ORIGINAL RESEARCH PAPER

Internal Medicine

IDIOPATHIC ABDOMINAL COCOON - A RARE CAUSE OF SMALL BOWEL OBSTRUCTION

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ABSTRACT

The idiopathic form of sclerosing encapsulating peritonitis, also known as abdominal cocoon, is a rare entity of unknown cause that leads to intestinal obstruction due to complete or partial encapsulation of the small intestine by a fibro-collagenous membrane. As the initial clinical features are non-specific, they often remain unrecognized making it difficult to make a definite pre-operative diagnosis. Recurrent episodes of small intestinal obstruction along with relevant imaging findings and lack of other causative mechanisms, gives rise to a clinical suspicion. We report a young lady who presented to us with features suggestive of acute intestinal obstruction. Thorough enquiry revealed similar episodes on multiple occasions in the past which was temporarily relieved with conservative management. The need to keep an open mind regarding the uncommon causes of commonly encountered problems is the point of interest in our case.

INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) was first described by Owtschinnikow in 1907 as peritonitis chronic fibrosa incapsulata. (Mohanty et al., 2009) This rare condition of unknown etiopathology is predominantly prevalent in adolescent girls living in the tropics. Whether idiopathic or secondary, SPE can be distinguished by a thick grayish-white fibrotic membrane, completely or partly encasing the small bowel, and can extend to engage other organs like the large bowel, liver and stomach. The idiopathic form, also known as abdominal cocoon, was first observed by Foo et al in 1978. Although post-operative adhesions are the culprit in 3 out of 5 patients with obstruction of the small intestine, merely 6% of the cases are unusual.

Retrograde menstruation with a superimposed viral infection, retrograde inflammation of the peritoneum and gynaecological pathology inducing cell-mediated immunological tissue damage are some of the hypotheses that have been proposed to explain this rare entity. However, since there are reports of males, pre-menopausal females and children who have been affected by this condition, these hypotheses hold extremely poor credibility. (Xu et al., 2007) Further theories are needed to account for idiopathic SEP.

Pre-operative diagnosis of abdominal cocoon is difficult and warrants a high index of clinical suspicion. The diagnosis should always be considered in a patient who reports episodes of abdominal pain, nausea, vomiting and unintentional loss of weight. With the help of imaging techniques such as ultrasound and CT scan, a pre-operative diagnosis can be made to prevent unnecessary bowel resection. Release of the small intestine after cautious dissection and excision of the cocoon will lead to complete recovery.

Case Report

In November 2022, a 31year old lady presented to our emergency with complaints of severe generalized abdominal pain since 24 hours. This was associated with 14 episodes of vomiting over the same duration. The vomitus initially contained food particles but later turned bilious. There were similar episodes occurring every 2-3 months ever since her pre-pubertal days, with spontaneous resolution within 48 hours. At the age of 17years, following one such episode, she underwent appendicectomy. However her symptoms persisted post-operatively.

She was empirically managed at ER with intravenous (IV) fluids, anti-spasmodic agents and anti-emetics; blood investigations were non-contributory. Abdominal ultrasound gave the impression of prominent central bowel loops and minimal ascites.

Contrast enhanced CT scan showed dilated proximal and mid ileal loops with evidence of a transition at distal ileal level, a stricture / band obstruction at the level of the pelvic brim near midline, small bowel fecal sign in the distal ileal loop, the terminal most ileum along with the ileocecal junction and the entire colon was found to be collapsed - features suggestive of small bowel obstruction, and small free fluid in the abdomen. ***Fig. 1***

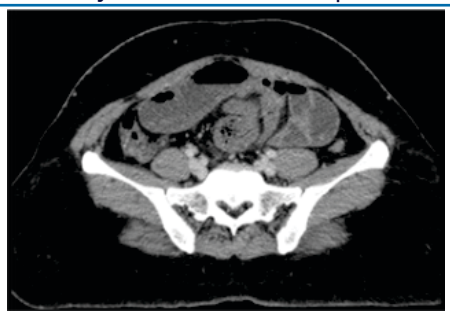

Fig.1 Venous phase of CT abdomen showing transition at distal ileal level

In view of worsening symptoms, emergency exploratory laparotomy was planned. With all aseptic precautions and adequate antibiotic coverage, the abdomen was explored – dilated small bowel loops, cocoon formation involving whole of the mid and terminal ileum, and clotted mucinous fluid in the peritoneal cavity were noted. The intestinal cocoon was released, decompression and toileting was done under general anaesthesia. Analysis of the aspirated fluid revealed normal study. Histopathological examination of wall of the cocoon showed dense poorly cellular fibrous tissue with congested vessels of varying caliber and a cyst lined by ciliated columnar epithelial cells. Post-operatively, she underwent an uneventful recovery and has experienced no further symptoms in the last 4months post-discharge. ***Fig. 2, 3***

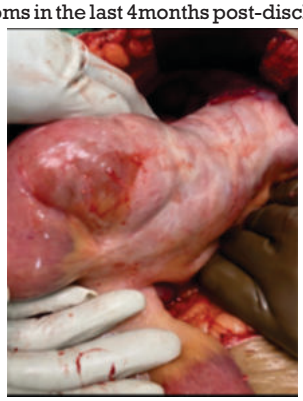

Fig. 2 Intraoperative finding: mid and terminal ileum encased by the peritoneum

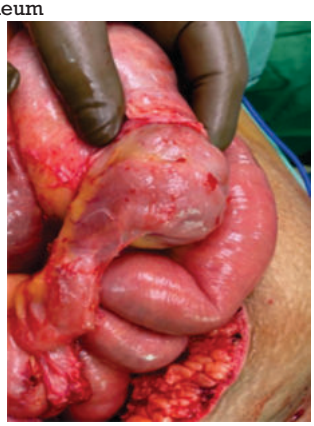

Fig. 3 Intraoperative finding: releasing the intestine from the abdominal cocoon.

DISCUSSION

Abdominal cocoon is often associated with other embryologic abnormalities like hypoplasia of the greater omentum and malformation of the mesenteric vessels, hence developmental abnormality may be a probable etiology. Ambulatory chronic peritoneal dialysis (PD) has been known to be associated with secondary SEP. (Afthentopoulos et al., 1998) Those undergoing PD may complicate as SEP which in turn leads to decreased ultrafiltration and ultimately intestinal obstruction. Uncommon causes of secondary SEP include a past history of abdominal surgery, subclinical primary viral peritonitis, repeated episodes of peritonitis, use of medications such as beta-blocker and peritoneo-ventricular shunting. Abdominal tuberculosis, sarcoidosis, familial Mediterranean fever, intraperitoneal chemotherapy, cirrhosis, liver transplantation, malignancy of the gastrointestinal tract, luteinized ovarian thecomas, endometrial pathology, deficiency of protein S, rupture of dermoid cyst and presence of fibrogenic foreign material are some of the extremely rare causes of secondary SEP. (Tannoury JN et al., 2012)

Depending on the extent of membrane encasement, SEP can be categorized into 3 types - in type 1 SEP, the small intestine is partially encased by the membrane, the whole small intestine is encapsulated by the membrane in type 2, whereas, the entire small intestine as well as other intra-peritoneal organs such as stomach, colon, liver and ovaries are inside the membrane in type 3. (Xia et al., 2018) In a systematic review of 118 patients, it was reported that type 1 occurred in 43%, type 2 was seen in 31% and only one-fourth of the patients had type 3 SEP. (Machado, 2016) With the help of radiological imaging as well as intra-operatively, type 1 abdominal cocoon was observed in our patient.

The most commonly reported manifestations of intestinal obstruction in cases of SEP are abdominal pain 72%, distended abdomen 44%, mass in the abdomen 30%, and nausea/ vomiting, as reported by Machado et al. in 2016. The mean age of presentation in this study was 39 years. The condition was present in acute, subacute or chronic form. (Machado, 2016)

Thorough medical history, complete physical examination, a high index of suspicion complemented with a laboratory and radiological investigations is needed to make a timely diagnosis. Internal hernia is a close differential diagnosis to SEP with a similar CT finding; however, the membrane-like sac is usually absent in the scan. Conventionally, SEP is an intra-operative diagnosis with a thick membrane enclosing the intestine; however, recent advances in radiological imaging and interpretation have made possible pre-operative diagnosis of this condition. Diagnosis prior to surgery can help tailor the procedure and prevent any unnecessary operative intervention or bowel resection.

Sonography reveals dilated loops of small intestine, trilaminar appearance of the intestinal wall, membrane formation, and attachment of the bowel to the posterior abdominal wall. (Tannoury JN et al., 2012) A highly sensitive radiological modality in current use is the abdominal CT scan where we can visualize several radiological characteristic signs of SEP such as small bowel dilatation at the midline enveloped by a sac or a thick membrane, small amounts of encapsulated effusion in the sac, intestinal obstruction, thickened mesentery, absent or hypoplastic greater omentum, calcified small intestinal wall, enlarged lymph nodes etc. (Karona et al., 2021:Yu et al., 2019)

Exploratory laparotomy with complete membrane excision and adhesiolysis is the conventional approach to cases with complete bowel obstruction. In order to avoid iatrogenic intestinal or vessel injury, it is of utmost importance that manipulation during surgery be done delicately and precisely to completely release the neck of the sac along the duodeno-jejunal junction and superior mesenteric vessel. (Dave et al., 2019) Conservative approach towards management of intestinal obstruction may be attempted in a hemodynamically stable patient with virgin abdomen.

CONCLUSION

Non-specific clinical presentation of intestinal obstruction should raise an alert regarding the possibility of existing

abdominal cocoon. A high index of clinical suspicion along with appropriately timed radiological investigations with increase the chance of pre-operative diagnosis of abdominal cocoon. The aim of this article is to create awareness of this condition typically presenting with features of intestinal obstruction. Intra-operative finding of a thick membrane encasing the bowel stamps the diagnosis. Extensive studies are the needed to elucidate the etiology of idiopathic SPE. Further studies on the pathogenesis of secondary SEP and well as their outcome can help explore the surgical and conservative treatment options of this condition.

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