustered within a thick membrane in a concertina fashion. An additional typical feature is the ultrasound trilaminar sign, consisting of the superficial hyperechoic peritoneal membrane, a middle hypoechoic layer of bowel wall, and the deep hyperechoic layer produced by bowel gas/content. Ascites aid in visualization of the clustered bowel loops (appearing like a cauliflower) and the trilaminar sign, with high resolution afforded using the linear-array probe. An attempt at dispersing the clustered loops by direct probe pressure is unsuccessful as they are fixed [17–19]. The abdominal sonography (Fig. 2) of our first case are consistent with these features.

Contrast-enhanced CT also shows similar findings but with greater clarity. Overall, CT is an excellent modality for small bowel obstruction, with high sensitivity and specificity for diagnosis, detecting causes and complications, and predicting surgical intervention [20]. CT shows the abnormal position and location of the affected segment of small bowel loops, which are central, conglomerated, and encased by a soft tissue density layer or sac-like structure. CT is the most sensitive modality to outline the complete extent of the encasing membrane. Thick (>2 mm) and uniformly enhancing smooth peritoneum is also seen. The affected bowel loops may be dilated with mural thickening. Angulation, kinking, and tethering are important to report as they signify interloop adhesion and confer a greater surgical risk. Proximal dilation, nature, and volume of ascites and loculated collections can also be seen. Calcifications of varied morphologies can also occur. In cases of SEP secondary to TB, omental thickening, nodules, as well as matted lymphadenopathy, are seen. Recognition of SEP is important as interloop fibrosis and adhesion are usually severe, entailing adhesiolysis that carries a higher risk of iatrogenic injury. Ensuing complications such as bowel ischemia (nonenhancing) and bowel perforation (free peritoneal gas) are rare but can be seen. CT can be used to identify complications of SEP such as bowel gangrene (lack of normal bowel enhancement) and bowel perforation (pneumoperitoneum, oral contrast material extravasation). These complications are relatively rare because the thickened peritoneal membrane encapsulates and compresses the bowel from the exterior, with preservation of the central vascular pedicle (as opposed to mimics such as internal hernia, in which the pedicle is twisted, leading to early ischemia). CT can reliably depict these complications when they occur and can help determine management. Owing to its excellent depiction of the anatomic abnormality and the complications, contrast-enhanced CT is regarded as the modality of choice in the imaging of SEP. CT is the modality of choice in the imaging work-up of SEP [5,13,21–23]. MRI shows similar findings but does better at identifying abnormal peritoneal enhancement and the separation of the bowel loops from the capsule by cine imaging [1,5].

Peritoneal encapsulation and internal hernia are 2 important differential diagnoses that have a similar morphology to SEP. The former, unlike SEP, is a noninflammatory congenital accessory membrane between the omentum and mesocolon. It is usually an incidental discovery during laparotomy. Internal hernia is a reasonable differential, as clustered bowel loops are also present. But the absence of an encasing membrane, fixed anatomic location, and secondary effects on mesenteric vessels (twisting, crowding, stretching) help in the differentiation of internal hernia and SEP. But it is worth noting that vascular complications are more common with internal hernia than SEP, as the covering mantle affords protection of the vascular pedicle in the latter. The same mechanism is also protective against perforation [1,5,17].

There are no agreed-upon treatment regimens for idiopathic SEP, as the condition is rare [13]. The patient's symptoms command treatment options. With an early diagnosis of SEP possible, conservative measures such as watchful waiting, bowel rest, and nasogastric decompression for mild symptoms can be followed. If conservative therapies do not help, various anti-inflammatory and antifibrotic drugs are used alone or in combination, and options include corticosteroids, Tamoxifen, and Colchicine (both have an anti-transforming growth factor beta effect [1,13].

For severe symptoms, surgery is required, the goals of which are removing the encasing membrane, releasing adhesions, and relieving obstruction. The most appropriate procedure for idiopathic SEP is membrane excision and adhesiolysis. Total membrane excision diminishes the chances of recurrence. Bowel resection and anastomosis is associated with

short small bowel, enterocutaneous fistula, and sepsis; hence, it should be carried out for gangrenous bowel only, which is rare in SEP. Unnecessary resection and anastomosis increases morbidity and mortality [1,3,12,24]. Preoperative nutritional support is a significant factor for better treatment outcome [3]. The main complications are postsurgical inflammatory intestinal obstruction (within the first 30 days) and chronically, adhesive intestinal obstruction [3,12].

In conclusion, idiopathic SEP is a rare condition that can easily be confused with other, more common causes of intestinal obstruction. It should be considered in patients with prolonged symptoms of bowel obstruction, in the absence of other causes. Imaging suggests the diagnosis preoperatively, enabling the use of conservative surgical procedures to reduce post-operative complications.

Patient consent

Written informed consent was obtained from the two patients for anonymized information to be published in this article.

REFERENCES

[1] Akbulut S. Accurate definition and management of idiopathic sclerosing encapsulating peritonitis. World J Gastroenterol 2015;21(2):675–87.
[2] Tannoury JN, Abboud BN. Idiopathic sclerosing encapsulating peritonitis: abdominal cocoon. World J Gastroenterol 2012;18(17):1999–2004.
[3] Li N, Zhu W, Li Y, Gong J, Gu L, Li M, et al. Surgical treatment and perioperative management of idiopathic abdominal cocoon: single-center review of 65 cases. World J Surg 2014;38(7):1860–7.
[4] Mohammed F, Abdulkarim M, Ibn Yasir A, Taleballah O, Shani D, Salih N. Abdominal cocoon syndrome, a case report of a rare disease entity causing intestinal obstruction. Int J Surg Case Rep 2021;87:106401.
[5] Singhal M, Krishna S, Lal A, Narayanasamy S, Bal A, Yadav TD, et al. Encapsulating peritoneal sclerosis: the abdominal cocoon. Radiographics 2019;39(1):62–77.
[6] Gurleyik G, Emir S, Saglam A. The abdominal cocoon: a rare cause of intestinal obstruction. Acta Chir Belg 2010;110(3):396–8.
[7] Bell CM, Dart BW. Abdominal cocoon-an unusual case of bowel obstruction. Am Surg 2016;82(11):308–9.
[8] Al Saied G, Hassan AZ, Ossip M, Hassan AZ. Idiopathic sclerosing encapsulating peritonitis. Case report and review of literature. Eur Surg 2010;42(2):103–6.
[9] Maguire D, Srinivasan P, O'Grady J, Rela M, Heaton ND. Sclerosing encapsulating peritonitis after orthotopic liver transplantation. Am J Surg 2001;182(2):151–4.
[10] Brown MC, Simpson K, Kerssens JJ, Mactier RAScottish Renal Registry.. Encapsulating peritoneal sclerosis in the new millennium: a national cohort study. Clin J Am Soc Nephrol 2009;4(7):1222–9.
[11] Foo KT, Ng KC, Rauff A, Foong WC, Sinniah R. Unusual small intestinal obstruction in adolescent girls: the abdominal cocoon. Br J Surg 1978;65(6):427–30.
[12] Wei B, Wei HB, Guo WP, Zheng ZH, Huang Y, Hu BG, et al. Diagnosis and treatment of abdominal cocoon: a report of 24 cases. Am J Surg 2009;198(3):348–53.
[13] Singh H, Irrinki S, Yadav TD, Kumar H, Kharel J, Dhaka N, et al. Surgical outcomes in patients with abdominal cocoon: series of 15 patients. World J Surg 2019;43(9):2143–8.
[14] Kaushik R, Punia R, Mohan H, Attri AK. Tuberculous abdominal cocoon – a report of 6 cases and review of the literature. World J Emerg Surg 2006;1:18.
[15] Hu Q, Shi J, Sun Y. Abdominal cocoon with intestinal perforation: a case report. Front Surg [Internet] 2021;8. [cited 2023 November 26]. (accessed February 14, 2024). Available from: https://www.frontiersin.org/articles/10.3389/fsurg.2021.747151/full.
[16] Sieck JO, Cowgill R, Larkworthy W. Peritoneal encapsulation and abdominal cocoon. Case reports and a review of the literature. Gastroenterology Jun 1983;84(6):1597–601.
[17] Vijayaraghavan SB, Palanivelu C, Sendhilkumar K, Parthasarathi R. Abdominal cocoon: sonographic features. J Ultrasound Med 2003;22(7):719–21.
[18] Rokade ML, Ruparel M, Agrawal JB. Abdominal cocoon. J Clin Ultrasound 2007;35(4):204–6.
[19] Indiran V, Ethiraj D. Ultrasound trilaminar sign of abdominal cocoon. GE Port J Gastroenterol 2021;29(6):442–3.
[20] Li Z, Zhang L, Liu X, Yuan F, Song B. Diagnostic utility of CT for small bowel obstruction: systematic review and meta-analysis. PLoS One 2019;14(12):e0226740.
[21] Hur J, Kim KW, Park MS, Yu JS. Abdominal cocoon: preoperative diagnostic clues from radiologic imaging with pathologic correlation. Am J Roentgenol 2004;182(3):639–41.
[22] Gupta S, Shirahatti RG, Anand J. CT Findings of an abdominal cocoon. Am J Roentgenol 2004;183(6):1658–60.
[23] Ethiraj D, Indiran V. Abdominal cocoon: "cauliflower sign" on contrast-enhanced computed tomography scan. GE Port J Gastroenterol 2020;28(1):76–7.
[24] Çolak Ş, Bektaş H. Abdominal cocoon syndrome: a rare cause of acute abdomen syndrome. Ulus Travma Acil Cerrahi Derg 2019;25(6):575–9.