

## CASE REPORTS

# Abdominal Cocoon – A Case Report with Short Review of Literature

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### Abstract:

Abdominal cocoon is a rare entity where there is encapsulation of the small bowel by a fibrous membrane causing clustering of the bowel. The exact aetiology is unclear. We report a case of a 15 year old girl with a history of recurrent colicky abdominal pain in the right lower abdomen. Clinical diagnosis was recurrent appendicitis. She underwent laparotomy for appendicular lump. Histopathological examination of the resected lump grossly and histologically revealed the features of abdominal cocoon.

**Key Words:** Abdominal Cocoon, Sclerosing encapsulating peritonitis, Small bowel.

[BSMMU J 2009; 2(2): 81-84]

### Introduction:

Cocoon is a covering of silk threads that some insects make to protect themselves before they become adult. Abdominal cocoon is a peculiar morphologic entity characterized by encasement of part or the whole of the small bowel by a fibrocollagenic cocoon like case<sup>1-3</sup>. It is a rare condition of unknown aetiology that primarily affects adolescent girls living in tropical and subtropical regions<sup>4</sup>. Abdominal cocoon usually presents as recurrent acute or sub acute intestinal obstruction with or without a mass<sup>3</sup>. About 60 articles dealing with this topic was found in Medline search till date<sup>2</sup>. Here we report a case of abdominal cocoon in a young girl with history of intermittent colicky pain in right lower abdomen for about a year. Clinically it was diagnosed as recurrent appendicitis. Laparotomy was done and resection of the lump was performed and the specimen was sent for histopathology with a provisional diagnosis of intestinal tuberculosis.

### Case Report

A 15 year old girl from Narsingdi presented with a history of intermittent colicky pain in the right lower abdomen for one year. The pain aggravated after taking food. There had been 7 to 8 episodes of similar attack of pain during last one year which was relieved by taking medications (homoepathic drugs). She had a history of weight loss and occasional fever. During her last attack she consulted a local physician. Clinical diagnosis was recurrent appendicitis. On examination she was found anaemic, pulse rate was 68/min and blood pressure was 100/70 mm Hg. Routine laboratory tests revealed a total leucocyte count of 6500 cell/cmm,

polymorphonuclear leucocytes were 69% and lymphocytes were 29%. Her haemoglobin was 12.6gm/dl and ESR was 10 mm and a normal urine analysis. Ultrasonogram (USG) of the whole abdomen was done. There was a mixed echogenic lump in the right iliac region (64mm X 43mm) which gave an impression of appendicular lump. The whole abdomen was otherwise normal. She was admitted to a local clinic and underwent laparotomy in April 2009. On opening the abdomen a huge mass was found in the terminal ileum. Resection of the terminal ileum with caecum and proximal part of the ascending colon was done followed by ileocolonic anastomosis, as stated in the operation note. Recovery of the patient was uneventful.

The surgical specimen was sent to the Department of Pathology, BSMMU with a provisional diagnosis of intestinal tuberculosis. On gross examination there was a long loop of intestine measuring 26 cm in length. Central portion of the loop was dilated measuring 10 cm in diameter (Fig.-1), located 10cm from the proximal resection margin



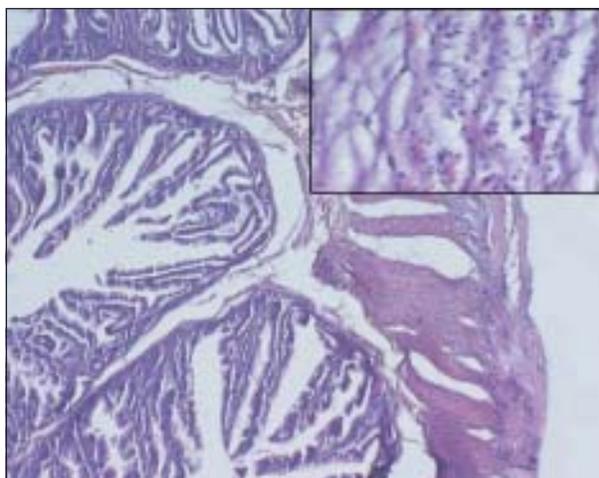
**Fig.-1:** Gross view of the resected intestine

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and 6cm from the distal resection margin. Caecum or ascending colon could not be identified. The serosa was apparently unremarkable. On opening the dilated portion showed multiple luminal structures clustered together and covered by thick fibrous tissue. Majority had thickened mucosa resembling intestinal mucosa and few had thin mucosa (Fig.-2). The average diameter of the luminal structures was about 2.5cm. Three lymph nodes were found. Multiple sections were given from the luminal structures including the lymph nodes. On microscopic examination the sections from the luminal structures revealed only lymphoid hyperplasia in the submucosa. There was moderate infiltration of chronic inflammatory cells in the serosa with fibrosis (Fig 3). Lymph nodes revealed features of reactive changes. There was no evidence of granuloma or malignancy. The diagnosis was 'Primary Abdominal Cocoon'.



**Fig.-2:** Cut section of the dilated part of the intestine



**Fig.-3:** Microscopic section of the intestinal wall shows lymphoid hyperplasia in the submucosa (H & E X 200). Inset shows microscopic section of the serosa showing thickening with chronic inflammatory cell infiltration (H & E X400)

### Review of literature

Abdominal cocoon is a rare condition<sup>1,3-7</sup> that refers to total or partial encapsulation of the small bowel by a thick fibrocollagenous membrane or cocoon like membrane and adhesions causing clustering of the bowel with local inflammatory infiltrate leading to acute or chronic bowel obstruction<sup>1,3,6-8</sup>. Occasionally the large bowel, stomach, liver or other abdominal organs may be involved<sup>2,3,6</sup>.

### Historical background

Abdominal cocoon was first described in 1907 by Owstchinnikow as peritonitis chronica fibrosa incapsulata<sup>3</sup>. Brown et al.<sup>9</sup> described the condition as a complication of long term treatment with beta adrenergic blocking agents. The abdominal cocoon was first described and named in 1978 by Foo et al.<sup>10</sup> Abdominal cocoon, a rare condition primarily affects young females from tropical and subtropical regions<sup>4,7</sup> but adult case reports from temperate zones and in both genders can be encountered in literature<sup>3,4,7</sup>. A Medline search revealed that in English literature approximately 47 cases have been reported till 2006<sup>4</sup>. Another study reported about 60 articles dealing with this topic found in Medline search<sup>2</sup>. This condition has been variously described as sclerosing peritonitis, encapsulating peritonitis or sclerosing encapsulating peritonitis (SEP)<sup>5,11</sup>.

### Aetiological factors

Abdominal cocoon is of two types primary or idiopathic and secondary<sup>2,3,5,12</sup>. The primary or idiopathic abdominal cocoon is a rare condition mainly described in young girls from tropical regions<sup>2,3,12</sup>. To explain the aetiology and the formation of the membrane of this condition a number of hypotheses have been proposed. These include retrograde menstruation with a superimposed viral infection<sup>1,3,4,8,10</sup> retrograde peritonitis via fallopian tubes, and cell mediated immunological damage incited by gynaecological infection<sup>13</sup>. However none of these hypotheses explain the characteristic age group, sex, and geographical distribution of this disease and there is no objective evidence to substantiate them<sup>4</sup>.

Secondary abdominal cocoon has been reported following long term use of the beta blocker practolol<sup>9,14,15</sup> or associated with sarcoidosis, SLE, liver cirrhosis, chronic ambulatory peritoneal dialysis (CAPD), intraperitoneal instillation of drugs, leiomyomata of the uterus, ovarian endometriosis<sup>16</sup> or tumours of the ovary, tuberculous pelvic inflammatory disease<sup>2,3,5,6,14</sup>. These conditions may predispose patients to peritoneal irritation and inflammation, which as a final effect leads to peritoneal fibrogenesis<sup>3,6</sup>.

### Clinical features

Patients usually presents with features of acute/subacute small bowel obstruction, symptoms of chronic obstruction, progressive nausea and vomiting, weight loss and/or colicky pain associated with an abdominal lump<sup>2-4,6,7,16</sup>. A preoperative diagnosis is almost never made and the non specific and intermittent symptoms may result in delay in diagnosis<sup>4</sup>.

Most cases are diagnosed incidentally at laparotomy although a preoperative diagnosis is purported feasible by a combination of barium follow through (concertina pattern or cauliflower sign and delayed transit of contrast medium)<sup>3,5-7</sup> and computed tomography of abdomen may be more diagnostic<sup>3</sup> demonstrating small bowel loops congregated to the center of abdomen encased by a soft tissue density mantle, peritoneal thickening, calcification, peritoneal enhancement, small bowel tethering and loculated fluid collection<sup>1,5,6,8</sup>. USG may show clumping of bowel loops with the bowel surrounded by a thick rim of hypochoic tissues<sup>6</sup>. However, preoperative diagnosis of abdominal cocoon requires a high index of clinical suspicion<sup>7</sup>, supported by clinical data and image findings indicative of the condition<sup>16</sup>.

Clinicians must rigorously pursue a preoperative diagnosis, as it may prevent a 'surprise' upon laparotomy and unnecessary procedures for the patient such as bowel resection<sup>7</sup>. Most cases were diagnosed when a laparotomy was performed for obstructive symptoms<sup>9</sup>. The characteristic findings is that of the encasement of the whole or part of the small bowel by a thick shiny membrane, aptly simulating cocoon. The loops of the small bowel remain stuck together by filmy soft adhesions separated easily by blunt or sharp dissections from the cocoon<sup>4,12</sup>. Histopathological examination of the encapsulating membrane shows thickened fibrocollagenous tissue with or without lymphocytic and plasma cell infiltration<sup>2-5</sup>. Regional lymph nodes demonstrate non specific reactive hyperplasia<sup>10,12</sup>. The final diagnosis of abdominal cocoon is usually made based on intraoperative and histological findings<sup>1</sup>.

### Management

As conservative management often fails<sup>3</sup> surgery remains the cornerstone in the management of abdominal cocoon<sup>4</sup>. Surgery includes careful dissection and excision of the thick sac with release of small intestine and adhesiolysis of small bowel loops<sup>3-5,7,16</sup> which leads to complete recovery<sup>1,4,7</sup>. Resection of bowel is unnecessary and increases morbidity and mortality and is indicated only if it is nonviable<sup>7</sup>.

### Differential diagnosis

Abdominal cocoon or sclerosing encapsulating peritonitis (SEP) may be confused with a developmental anomaly where the whole of the small bowel is encased in a thin membrane. The clinical symptoms differ from those of the abdominal cocoon in that the patients are mostly asymptomatic and the findings are incidental and late in life<sup>5,12</sup>.

### Prognosis

The prognosis of abdominal cocoon after surgery seems excellent and no recurrence has been described<sup>4,17,18</sup>.

### Conclusion:

Although abdominal cocoon is a rare entity, it can be diagnosed preoperatively as it may have a distinct appearance on barium follow through and CT of the abdomen and also with high index of clinical suspicion. In this case however only USG was done which gave an impression of an appendicular lump.

This case report intends to raise an awareness and enable earlier preoperative diagnosis and prevent unnecessary bowel resection as careful dissection and excision of the thick sac with release of small intestine and adhesiolysis of small bowel loops leads to complete recovery.

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