

# Abdominal cocoon associated with endometriosis

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## Abstract

**Abdominal cocoon is a rare cause of intestinal obstruction in adults. Diagnosis is usually established at laparotomy in patients with recurrent attacks of non-strangulating small bowel obstruction. A 40-year-old infertile Brazilian woman with intestinal obstruction and massive haemoserous ascites, due to coexistent ovarian endometriosis and abdominal cocoon, is reported. Abdominal pain, nausea, vomiting and a palpable mass, in addition to imaging of small bowel obstruction and thickened peritoneum, raised diagnostic suspicion. Higher awareness allows for early diagnoses and yields better results during management.**

**Keywords: abdominal cocoon, ascites, endometriosis, haemoserous ascites, intestinal obstruction, peritoneal encapsulation syndrome**

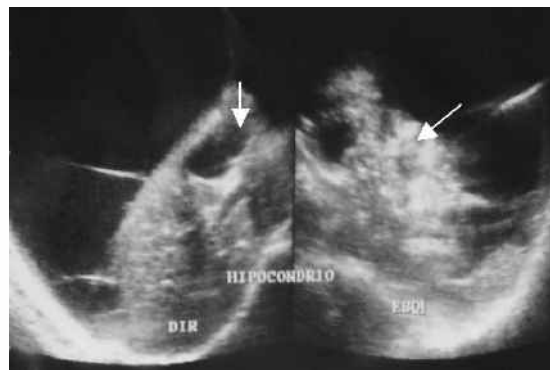
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## INTRODUCTION

Encapsulating peritonitis is an infrequent cause of intestinal obstruction in adult patients. The peritoneal membrane, which involves the mesentery, intestines, stomach, liver, gallbladder, spleen, and pelvic organs, was first described and named the abdominal cocoon (AC) by Foo et al in 1978.<sup>(1)</sup> We report a middle-aged Brazilian infertile woman, who had intestinal obstruction and massive haemoserous ascites due to coexistent AC and left ovarian endometriosis. Endometriosis has rarely been associated with AC or haemoserous ascites.

## CASE REPORT

A 40-year-old Brazilian woman was admitted for complaints of upper abdominal pain, vomiting and a weight loss of 11 kg in two months. There were no renal, cardiac, respiratory or urinary changes. She was a nulligravida, and in the last year, during laparoscopic investigation for a longstanding amenorrhoea, the diagnosis of ovarian endometriosis was established. She used a GnRH-analogue for six months (goserelin



**Fig. 1a** US image shows the heterogeneous mass (arrows) in the epigastrium and right hypochondrium.



**Fig. 1b** Abdominal CT image shows the small bowel loops clustered together (arrows).

3.6 mg dose every 28 days), and persisted without symptoms until two months before admission. There was no history of any other medication, significant illness or abdominal surgery. She was afebrile, with a body mass index of 14.7 kg/m<sup>2</sup>, ascites and a palpable mass in the mesogastrium. Laboratory data showed a hypochromic microcytic anaemia. Electrolytes, renal, liver and thyroid functions, and urinalysis were within the normal ranges; and the tuberculin skin test showed strong reaction. The erythrocyte sedimentation rate, C-reactive protein, antinuclear factor, C3 and C4 complement levels were unremarkable. Diagnostic abdominal tap showed haemoserous fluid with 0.3

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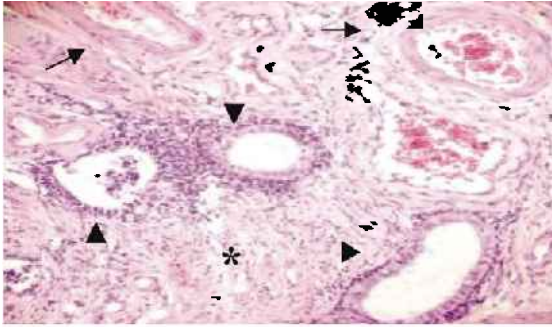
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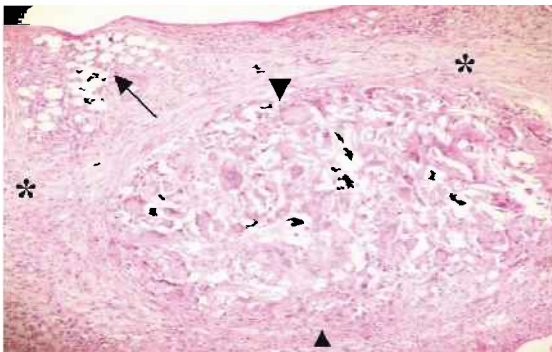
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**Fig. 2a** Photomicrograph of the biopsy specimen shows intense fibrosis (asterisk) involving endometrial glands (arrowheads) and inspissated vessels (arrows) in the peritoneal membrane (Haematoxylin & eosin,  $\times 150$ ).



**Fig. 2b** Photomicrograph of the biopsy specimen shows another aspect of the conspicuous peritoneal fibrosis (asterisks), with residual fat cells (arrow), mononuclear infiltrates and giant cell granuloma (arrowheads) (Haematoxylin & eosin,  $\times 150$ ).

serum-ascites gradient of albumin; 160 leukocytes/dL (76% mononuclear); albumin 3.5 g/dL and globulin 0.9 g/dL; without bacteria or malignant cells; normal adenosine deaminase titre; and negative culture for *Mycobacterium tuberculosis*. Chest radiograph was normal. Abdominal ultrasonography revealed gross loculated ascites and a huge heterogeneous mass in the epigastric and right hypochondrial areas (Fig. 1a). Abdominal computed tomography (CT) showed massive ascites and distended small bowel loops clustered together into the right upper quadrant and mid-abdomen (Fig. 1b).

Laparoscopy and exploratory laparotomy were performed, and multiple samples were obtained from the conspicuous membrane and cavity lining for histopathological studies (Figs. 2a–b). At laparoscopy, a cocoon-like white thickened peritoneum was seen encasing small intestine loops, in the presence of haemorrhagic ascites. Without the option of resolution of the symptoms with conservative measures, exploratory laparotomy was the next therapeutic and complementary diagnostic procedure. Neither

gut malrotation nor congenital accessory peritoneal sac was found. The AC was partially removed, and adhesiolysis of the intestinal loops was performed without bowel resection. It was worth noting that there was a lack of fibrous tissue covering the aorta, vena cava, renal pedicles, ureters and psoas muscles. In spite of the total parenteral nutrition and intensive care facilities, she developed cachexia and recurrent episodes of intestinal obstruction, with several enterocutaneous fistulae, and died as a consequence of peritonitis and sepsis about five months later.

## DISCUSSION

In this case, the characteristic cocoon-like white thickened membrane was found encasing the small intestine. Moreover, the sclerosis of visceral peritoneum, lymphohistiocytic infiltrates, giant cells, and vessel changes were similar to the classical findings described in AC. This is a rare cause of intestinal obstruction in adults, usually found at laparotomy in patients with recurrent small bowel obstruction.<sup>(2,3)</sup> Although sometimes used interchangeably, three main distinct hypotheses must be considered in the present case: (1) peritoneal encapsulation, a very rare congenital condition first described in 1868 by Cleland, which is characterised by the small intestine lying behind a saccular accessory peritoneal membrane; (2) sclerosing encapsulated peritonitis, first described by Owtshinnikow in 1907 and associated with peritoneal dialysis, beta-blocker use, recurrent peritonitis, peritoneal shunting, sarcoidosis and systemic lupus erythematosus, which is characterised by a thick greyish-white fibrous membrane that involves the small intestinal loops; and (3) abdominal cocoon.<sup>(1,2)</sup>

Although the aetiology of AC remains unclear, this entity is characterised by the presence of a white dense peritoneal membrane, which totally or partially encases the small bowel loops. The pathogenesis is not well known, but mesothelial and inflammatory cells, fibroblasts and cytokines play a role in the development of peritoneal fibrosis and neoangiogenesis.<sup>(4)</sup> Interestingly, retrograde menstruation has been postulated as a predisposing factor in AC,<sup>(2,5)</sup> and could have played a role in the pathogenesis of the ovarian endometriosis in our patient.<sup>(6)</sup> In spite of the treatment with goserelin for six months, our initial concern was about the persistence of endometriosis in a middle-aged infertile woman with haemorrhagic ascites. However, this is an uncommon cause of massive haemorrhagic ascites and has been rarely associated with AC.<sup>(5,7)</sup> A major diagnostic concern was primary ovarian tumour or endometriosis-associated ovarian cancer.<sup>(6)</sup> Nevertheless, the histopathological studies ruled out the hypotheses of malignancy.

Conditions associated with peritoneal fibrosis include beta-blocker use, peritoneal dialysis, abdominal surgery, peritoneal malignancy, drug or particulate matter, peritonitis, autoimmune reaction, peritoneal catheter, liver cirrhosis, sarcoidosis, endometriosis, and tuberculosis.<sup>(2,3,5,8)</sup> As the PPD skin test yielded strongly reactive results in a cachetic patient from a developing country, another concern highlighted was the possibility of encapsulating tuberculous peritonitis. In fact, in addition to an eventual association with AC, peritoneal tuberculosis can also mimic advanced ovarian cancer.<sup>(8,9)</sup> However, tuberculosis was excluded, based on the histopathological and bacteriological data. Additionally to the absence of a systemic connective impairment, no indicative signs of coexistent retroperitoneal fibrosis could be found in this patient.

The preoperative diagnosis of AC requires a high index of suspicion, supported by clinical data and imaging findings indicative of the condition. However, most cases are diagnosed at exploratory laparotomy. Clinical presentation includes intestinal obstruction, weight loss and the presence of an abdominal mass. Ultrasonography and CT classically show bowel loops encased within a conspicuous membrane, mural thickening and calcifications. Removal of the cocoon tissue and adhesiolysis have been the effective treatments, but extensive resections

carry high morbidity and mortality rates.<sup>(2,3,5,8)</sup> Associated measures include small bowel intubation, total parenteral nutrition, steroids and cyclophosphamide or azathioprine, with or without colchicine.<sup>(3,4)</sup>

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