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Abdominal cocoon syndrome or sclerosing encapsulating chronic peritonitis, a rare cause of intestinal obstruction in the adult

Síndrome del capullo abdominal o peritonitis crónica esclerosante encapsulada primaria, una causa rara de obstrucción intestinal en el adulto

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Palabras clave: *Obstrucción intestinal, abdomen en capullo, peritonitis crónica esclerosante encapsulada.*

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ABSTRACT

Introduction: Sclerosing encapsulating chronic peritonitis is a rare cause of small intestine obstruction. Primary presentation of abdominal cocoon syndrome is an uncommon malformation in development characterized by the envelopment of the small intestine, compromising totally o partially the full length of the intestine, secondary to a fibrotic tissue covering of the intestinal loops. This membrane is thick and fibrotic, resembling a cocoon; involving the small intestine, in other cases, it can reach the stomach, colon, liver, and spleen. **Case report:** A 35-year-old man presented to the ER with a history of abdominal pain three months before admission. His symptoms had intensified progressively. A surgical approach was decided, first by laparoscopy. The intestinal loops were covered by fibrotic tissue, compromising almost 75% of the small intestine. Abdominal cocoon syndrome is rare. Knowledge of this kind of pathology is important because it mimics common causes of the surgical abdomen.

RESUMEN

Introducción: *La peritonitis crónica esclerosante encapsulada es una causa rara de obstrucción del intestino delgado. La forma primaria o abdomen en capullo es una extraña malformación del desarrollo caracterizada por el revestimiento de la totalidad o parte del intestino delgado por una vaina de espesor del peritoneo visceral. La membrana es gruesa y fibrótica que recuerda un capullo, la cual involucra intestino delgado y en ocasiones alcanza a cubrir estómago, colon, hígado y bazo.* ***Reporte de caso:*** *Masculino de 35 años de edad en el Centro Médico ISSEMyM Toluca, con dolor abdominal de tres meses de evolución, con múltiples visitas al departamento de urgencias y características clínicas inespecíficas; debido al empeoramiento de su sintomatología, se decidió una exploración quirúrgica inicialmente de manera laparoscópica, con evidencia de asas intestinales con tejido fibrótico que envolvía 75% del intestino delgado. El síndrome de abdomen en capullo es raro; sin embargo, el conocimiento de este tipo de patología es de importancia para el cirujano, ya que su presentación puede ser similar a otras causas de abdomen quirúrgico.*

INTRODUCTION

Chronic sclerosing encapsulating peritonitis (CSEP) is a rare cause of small bowel obstruction. First described by Owtschinnikow in 1907, it was initially named chronic fibrosing peritonitis incapsulata. Foo in 1978 named it cocoon abdomen (CA).[1,2] In the medical literature a total of 50 cases have been reported.[3,4]

CSEP is classified according to its etiology into primary and secondary to other diseases, e.g., chronic ambulatory peritoneal dialysis patients, chronic infections, etc. The primary form is a rare developmental malformation characterized by the lining of

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all or part of the small intestine by a thick sheath of accessory peritoneum, which mainly affects young women from tropical or subtropical areas.[5,6] The membrane is thick and fibrotic and resembles a cocoon, which involves the small intestine and sometimes extends to cover the stomach, colon, liver, and spleen.[7-9]

The clinical presentation of primary CSEP or cocoon abdomen syndrome can manifest as intestinal occlusion, either in acute or subacute episodes. Its main characteristic is its recurrent form, associated with abdominal distension, weight loss, nausea, hyporexia. It sometimes manifests as an abdominal mass at the time of physical examination.[10] On occasions, it is asymptomatic, an incidental finding at laparotomy for other causes.[11]

We present the case of a patient with recurrent intestinal obstruction, with cocoon abdomen or primary CSEP. We highlight the diagnostic approach, surgical treatment, and histological confirmation of this rare pathology.

PRESENTATION OF THE CASE

A 35-year-old male, with no previous surgical, allergic, transfusion, traumatic, chronic degenerative, or infectious history. He started suffering three months before hospitalization and assessment, with colicky abdominal pain of insidious and progressive onset, located in the epigastrium and mesogastrium, intermittent, with an intensity of 4/10 reaching up to 8/10 on a visual analog scale (VAS). It started 15 days before hospitalization, with no exacerbating or extenuating factors, with clinical improvement after medical management with antispasmodics at the time of hospitalization. It was accompanied by nausea and vomiting of gastric contents. He reported a weight loss of about 19 kg.

Physical examination revealed a patient with a thin complexion, borderline hydration, pain facies at the time of colic with uncomplicated ambulation. His vitals were heart rate 75 BPM, respiratory rate 16 per minute, blood pressure 110/70 mmHg, oxygen saturation of 94% without supplemental support. The thorax had symmetric amplexion and amplexation, audible bilateral vesicular murmur. The abdomen was soft and depressible, with mild pain on deep palpation in the epigastrium and mesogastrium, and hypoactive peristalsis. On rectal examination a normotonic anal sphincter, with no evidence of tumor, and fecal debris. Limbs were normal, without alterations of sensitivity.

The laboratory reported leukocytes 11.37 × 103/μl, hemoglobin 17.4 g/l, hematocrit 33.8%, glucose 85 mg/dl, urea 35 mg/dl, creatinine 0.9 mg/dl, sodium 145 mmol/l, potassium 4.3 mmol/l. No other alteration in the rest of the reported values. The abdominal X-ray showed enlarged small bowel loops, hydro-aerial levels in the epigastrium and mesogastrium, with no evidence of abdominal air. Intestinal transit with water-soluble medium and oral and intravenous contrast tomography of the abdomen revealed distension of small bowel loops at different levels, gas in the rectal ampulla, passage of contrast medium in front of the mesenteric artery, with an arterial angle greater than 25º, a mesenteric clamp syndrome (Wilkie syndrome) was ruled out. Magnetic resonance was performed in which the four portions of the duodenum were found distended with enlargement of the proximal jejunum. Endoscopy showed an esophagitis grade C of Los Angeles, chronic follicular gastropathy, retentive stomach, distension in the second and third duodenal portions. This led to the diagnosis of partial intestinal obstruction *(Figure 1)*.

A diagnostic laparoscopy was decided due to a lack of resolution and diagnostic certainty of an intestinal occlusion syndrome. A laparoscopic approach was used with a 10 mm umbilical port with Hasson or open technique. Peritoneal mesenchymal fibrotic mesenchymal tissue was found fixing the intestinal loops from the Treitz angle to the ileocecal valve thus limiting its movement. Distension was present forming a second pseudo peritoneal pouch, with serohematic fluid (40 ml). The surface of the pseudo-pouch was not granulomatous and separated from the greater peritoneal cavity *(Figure 2)*. It was decided to convert to open surgery, through a supra infra umbilical midline incision. A

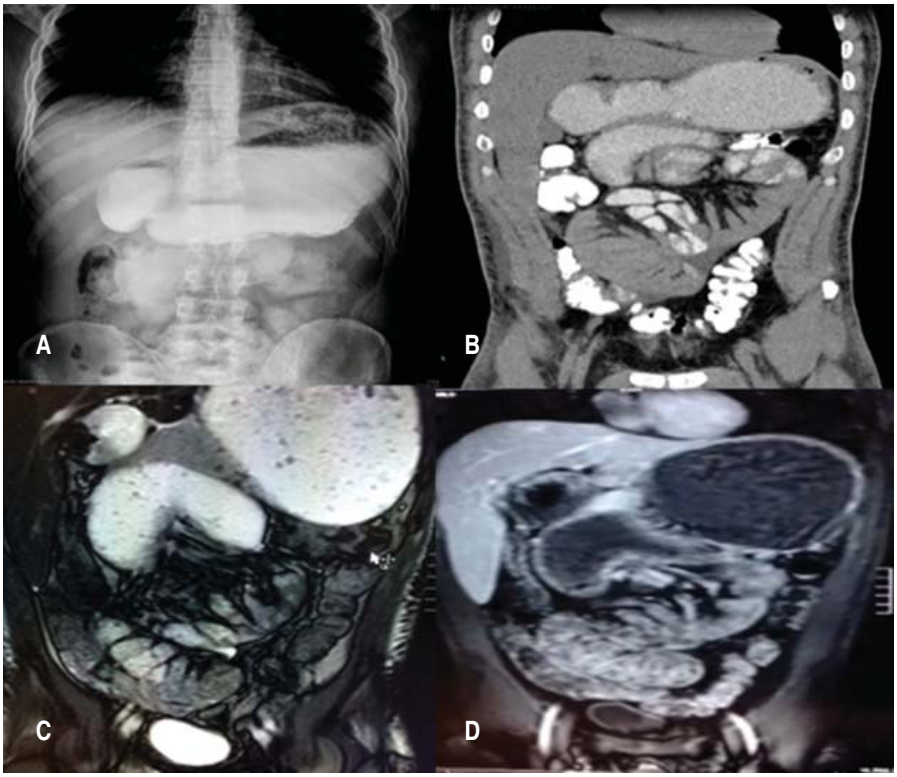

Figure 1:

(A) *Soluble medium bowel transit.* ***(B)*** *Contrast oral and IV tomography of the abdomen, distension of small bowel loops at different levels.* ***(C and D)*** *Magnetic resonance enterography showing the four portions of the duodenum distended and enlargement of the proximal jejunum.*

sample of cavity fluid was taken, then the release of fibrotic tissue which surrounded 75% of the small bowel from the jejunum to the terminal ileum. A separation plane was identified between the mesenchymal tissue and the intestinal visceral peritoneum *(Figure 3)*. There were no ischemic changes in the bowel loops, which had preserved motility, and no enlarged lymph nodes in the mesentery. A Jackson-Pratt drainage was placed into the pelvic cavity and the abdomen closed.

The patient evolved well. He mentioned symptomatic improvement as compared to the preoperative state. Oral feedings started 24 hours after the procedure. Only serohematic fluid came through the drainage during the first days. On radiological control, standing and decubitus abdominal films showed adequate intestinal gas distribution. The patient was discharged on the third postoperative day and sent to the hospital outpatient clinic for follow-up and control. At one year follow-up, he had gained weight and had no clinical evidence of recurrence of obstructive symptoms. The histopathological report was of a serous mesothelial cyst, with moderate chronic inflammatory changes in the omentum and peritoneal fluid *(Figure 4)*.

DISCUSSION

Since its initial description in 1907 as Primary CSEP and the denomination of cocoon abdomen by Foo in 1978, about 50 cases have been reported. Because of its histological characteristics, CSEP has been linked to chronic inflammatory processes of the abdominal cavity, i.e., mesenchymal tissue with an inflammatory reaction, and chronic and acute inflammatory cells. It is not possible to identify a factor associated with CSEP or cocoon abdomen. It may be secondary to various ailments.[12]

The primary stimulus that triggers the inflammatory reaction is unknown. Some authors suggest that it is associated with gynecologic conditions such as subclinical primary viral peritonitis, or secondary to an immunologic reaction after gynecologic infections, and/or due to retrograde

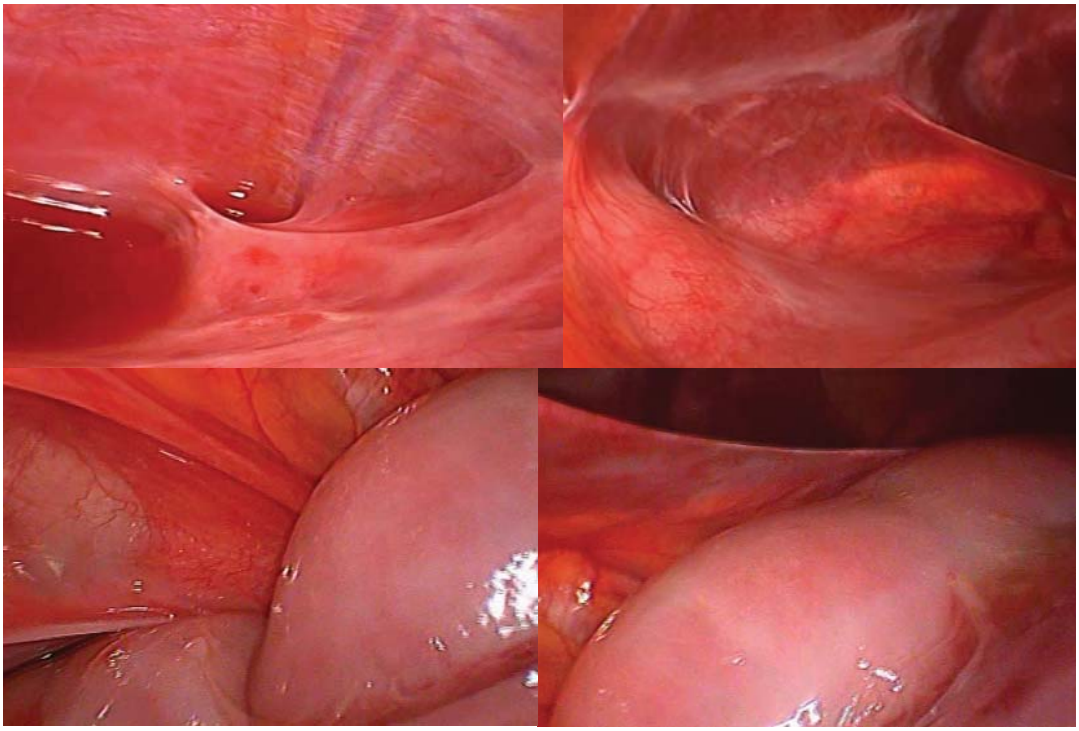

Figure 2: *Laparoscopic view of the abdominal cavity showing intestinal loops covered by a smooth, non-granulomatous surface separating the peritoneal cavity.*

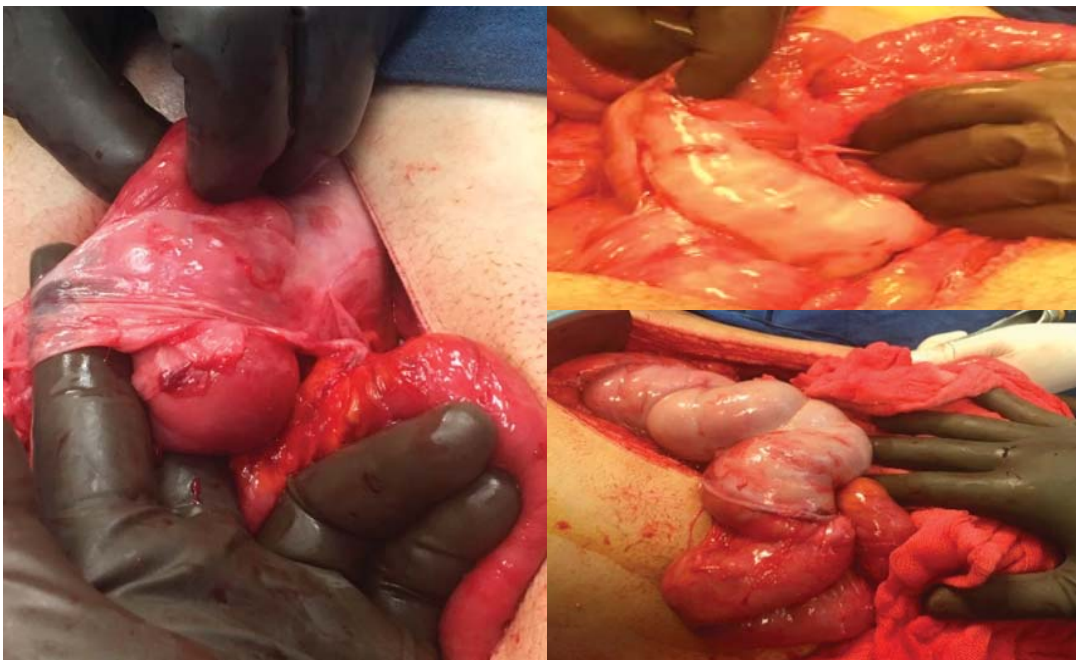

Figure 3: *Intestinal loops with fibrotic tissue enveloping 75% of the small intestine from the jejunum to the terminal ileum.*

menstruation. There is little support for these theories since cocoon abdomen also occurs in premenopausal women, children, and men, as in the case presented here.

Secondary CSEP has been reported in association with beta-blocker intake, chronic ambulatory peritoneal dialysis, sarcoidosis, systemic lupus erythematosus, liver cirrhosis, constrictive pericarditis under propranolol therapy, intraperitoneal instillation of drugs, uterine myomatosis, endometriosis, ovarian tumors, tuberculosis, and recurrent peritonitis.

The clinical presentation of these cases is associated with bowel obstruction. However, it varies from asymptomatic cases to acute (5%) or subacute (2%). Most characteristically cases are recurrent.[13]

Seventy percent of cases of CSEP are diagnosed incidentally during laparotomy, the diagnosis is difficult to reach before surgery because lab tests results are usually normal and imaging findings are often nonspecific. Simple abdominal films may show data compatible with intestinal obstruction, such as hydro-aerial levels in intestinal loops and distension, or abnormal distribution of the air pattern in the gastrointestinal tract. In the case we report, the abdominal CT scan showed a conglomerate of thin loops surrounded by a wall of dense tissue. In any case, the preoperative diagnosis of CA requires a high degree of clinical suspicion.[14]

Confirmation of the diagnosis is made during exploratory laparotomy by taking tissue samples for pathology analysis. The usual finding is a conglomerate of intestinal loops surrounded by a dense membrane, with a transition in the diameter of the loops at the point of obstruction.[15] We did not find a clear site of transition but the release of the peritoneal lining improved the motility of intestinal contents, and the obstructive picture did not recur after surgery.

Surgery is the treatment of choice, which should include dissection of the peritoneal membrane and extensive adhesiolysis. There is generally no need for bowel loop resection unless these show irreversible ischemic changes or perforations of more than 50% of the circumference, which increases morbidity and mortality. Long-term postoperative prognosis with an adequate evolution without complications in 95% of cases.

In rare causes of intestinal obstruction, the diagnosis of CA is made. Patients may report recurrent abdominal pain accompanied by postprandial vomiting, symptoms that can be confused with other causes of intestinal obstruction such as superior mesenteric artery clamp syndrome, pyloric hypertrophy, or a duodenal tumor, which should be ruled out in the study protocol.

Our patient presented with no surgical or chronic degenerative history, nor non-specific digestive symptoms. The endoscopic study

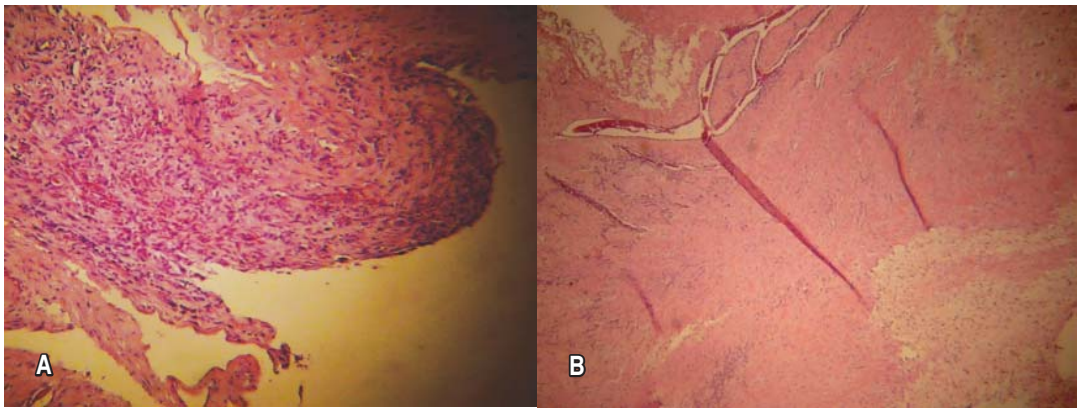

Figure 4: *Histopathological report.* ***(A)*** *Serous mesothelial cyst, omentum, with moderate chronic inflammatory process.* ***(B)*** *Peritoneal fluid, with moderate chronic inflammatory process.*

Table 1: CA cases reported in the literature, diagnostic tool and intraoperative findings.[17,18]

	Author and year	Age in years	Sex	Diagnostic tool	Intraoperative findings
1	Salamone et al 2013	45	M	CT scan and surgery	Small bowel and encapsulated omentum
2	Patel et al 2013	26	M	Surgery	Portion of encapsulated small bowel
3	Yeniay et al 2011	71	F	Surgery	Portion of encapsulated small bowel
4	Ranganathan et al 2003	25	M	Surgery	Portion of encapsulated small bowel
5	Oymacı et al 2013	32	F	Surgery	Portion of encapsulated small bowel
6	Madan Karthik Raj, 2013	30	M	CT scan and surgery	Small bowel entirely encapsulated
7	Sharma et al 2013	42	M	CT scan and surgery	Portion of encapsulated small bowel
8	Gupta et al 2013	40	M	CT scan and surgery	Small bowel entirely encapsulated
9	Narmadha et al 2014	48	F	Surgery	Small bowel entirely encapsulated
10	Çağlar et al 2013	36	F	Surgery	Small bowel entirely encapsulated

suggested obstruction of the second duodenal portion. This the study was completed with intestinal transit films and contrasted CT scans, to rule out other causes of intestinal obstruction. CT scan has a sensitivity and specificity of up to 93%, although in our case it only showed unusual distension of the proximal jejunal loops. Intestinal NMR shows a typical "cauliflower" image.

The definitive diagnosis is usually made during surgical treatment and with the histopathological features of the resected tissue *(Table 1)*.

To rule out infectious processes, tissue collection and intraperitoneal fluid cultures should be included during surgery.[16]

CONCLUSION

The diagnosis of recurrent intestinal obstruction is made by integrating repeated clinical, biochemical, and radiological data. CT scans and intestinal magnetic resonance imaging are useful to rule out other causes of intestinal obstruction. The cocoon abdomen syndrome other rare conditions are usually incidental findings during surgical treatment, confirmed by histopathologic analysis. The cause of this disease remains unknown.

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