An association between transmesenteric internal hernia and abdominal cocoon syndrome: A case report

Tansu Altintas*, İnanç Şamil Sarıcı, Mustafa Uygar Kalaycı
Kanuni Sultan Süleyman Training and Research Hospital, Department of General Surgery, Istanbul, Turkey

**ABSTRACT**

**INTRODUCTION:** Transmesenteric internal hernia is defined as the herniation of the small intestine from a mesenteric defect in the abdominal cavity, and abdominal cocoon syndrome is the partial or entire encapsulation of the small bowel like the shape of an accordion, by a fibrocollagenous membrane.

**PRESENTATION OF CASE:** A 32-year old male patient applied with complaints of abdominal pain, nausea, and vomiting bile. Signs visualized in the abdominal computer tomography were as follows: gatto formation of the small intestinal loops and suspected of an internal hernia. In the operation, a membrane was detected encapsulating the entire intestine resembling a tube, making the intestines to appear like an accordion and an opening was present in the small intestinal mesentery. The intestine was separated from the defect, and placed in its normal anatomical position. The defect in the mesentery was closed and the encapsulating membrane was removed from small intestine. Intestinal resection was not required.

**DISCUSSION:** Internal hernias comprise less than 1% of all intestinal obstructions, and are formed by the herniation of the intestine and mesentery into the opening of the visceral peritoneum or into the recessus. Abdominal cocoon syndrome is a disorder characterized by the partial or total encapsulation of the small intestine by a thick and fibrotic membrane. Preoperative diagnosis is very difficult and is generally diagnosed during laparotomy exploration.

**CONCLUSION:** The association of internal herniation and abdominal cocoon syndrome is an extremely rare cause of mechanical intestinal obstruction. If not promptly diagnosed and treated, can lead to serious complications.

© 2016 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

1. Introduction

Internal herniations are defined as the protrusion of small intestinal segments into the anatomical or non-anatomical spaces within the peritoneum, and they include less than 1% of cases with intestinal obstruction [1,2]. Intestinal obstruction associated with internal herniation may exist in any age; however internal hernias due to congenital transmesenteric defects are mostly reported in pediatric age groups and frequently concomitant with intra-abdominal anomalies like intestinal atresia. In adults the defect is mostly acquired as a result of blunt abdominal trauma or previous intestinal and mesentery surgeries [3]. In the absence of an anamnesis, revealing a previous abdominal surgery, a preoperative diagnosis of these herniations is difficult and therefore rapidly progress to a life-threatening intestinal ischemia. Due to delays in diagnosis and treatment mortality rates can rise up to 20% [4].

Abdominal cocoon syndrome is a rare condition, which is the partial or complete encapsulation of the intestines by a fibro-collagenous membrane resembling an accordion, existing as an acute or subacute intestinal obstruction. Mostly observed in young adolescent girls, it was first defined in 1868 and exists with non-specific signs, generally diagnosed during surgery [5,6]. Causes of fibrosis have been claimed in its physiopathology and include the retrograde menstruation, previous abdominal surgery, use of beta-blockers, peritoneal dialysis, cirsrhosis, Lee Veen shunt and idiopathic cases exist congenitally in which no cause leading to cocoon formation can be determined. Both transmesenteric internal herniation and abdominal cocoon syndrome causes of intestinal obstruction that rarely exist on their own.

We present here an extremely rare case of an association of transmesenteric internal herniation and abdominal cocoon syndrome, which we operated on due to signs of acute abdomen and mechanical intestinal obstruction, and was diagnosed during the operation.

*Corresponding author at: Bahçeşehir 1. Kism Mahallesi İskıtkule Caddesi Vaditepe Bahçeşehir 5. Bölge Sitesi K01 Blok No:33D iç kapı No:16 Başakşehir/Istanbul, Turkey.
E-mail address: issarici2015@gmail.com (T. Altintas).
2. Case report

A 32-year old male patient applied with complaints of abdominal pain, nausea, and vomiting bile, which had started the previous day. His medical history revealed absence of a known chronic disease or drug use, undescended testicle surgery at 8 years old, and a defecation habituation of once every two days. His family history showed no specific property. His physical examination body temperature 37.9 °C, pulse 100/min, blood pressure 120/70 mmHg, and respiratory rate 16/min. A slight distension, and a generalized tenderness that was more marked in the epigastric area were detected in the abdominal examination; abdominal defense was absent and rebound phenomenon was found to be positive. Direct abdominal X-ray taken in the upright position revealed several air-fluid levels. Abdominal ultrasonography evaluation revealed no pathological sign, except a slight dilation of the intestinal loops. Laboratory analysis revealed the following: leukocyte 6570/mm³, Hb 14.2 gr/dl, Plt 246,000/mm³, CRP 8.9 mg/L. In abdominal CT minimal fluid in the perisplenic area, gatto formation of the small intestinal loops in the abdominal midline and left lateral and an image in the right side suspected of an internal hernia (Figs. 1 and 2).

The patient underwent surgery for the mechanical intestinal obstruction. The small intestine was dilated in appearance and marked abdominal adhesions were observed. A membrane was detected encapsulating the entire intestine resembling a tube, making the intestines to appear like an accordion (Fig. 3). In the right, an opening was present in the small intestinal mesentery; the rest of the small intestine was observed to herniate from this opening, together with the encapsulating membrane (Fig. 4). The intestine was separated from the defect, and placed in its normal anatomical position. The defect in the mesentery was closed. During the first exploration the length of the small intestine was observed about one meter; this length was discovered to be 3.5–4 m following the removal of the encapsulating membrane. Intestinal resection was not required.

Histopathological examination of the membrane revealed fibrous connective tissue with chronic inflammation, congestion, and edema, showing scleral hyaline changes. Bowel movements began on the post-operative fifth day. The patient was discharged from the hospital with recovery, on the post-operative seventh day. In his follow-ups after hospitalization, no abnormalities existed in the gas or feces discharge; however, he experienced loss of appetite, nausea, vomiting, indigestion, and about 8 kg of weight...
loss. His complaints lasted for nearly two months, showing a gradual decrease. In the period following these two months, he felt relieved, and regained his previous weight.

3. Discussion

Internal hernias comprise less than 1% of all intestinal obstructions, and are formed by the herniation of the intestine and mesentery into the opening of the visceral peritoneum or into the recessus [2]. Internal herniations are so named due to the openings they pass through, or the position of the herniated organ. According to these considerations, they exist in different types, as paraduodenal, foramen of Winslow, pericecal, intersigmoid, transmesenteric, transomental, and retroanastomotic. The type most frequently seen is the paraduodenal hernia, which forms 53% of all internal herniations [7]. In some series, the transmesenteric type is more frequently observed in children [4]. When replaced and gath- ered segments of the small intestine, and rotational movements of the mesenteric vascular structures supplying these intestinal segments are visualized in an abdominal CT, internal herniation has to be suspected. The time duration for the progress of an intestinal obstruction to the intestinal ischemia may be short. Therefore, losing time through conservative treatment may be life threatening in these cases. The clinical status of the patient has to be the primary guide when deciding on surgery. Morbidity and mortality can be reduced by a last minute intervention [4].

Abdominal cocoon syndrome is a disorder characterized by the partial or total encapsulation of the small intestine by a thick and fibrotic membrane [8], incidences of which are reported to occur between 0.4% and 5.5% [9]. Idiopathic abdominal cocoon occurs congenitally, in which no cause leading to cocoon formation can be detected [8]. Secondary abdominal cocoon formation is determined more frequently, as a result of abdominal surgery, the use of beta-blockers (practolol), peritonitis related with peritoneal dialysis, Lee Veen shunt, cirrhosis and irregular menstruation. Its etiology includes various acquired factors; however the common fact for all is the existence of a condition that leads to an intraabdominal reaction [8,10]. Preoperative diagnosis is very difficult and is generally diagnosed during laparotomy exploration [7]. Laboratory findings do not reveal any sign of it being specific to the abdominal cocoon [11]. Routinely taken direct abdominal graphies in the upright position do not indicate a specific sign, except the gas-fluid levels [11]. In CT, the gathering of small intestinal segments in a single area, and a membrane with soft tissue density surrounding them, are the classic signs. Other CT signs include the collection of ascites or locu- lar fluid, peritoneal thickening and contrast trapping, peritoneal calcifications, and the thickening of the intestinal wall [11].

As a conclusion, the association of internal herniation and abdominal cocoon syndrome is an extremely rare cause of mechanical intestinal obstruction. Its treatment includes the excision of the cocoon by careful dissection, the reduction of the hernia, the resection/anastomosis of the devitalized intestine, and repair of the mesenteric defect. Time duration for regaining the small intestinal functions is longer in these patients compared with other cases with ileus, and these functions become regular over a longer time. If not promptly diagnosed and treated, can lead to serious complications such as intestinal ischemia, perforation, sepsis and death. The most important goal of management is early identification of the disease which provides a better prognosis.

This case report is compliant with the SCARE Guidelines and criteria [12].

Conflicts of interest

The authors have no conflict of interest.

Funding

We have no source of funding for our research.

Ethical approval

Number of Ethics committee number: 286–2016. Kanuni Sultan Suleyman Training and Research Hospital Ethics Committee.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Writing the paper: İnanc Samil Sarici.

Consept or design: Mustafa Uygar Kalayci.

Data Collection: Tansu Altintas.

Guarantor

İnanc Samil Sarici accept full responsibility for the work and the conduct of the study, had access to the data, and controlled the decision to publish.

References

[1] O.A. Catalano, A. Bennicenga, M. Abbate, E. Tomei, M. Napolitano, A. Vanzulli, Internal hernia with volvulus and intussusception: case report, Abdom. Imaging 29 (2004) 164–165.
[2] A. Tékin, M. Sahin, T. Küçükakkartallar, A. Kaynak, Nadir bir iileus nedeni: paraduodenal herni, Genel Tip Derg. 17 (2007) 111–114.
[3] Z. Rehman, S. Khan, Large congenital mesenteric defect presenting in an adult, Saudi J. Gastroenterol. 16 (2010) 223–225.
[4] H.P. Fan, A.D. Yang, Y.J. Chang, CW. Juan, H.P. Wu, Clinical spectrum of internal hernia: a surgical emergency, Surg. Today 38 (2008) 899–904.
[5] M.H. Chew, I. Sophian Hadi, G. Chan, H.S. Ong, W.K. Wong, A problem encapsulated: the rare peri toneal encapsulation syndrome, Singapore Med. J. 47 (9) (2006) 808–810.
[6] M. Kaplan, N.M. Atabek, B. Salman, O. Durmus, A. Abbasova, X. Mustafayev, Bir olgu nedeniyle sklerozan enkapsule peritonit, Genel Tip Derg 12 (4) (2002) 147–150.
[7] A. Blachar, M.P. Federle, S.F. Dodson, Internal hernia: clinical and imaging findings in 17 patients with emphasis on CT criteria, Radiology 218 (2001) 68–74.
[8] D.O. Irabor, O. Atalabi, Case report: abdominal cocoon, West Afr. J. Med. 20 (October–December) (4) (2001) 265–267.
[9] Kawaguchi, et al., Encapsulating peritoneal sclerosis: definition, etiology, diagnosis, and treatment, Perit. Dial. Int. 20 (Suppl. (4)) (2000) 543–555.
[10] W.K. Elingtonham, H.J. Espriiner, C.W. Windsor, D.A. Griffiths, J.D. Davies, H. Babdeley, A.E. Read, R.J. Blunt, Sclerosing peritonitis due to practolol: a report on 9 cases and their surgical management, Br. J. Surg. 64 (April (4)) (1977) 229–235.
[11] J.D. Wig, S.K. Gupta, Computed tomography in abdominal cocoon, J. Clin. Gastroenterol. 20 (1998) 616–157.
[12] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, S.C.A.R.E. the Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. (2016) (article in press).

Open Access

This article is published Open Access at sciencedirect.com. It is distributed under the IJSCR Supplemental terms and conditions, which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.