ORIGINAL ARTICLE

Abdominal cocoon syndrome: A rare cause of acute abdomen syndrome

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ABSTRACT

BACKGROUND: A rare cause of acute abdomen or intestinal obstruction, the abdominal cocoon syndrome is also described in the literature as sclerosing peritonitis or sclerosing encapsulating peritonitis. Abdominal cocoon is characterized by the total or partial wrapping of the abdominal organs by a fibrous membrane. Although it is usually observed in young women, the etiology is unknown. The diagnosis is usually made during laparotomy. In this case series, we aimed to present seven patients diagnosed with abdominal cocoon syndrome during operation.

METHODS: The records of patients who underwent laparotomy for abdominal pain and/or intestinal obstruction in our hospital and diagnosed as abdominal cocoon during operation between January 2012 and November 2018 were retrospectively reviewed. The demographic characteristics of the patients, etiologic factors, surgical procedures, operative findings and follow-up of the patients were recorded.

RESULTS: Four out of seven patients who were operated for abdominal cocoon were male and 3 of them were female. The median age of patients was 61 (57–63) years in male and 39.6 (28–49) years in female. Six of the patients were operated in emergency conditions with the diagnosis of an acute abdomen or ileus. One of the patients was operated with the diagnosis of an intra-abdominal mass in elective conditions. In five out of seven patients, all of the small intestines were wrapped with a fibrous collagen capsule, while two of the patient intestines were partially wrapped with a fibrous collagen capsule. Four of the patients had no underlying disease, while one of the patients had Familial Mediterranean Fever (FMF), one had Endometriosis and one had beta-blocker medication. One patient who had small bowel necrosis and septic peritonitis were observed during the operation and died post operative 6th days. Post-operative complications were not observed in the follow-up of other patients and reoperation was not required due to recurrence.

CONCLUSION: Abdominal cocoon is a condition that is usually diagnosed during operation in patients that were operated for reasons, such as the acute abdomen or intestinal obstruction. When the diagnose delayed, death can be seen due to small bowel necrosis and septic complications. High clinical suspicion and radiological imaging are important in the preoperative diagnosis. Treatment is required adhesiolysis and excision of the fibrous membranes.

Keywords: Abdominal cocoon; endometriosis; ileus; peritoneal fibrosis.

INTRODUCTION

Sclerosing encapsulated peritonitis, a rare cause of acute abdomen or intestinal obstruction, was first described in 1868 and the definition of peritonitis chronica fibrosa incapsulate was used.[1] After 90 years of this identification, Foo et al.[2] used the term abdominal cocoon syndrome (ACS) for this disease, in 1978. The definition of primary sclerosing peritonitis or idiopathic sclerosing peritonitis is also used synonymously for this disease.[3]

ACS can be seen in primary form without any underlying cause and may be related to interventions, such as abdominal surgery, chronic peritoneal dialysis, liver transplantation, ventriculoperitoneal shunt, and some drugs.[4,5]

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Radiologically, contrast-enhanced Magnetic Resonans Imaging (MRI) can diagnose of abdominal cocoon better than computerised tomography (CT).[6]

Factors affecting the clinical signs of ACS are duration and severity of the disease, underlying causes and immunological condition of patient. ACS is most commonly characterised-by recurrent intestinal obstruction attacks.[7] However, in some patients, the entero-atmospheric fistula may be associated with more rare complications, such as necrosis of small intestine due to impaired blood flow and malnutrition due to long-term nausea and vomiting of patients.[8]

In most cases of ACS, is diagnosed intraoperatively. The diagnosis of ACS is very difficult with clinical evaluation.[9] Abdominal computerised tomography (CT) has a significant role in the pre-operative diagnosis of ACS.[10] Surgical intervention remains the most effective treatment option for ACS. In the surgery, excision of the fibrous capsule and adhezyolizis are performed.[11] Surgical intervention has many complications, such as iatrogenic bowel injury and recurrent ileus due to adhesions in the early period. In this study, we aimed to investigate the patients who were operated for acute abdomen or ileus between January 2012 and November 2018 and who had ACS detected during exploration.

MATERIALS AND METHODS

The files of the patients who were diagnosed with abdominal cocoon during the operation in our hospital were retrospectively reviewed between January 2012 and November 2018. The demographic characteristics of the patients, etiologic factors, surgical procedures and operative findings of the patients were recorded.

The seven patients operated for abdominal cocoon, four of them were male and three of them were female. The median age of the patients was 61 (57–63) in male and 39.6 (28–49) in female. Six of the patients were admitted to the emergency department with acute abdomen and ileus. Five patients underwent CT and one patient underwent direct abdominal radiography. CT findings of patients were shown in Figure 1 and 2. The patients had a mild leucocytosis ranging from 9.900 to 12300. One of the patients underwent elective surgery with the diagnosis of intraabdominal mass localised the mesenter of intestine. Demographic characteristics, pathology and follow-up of patients were shown in Table 1.

Informed consent was obtained from all the patients included in this study. This study was performed, and data were collected according to the ethical principles of the Declaration of Helsinki.

Statistical Analysis

Descriptive statistical methods, such as mean±standard de-

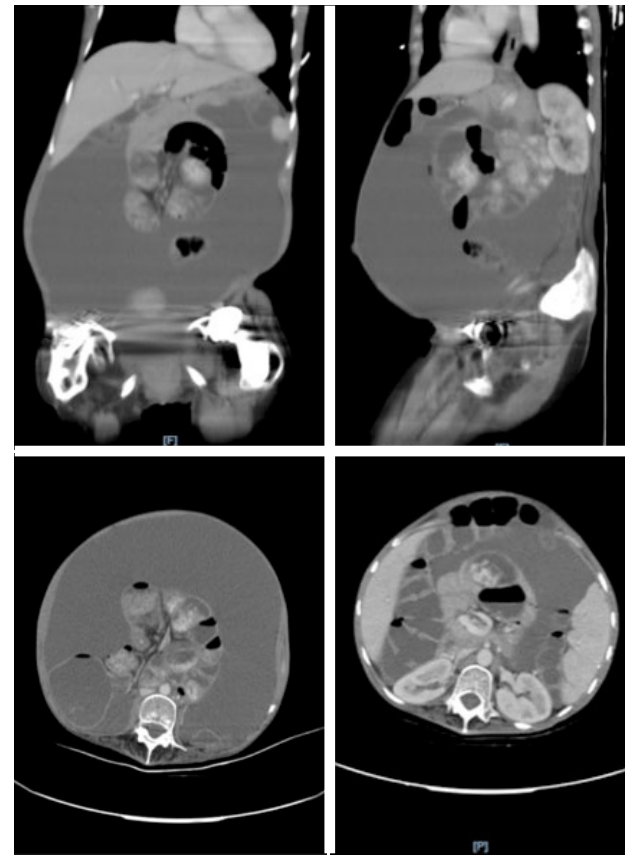

Figure 1. CT images of totally wrapped abdominal cocoon.

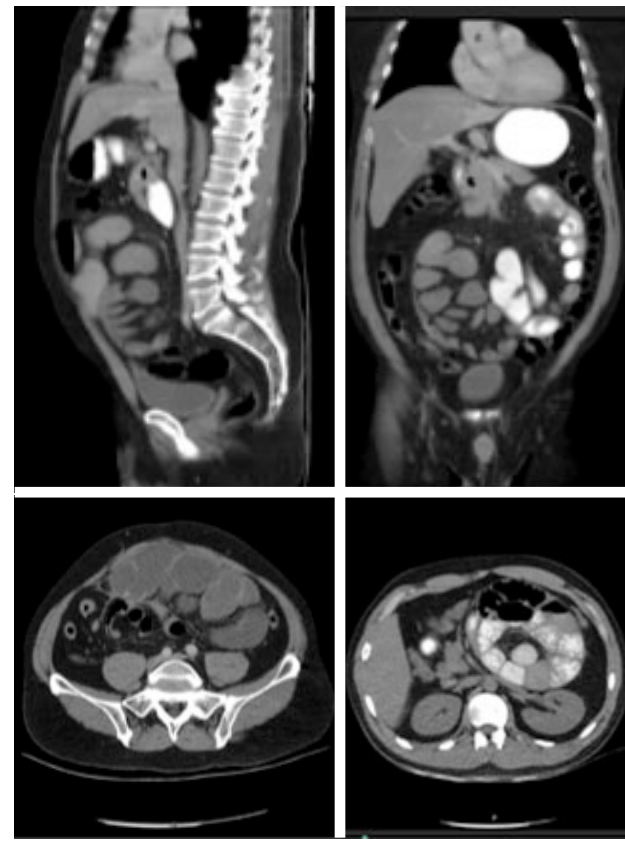

Figure 2. CT images of partially wrapped abdominal cocoon.

Table 1. Demographic characteristics and histopathology of patients

Sex	Age	Total capsule of intestine	Partial capsule of the intestine	Pathology	Etiology	Follow-up/months
M	63	(+)		Fibrous inflammatory material	Propranolol	75
M	57	(+)		Fibrous inflammation material	–	54
F	28		(+)	Endometriosis	Endometriosis	45
F	49	(+)		Inflammatory peritoneum	–	36
M	61		(+)	Fibrous inflammatory material	–	36
F	42	(+)		Fibrous inflammatory material	FMF	3
E	63	(+)		Inflammatory peritoneum	–	Exitus

viation and/or median (minimum–maximum) and frequency and percentage, were used for data evaluation. The statistical analysis was performed using the Statistical Package for Social Sciences (SPSS®) software package for Windows, version 17.0 (SPSS Inc., Chicago, Illinois, USA).

RESULTS

In two of the patients, all small intestines were partially wrapped with peritoneum, and there were adhesions between the small intestine. The other five patients were covered with fibrocollagenous tissue that formed on the intestines that adhered together due to inflammation. In three of these patients, fibrocollagen tissue covered all intestines, while in two patients a partial of the small intestine was covered with fibrocollagen. All patients underwent adhesiolysis and capsule excision. In one patient, the iatrogenic injury occurred in the transverse colon during adhesiolysis and repaired primarily. One patient had underwent partial omentectomy due to necrosis of omentum, and partial small bowel resection due to small bowel necrosis. This patient developed small bowel necrosis and abdominal sepsis and died 4 days after the operation. The median duration of hospitalization was 9.5 (4–14) days.

The pathologic examination of the excised capsules revealed in two patients with peritoneal inflammation, in four with fibrozis containing fat and connective tissue and one patient with endometriosis. When the etiologic factors and past medical history of patients were evaluated four of patiets had no etiological cause, one patient had medication of beta-blocker, one patient had endometriosis and one patient had FMF. The median hospitalisation in our study was 9.5 (4–14) days. The follow-up period of our patients was 35.57 (3–75) months and two patients were hospitalized for follow-up because of subileus attacks, but re-operation was not required.

DISCUSSION

The clinical manifestations of ACS is usually consisted of acute or subacute intestinal obstruction and sometimes acute abdomen. On physical examination, there is an increase of bowel sounds with beside of abdominal distention. The small intestines covered with a fibrocollagen membrane localised anywhere in the abdomen.[12] In this case, if there is no acid in the abdomen, non-mobile intestines may cause asymmetric abdominal distension in the physical examination. There are non-specific findings, such as nausea and vomiting, which are frequently recurrent spontaneously or respond to medical treatment when questioned. Chronic constipation, loss of appetite, weight loss and intraabdominal masses, perforation, or ischemia-related necrosis is rarely seen in these patients. Yip and Lee reported four main clinical features to standardize the preoperative diagnosis: a young female patient with intestinal obstruction with no apparent cause, having a background with similar attacks, having a bowel obstruction and bloating, and having a non-sensitive mass in the examination.[11,13] In many studies with a high number of cases, the rate of diagnosis during operation varies between 52.3% to 100%. On the other hand, the rate of patients diagnosed preoperatively varies between 16.7% and 48%.[9,14]

In our series, the number of males was higher than female patients, whereas female patients were younger than male patients. All of the patients had similar and recurrent acute episodes, and one patient had a diagnosis of intraabdominal mass. However, we could not detect asymmetric distention in the abdomen.

The most common form of ACS is idiopathic form and its exact incidence is unknown. The secondary form occurs in various situations. These include familial Mediterranean fever (FMF), endometriosis, Peritoneal Dialysis (PD), ventriculoperitoneal or peritoneo-venous shunts, liver transplantation, recurrent peritonitis, medication of proctolol and propranolol, beta-blockers, methotrexate, protein C deficiency, exposure of asbestos, intraperitoneal chemotherapy, intraperitoneal povidone - iodine contact, liver cirrhosis, gastrointestinal malignancy, fibrogenic foreign object, systemic lupus erythematosus, abdominal tuberculosis and granulomatous peritonitis caused by parasitic infectious.[3,9,15] The incidence of ACS in patients with peritoneal dialysis has been reported to be between 1.4% and 7.3%.[6]

The case series published in recent years, ACS is seen more frequent in tropical and subtropical regions, such as in China, India and Turkey, and idiopathic ACS is reported that more common in women than men.[9,16] Preoperative diagnosis is quite difficult. Because the clinical findings are nonspecific, the most useful method for diagnosis is multislice CT. In addition, abdominal X-ray can be used and abdominal ultrasonography, but they provide very limited information for diagnosis. In x-ray examination and ultrasonography, dilated small bowel loops, air-fluid levels, intraperitoneal echogenic bands and calcifications can be seen.[4,8,11,17] Accordion pattern or cauliflower sign can be seen in abdominal CT. Delay in the passage of the contrast medium through the small intestine, collection of the dilated gut in one region, membrane formation around the intestines, wall thickening, localized fluid collection, peritoneal thickening and contrast enhancement are other features that can be seen in CT. CT is useful in preoperative diagnosis.[8,18]

Evidence in the literature indicates that patients with minimal abdominal symptoms should be treated conservatively. Intestinal resting, nasogastric decompression, and parenteral nutrition support are recommended for these patients.[3,9,19] Improving the nutritional status of these patients is of great importance as it can improve the response to conservative treatment or prevent subsequent surgical complications, such as infection and fistula. For patients not responding to conservative treatment, drugs, such as tamoxifen, steroids, colchicine, azathioprine and mycophenolic acid, can be used.[3,9,20] Common characteristics of these drugs are anti-inflammatory and anti-fibrogenic. One of the patients in our series had a history of FMF for 15 years and the patient was using regular colchicine. ACS was present in the patient despite the anti-inflammatory and anti-fibrogenic properties of the colchicine.

If the brid ileus or internal herniation is excluded, these patients with symptoms of bowel obstruction may be candidates for surgery if not respond to conservative treatment. The preferred method for the treatment of ACS is total or partial removal of the membrane.[3,8,9,11] Resection is rarely required, and anastomosis should be considered with caution in these patients.[3,9,21] There are a limited number of publications suggesting that mycophenolate mofetil may be useful in patients with frequent recurrent symptoms after surgery of membrane excision and adhesiolysis by laparoscopic approach in ACS.[16,22,23] Adhesiolysis and partial or full capsule excision were performed in all cases. In one patient, partial small bowel resection and end jejunostomy were performed due to the necrosis that arises from a vascular failure in the small intestine. In another patient, omentectomy was performed due to omentum necrosis. In one of the patients, colon perforation occurred during adhesiolysis and primary repair was performed. The patient was discharged without any problem.

CONCLUSION

Abdominal cocoon is usually diagnosed during operation in patients operated for reasons such as acute abdomen or obstruction. In late diagnosed cases, death can be seen due to small bowel necrosis and septic complications. High clinical suspicion and radiological imaging are important in the preoperative diagnosis. The treatment is consisted of surgical adhesiolysis and membrane excision.

Conflict of interest: None declared.

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ORİJİNAL ÇALIŞMA - *ÖZET*

Abdominal koza sendromu: Akut karın sendromunun nadir bir nedeni

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AMAÇ: Literatürde nadir görülen bir akut karın veya bağırsak tıkanıklığı nedeni, abdominal koza sendromu sklerozan peritonit veya sklerozan enkapsüle peritonit olarak tanımlanmaktadır. Abdominal koza, karın organlarının lifli bir zar ile tamamen veya kısmen sarılması ile karakterizedir. Genellikle genç kadınlarda görülmesine rağmen etiyoloji bilinmemektedir. Tanı genellikle laparotomi sırasında yapılır. Bu yazıda, operasyon sırasında abdominal koza tanısı almış yedi hasta sunuldu.

GEREÇ VE YÖNTEM: Hastanemiz genel cerrahi kliniğinde Ocak 2012 ile Kasım 2018 tarihleri arasında karın ağrısı ve/veya bağırsak tıkanıklığı nedeniyle laparotomi yapılan ve ameliyat sırasında abdominal koza tanısı alan hastaların kayıtları geriye dönük olarak incelendi. Hastaların demografik özellikleri, altta yatan etiyolojik faktörler, cerrahi işlemler, bulguları ve takipleri kaydedildi.

BULGULAR: Abdominal koza nedeniyle ameliyat edilen 7 hastadan dördü erkek, 3'ü kadındı. Ortanca yaş erkeklerde 61 (57-63), kadınlarda 39.6 (28-49) idi. Akut karın veya ileus tanısı ile altı hasta acil durumlarda ameliyat edildi. Elektif koşullarda karın içi kitle tanısı alan bir hasta ameliyat edildi. Yedi hastadan beşinde, ince bağırsakların tamamı bir fibrokollajen kapsül ile kaplıyken, 2 hastada bağırsaklar kısmen fibrokollajen kapsül ile kısmen sarıldı. Hastaların dördünde altta yatan hastalık yoktu, bir tanesinde Ailesel Akdeniz Ateşi (AAA), birinde endometriozis, birisinde beta-bloker kullanımı vardı. Ameliyat sırasında ince bağırsak nekrozu ve septik peritonit saptanan hasta 6 gün sonra kaybedildi. Diğer hastaların takibinde komplikasyon görülmedi ve hiçbir hasta nüks nedeniyle tekrar ameliyat edilmedi.

TARTIŞMA: Abdominal koza, akut karın veya bağırsak tıkanması gibi nedenlerle ameliyat edilen hastalarda genellikle ameliyat sırasında teşhis edilen bir durumdur. Geç tanı konulan olgularda ince bağırsak nekrozu ve septik komplikasyonlar nedeniyle ölüm görülebilir. Preoperatif tanıda yüksek klinik şüphe ve radyolojik görüntüleme önemlidir. Tedavi, fibrokollajen membranın eksizyonu ve adezyolizisdir.

Anahtar sözcükler: Abdominal koza, endometriozis; ileus; periton fibrozisi.

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