

A RARE ETIOLOGY OF INTESTINAL OBSTRUCTION: ABDOMINAL COCOON SYNDROME

NADİR GÖRÜLEN BİR İLEUS SEBEBİ: ABDOMİNAL KOZA SENDROMU

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SUMMARY

Introduction: Abdominal cocoon syndrome is one of the rare causes of intestinal obstruction, intestinal hemorrhage, and perforation. The small bowel is encapsulated completely or particularly by the fibro-collagenous membrane which was called the abdominal cocoon and it was reported mainly in young adolescent women as a cause of small bowel obstruction. Preoperative diagnosis is very difficult and most common it was diagnosed as the abdominal cocoon with preoperative findings.

Case: A 46-year-old male patient admitted emergency service with abdominal pain, no gas, and stool discharge. His physical examination was compatible with acute abdomen and mechanic intestinal obstruction. He was hospitalized and operated with the presumptive diagnosis of intestinal obstruction. Operative findings were consistent with abdominal cocoon, adhesiolysis was performed. Patient's postoperative course was unremarkable, and he was discharged on postoperative 8th day. Follow-up 28 months showed no clinical evidence of recurrence. The pathology was reported as fibrosis, inflammation and fibrin exudation, compatible with the abdominal cocoon.

Conclusion: Here in, we present the etiology, management and the treatment of abdominal cocoon syndrome which was a rare cause of mechanic bowel obstruction among male patients and difficult to diagnose preoperatively.

ÖZ

Giriş: Abdominal koza sendromu intestinal obstruksiyon, intestinal hemoraji ve perforasyonun nadir görülen sebeplerinden biridir. Abdominal kozada incebarsaklar tamamen veya kısmen abdominal koza olarak adlandırılan fibrokollajenöz yapıda bir membranal kaplı olup, bu hastalık daha çok genç adölesan kadınlarda intestinal obstruksiyon sebeplerinden biri olarak bildirilmiştir. Preoperatif tanı koymak güç olup, genellikle peroperatif bulgular eşliğinde tanı konulabilmektedir.

Olgu: 46 yaşında erkek hasta, acil servise karın ağrısı, bulantı, kusma ve gaz gayta çıkaramama şikayetleri ile başvurdu. Fizik muayenesi akut batın ve mekanik barsak obstruksiyonu ile uyumluydu. Hasta intestinal obstruksiyon ön tanısı ile acil operasyon amaçlı hospitalize edildi. Peroperatif görünümü abdominal koza ile uyumlu olduğu saptandı ve bridektomi yapıldı. Hastanın postoperatif takibi sorunsuz seyretti ve postoperatif

sekizinci gün taburcu edildi. Hastanın postoperatif 28 aylık takibinde klinik olarak nüks bulgusu gözlenmedi. Patoloji spesimeni, inflamasyon, fibrozis, fibrin eksudaları şeklinde olup, abdominal koza sendromu ile uyumlu olarak raporlandı.

Sonuç: Biz burada, nadir bir mekanik barsak obstruksiyonu sebebi olup, erkeklerde daha nadir görülen, preoperative teşhisi zor olan abdominal koza sendromunu, etyolojisini ve tedavi yönetimini sunmayı planladık.

INTRODUCTION

Abdominal cocoon syndrome is one of the rare causes of intestinal obstruction which was first described as abdominal cocoon by Foo et al. in 1978(1). It describes the small bowel is encapsulated completely or partly by the fibro-collagenous membrane. Several theories have been suggested for the etiology of this disease, but most of the cases had still unknown etiology (2). It is most commonly seen in adolescent girls of the tropical and subtropical region though, a few cases of the male have also been reported in the literature as in our case(1,3).

CASE

A 46-year-old male patient admitted to the emergency department with abdominal pain, vomiting, nausea and no gas and stool discharge persisting for two days. He has no history of operation and chronic illness except tuberculosis of a lung with treatment. On physical examination, there were signs of peritoneal irritation all quadrants of the abdomen with rebound tenderness and abdominal distention. Air-fluid levels were seen in plain abdominal X-rays. The patient underwent computed tomography (CT) abdomen which showed localized clumping of the small bowel in center of the abdomen with matting. The loops were seen within the thin-walled sac-like structure containing localized fluid. Mostly part of jejunal loops was embedded within the sac (Figure 1). The patient was then explored based on the diagnosis of the acute abdomen. Preoperative 50 cm. from treitz ligament to ileocecal valve; all small bowels were found to be under a thick and dense membrane and the obstruction site causing the symptoms was a short, small bowel segment (Figure 2). After adhesiolysis and resection of the membrane, the bowels were freed, leaving no narrowed segment. Simply adhesiolysis was performed with no resection. The postoperative

course was uncomplicated. The patient was allowed to feed via the oral route by the 4th postoperative day, and stool discharge occurred on the 4th postoperative day. He was discharged on the postoperative 8th day. Follow-up 28 months showed no clinical evidence of recurrence. The pathological findings revealed fibrosis, chronic nonspecific inflammation, hyalinization, collagenase and fibrosis of peritoneal tissue. (Figure 3).



Figure 1. The loops were seen within thin-walled sac-like structure containing localized fluid. Mostly part of jejunal loops were embedded within sac (arrow)



Figure 2. Intraoperative findings show the bowels which were covered with a thick membrane

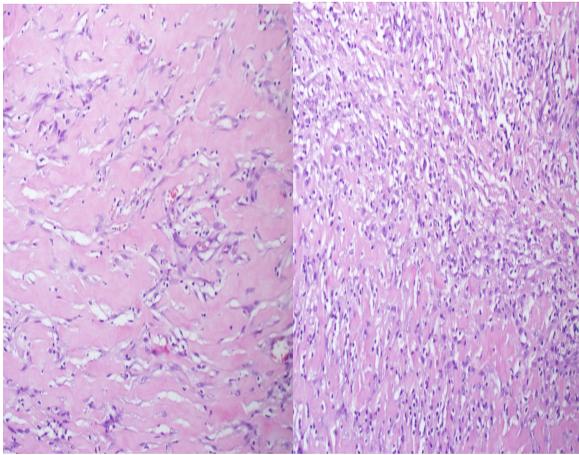


Figure 3. Revealed fibrosis, chronic non spesific inflammation, hyalinization, collaginased and fibrosis of peritoneal tissue.

DISCUSSION

Sclerosing encapsulated peritonitis (SEP), primer sclerosing peritonitis, sclerotic thickening of the peritoneal membrane, peritoneal fibrosis is the same terminology for abdominal cocoon syndrome (4,5). SEP describes the condition of partial or total small bowel with fibro-collagenous membrane, which is one of the rare causes of intestinal obstruction. It can be life-threatening with complications of small bowel obstruction, such as perforation or hemorrhage. Because of the nonspecificity of its early clinical features, and delayed the diagnosis is very difficult before the patient develops bowel obstruction.

Preoperative diagnosis of the abdominal cocoon syndrome is usually difficult (4). There are no reliable predictive tests for EPS. Therefore, the high index of suspicion is required in the diagnosis of this condition. The Radiological examination also plays a key role in the identification of the disease. Computed tomography (CT) is widely considered as the gold standard to image SEP(6,7). Intraoperative and histopathology findings are usually considered for the final diagnosis of an abdominal cocoon, as most patients underwent surgical intervention without CT imaging. Although radiological imaging aids in preoperative diagnosis, the definitive diagnosis is usually confirmed by laparotomy. Clinical presentation of this syndrome mostly occurs as an acute abdomen that in most cases, requires surgical intervention.

Several theories have been suggested regarding the etiology of the disease, but the predisposing factor prompts the patient to proceed to SEP has not exactly known (2). Abdominal surgery, generalized peritonitis, intraabdominal malignancy, intraperitoneal chemotherapy, ventriculoperitoneal shunt, cirrhosis, abdominal tuberculosis, b-blockers, peritoneal lavage, cytoreductive surgery and HIPEC, using certain disinfectants such as povidin iyot and peritoneal dialysis has been stated as relatedto SEP (8,9). In our case, the patient had no risk factors for SEP. He had only medical treatment with rifampicin and isoniazid for tuberculosis of the lung with which can be a risk factor for the etiology.

The exact pathogenesis of SEP is not known, but it is assumed to be due to peritoneal irritation, and an increased level of IL-1, with the proliferation of fibroblasts, causes inflammation and fibrosis. As a result of inflammation, intraperitoneal fibrin-like material is released by fibrogenic cytokines forming thick shiny membranes encasing whole or part of bowel giving a characteristic appearance to the disease.

Surgical management of the abdominal cocoon had a wide variety such as; only explorative laparotomy, partial membrane excision + adhesiolysis, resection + anastomosis, resection+ anastomosis + protective enterostomy. All treatment ways can be used alone or in combination, due to patient-related factors (10-12). We performed careful dissection and excision of the thick sac with the release of the small intestine surgical removal of the membrane which leads to complete recovery with open surgery (10).Nowadays laparoscopic approach can be a better choice for the diagnose and treatment which has advantages and disadvantages (11,12).

The advantage of laparoscopic approachis that it can used for both diagnostic and therapeutic purposes(11). However, there is a possibility of making a bowel injury during placing the first trocar due to adhesions.To prevent this, the first trocar can be placed with the open method (12).

However, surgery is the first choice of the treatment for syptomtatic SEP, the drugs such

as tamoxifen, corticosteroids, azathioprine, myophenolatemofetil and sirolimus can be used in the conservative treatment of mildly obstructive or asymptomatic patients which reported in the limited cases (12,13).

In conclusion, abdominal cocoon syndrome should be kept in mind, especially patients who had risk factors with symptoms of intestinal

obstruction and acute abdomen. The diagnosis of SEP is primarily based on the clinical finding, and then generally confirmed with radiological signs especially by CT. The laparoscopic approach may be better for diagnose and the treatment.

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