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CASE REPORT

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Idiopathic sclerosing encapsulating peritonitis: An uncommon cause of intestinal obstruction in a virgin abdomen

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Abstract

Idiopathic sclerosing encapsulating peritonitis is a clinical entity characterized by partial or complete encasement of the digestive tract by a fibrous membrane. The preoperative diagnosis is difficult to establish. The diagnosis of sclerosing encapsulating peritonitis should be considered for patients without any surgical history and admitted for intestinal obstruction, especially for patients having peritoneal dialysis. We herein report the case of a 50-year-old man with idiopathic encapsulating peritonitis complicated by intestinal obstruction and ischemia. Idiopathic sclerosing encapsulating peritonitis is a rare disease. The diagnosis is made generally during a surgical procedure. Surgery seems to be the best management option for patients with severe signs of intestinal obstruction. Total resection of membrane avoids recurrences but it is associated with higher morbidity and mortality

KEYWORDS

idiopathic sclerosing encapsulating peritonitis, sclerosing peritonitis intestinal obstruction, small bowel obstruction

1 | INTRODUCTION

Idiopathic sclerosing encapsulating peritonitis is a very rare entity. In the literature, less than 200 cases have been reported. It is characterized by the envelopment of the digestive tract by a fibrous membrane. The encasement of the small intestines may cause intestinal obstruction. The pathophysiology has not been clearly understood. The preoperative diagnosis is difficult as symptoms are nonspecific. Radiological images may guide the diagnosis.

We herein report the case of a 50-year-old man with idiopathic encapsulating peritonitis complicated by intestinal obstruction and ischemia.

2 | CASE PRESENTATION

A 50-year-old man with no medical or surgical history. He presented to our hospital with complaints of abdominal pain, vomiting, severe constipation, and abdominal distension for a two-day duration. There was no history of recurrent intestinal obstruction.

On physical examination, the patient was hemodynamically stable. He had no fever, and no scar was found on his abdomen.

Abdominal examination revealed a distended abdomen, with a localized tenderness over the periumbilical region, and a palpable mass localized in the right lower

quadrant. Laboratory tests revealed a biological inflammatory syndrome.

An abdominal CT scan was done and showed two masses. The first mass was localized in the pelvis, formed by adhesions of bowel loops (the distal ileum segment) and encased within a thick membrane. The small bowel loops were not dilated but the CT scan was showing incarcerated loops with decreased bowel wall enhancement. Some peritoneal calcifications were detected (Figure 1).

The second mass was localized in the left flank, formed by adhesions of proximal jejunum, encased within a thick membrane, without signs of small bowel wall ischemia (Figure 2).

The diagnoses of sclerosing encapsulating peritonitis and internal congenital hernia were suspected. There were signs of intestinal ischemia of the first mass, leading to performing an emergency laparotomy.

A median laparotomy was done. The intraoperative findings revealed two capsule-like masses formed by a whitish, transparent membrane encompassing the small bowel with fibrous adhesion. There were no signs of generalized peritonitis.

The first capsule-like mass was voluminous, measuring 18 cm, a gentle dissection was performed. The membrane was excised, and adhesions between bowel loops were divided. The underlying bowel loops were devitalized (Figure 3). We performed a resection of the 10 cm necrotic bowel loops with intestinal anastomosis.

The second capsule-like mass was the result of adhesion of proximal jejunum encompassing by fibrous membrane. We could not resect all the membrane. (Figure 4) We performed a partial resection and lysis of adhesions. The bowel loops were healthy. We did not traumatize the digestive tract.

The postoperative Day 3, we diagnosed an enterocutaneous fistula. The patient was re-explored. We found localized peritonitis due to an anastomotic leak. We performed an ileocecal resection and both ileal and colonic stoma were performed.

The histopathological examination concluded to peritoneal fibrosis associated with a non-specific inflammation without any signs of malignancy. The diagnosis retained was idiopathic encapsulating and sclerosing peritonitis.

3 | DISCUSSION

Sclerosing encapsulating peritonitis is defined as a chronic fibro-inflammatory disease of the peritoneum. It is described as a thick fibrous membrane encasing the digestive tract. It was reported for the first time nearly one century ago,[1] several terms were used to describe this entity. In 1868, Cleland reported the first case, initially termed encapsulating peritonitis.[2] In 1921, Winnie termed it "zuckergussdarm", describing the whitish membrane enveloping the digestive tract.[3] In 1978, Foo coined the expression of "abdominal cocoon."[4]

Sclerosing encapsulating peritonitis is classified as primary form or secondary form based on pathophysiological characteristics of the encasing membrane.[1] Theories suggest that congenital and embryological abnormality was the principal cause of the primary form.[5] However, primary sclerosing encapsulating peritonitis is the idiopathic form, some hypotheses have related this form to retrograde menstruation and viral infections.[6,7]

The secondary form of sclerosing encapsulating peritonitis is the result of an abnormal chronic inflammation of the peritoneum leading to the formation of the membrane covered with collagen and fibrinogen.

Multiple potential causes of secondary sclerosing encapsulating peritonitis were reported in the literature, such as ventriculoperitoneal derivations, system diseases (sarcoidosis, lupus), Mediterranean fever, tuberculosis, and especially peritoneal dialysis.[1,4,6] According to the Japanese national registry, the incidence of this pathology is about 2.5% after 10 years of peritoneal dialysis. The inflammation in the peritoneal dialysis is due to the hypertonic solutions or infections.[8,9]

This disease may affect both sexes at any age.[10] The mode of establishing the diagnosis is very variable. It is based on anamnestic, clinical, and radiological arguments.

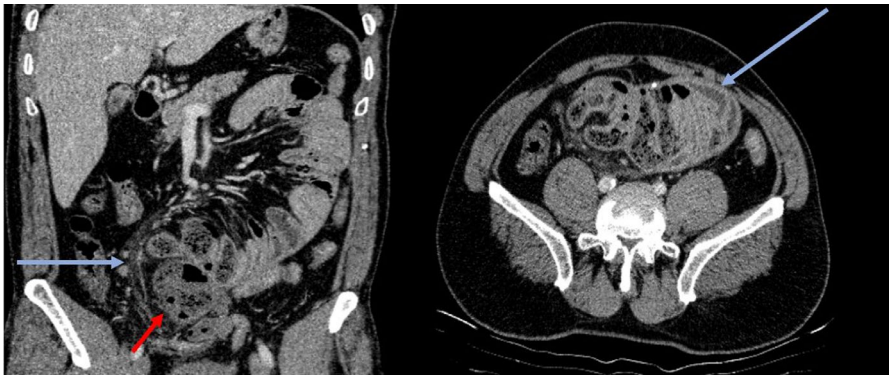

FIGURE 1 Axial and coronal view of the abdominal computed tomography scan showing the first mass formed by the adhesions of bowel loops (red arrow) encased within a membrane (blue arrow)

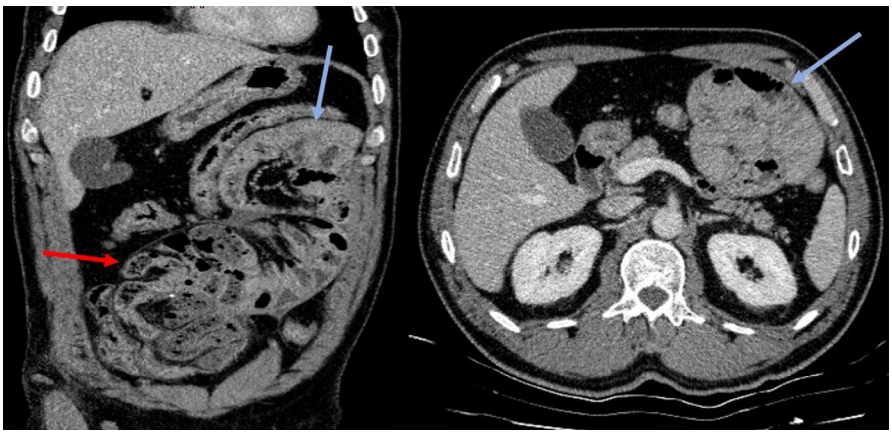

FIGURE 2 Axial and coronal view of the abdominal computed tomography scan showing the first mass of the pelvis (red arrow) and the second mass of the left flank (blue arrow)

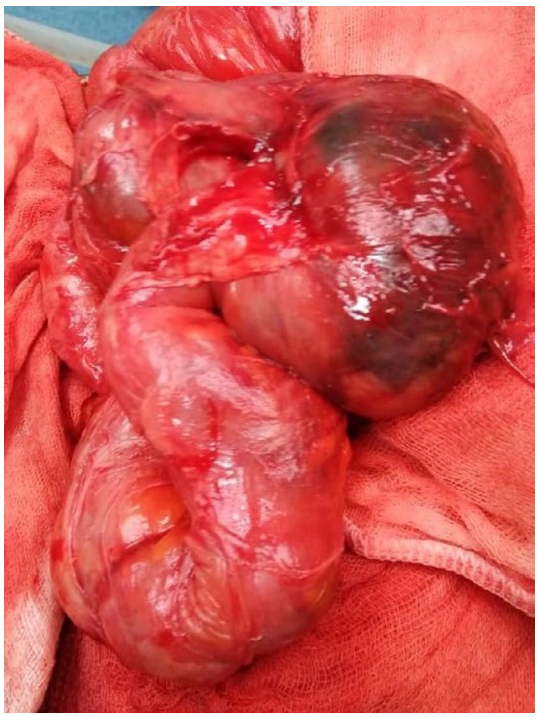

FIGURE 3 Intraoperative findings: the first capsule-like mass with a necrotic small bowel

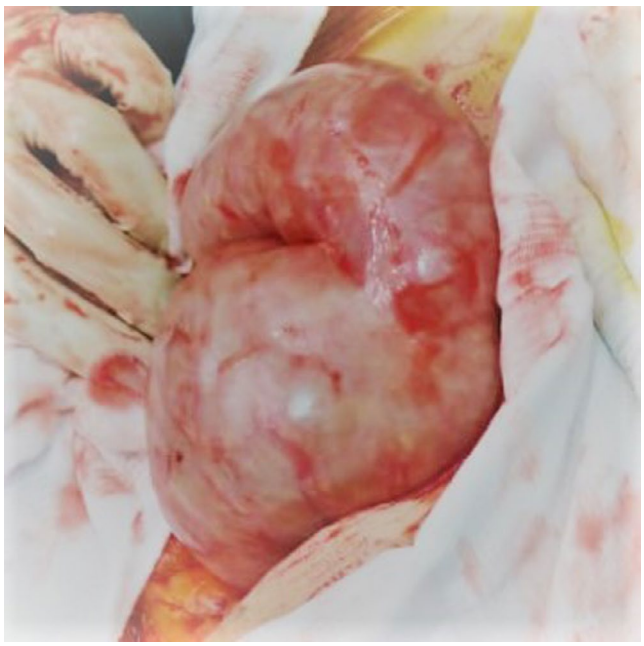

FIGURE 4 Intraoperative findings: the second capsule-like mass

About 47 % of patients were diagnosed perioperatively. In addition, few cases have been discovered at autopsy.[1,11]

Less than 200 cases have been reported in the literature, and some of these cases were associated with non-specific congenital anomalies such as situs inversus.[12] This rarity is confirmed by the largest review of Akbulut et al, which contains only 190 cases from 74 articles published in 23 countries.[5]

The origin of this pathology continues to be controversial and conjectural. The clinical presentation is non-specific. The most common symptom was abdominal distension (85%), followed by abdominal pain (72%). Only 6% of patients reported recurrent intestinal obstruction. Sometimes, an abdominal mass can be found on palpation[1]

The classification of Nakamoto has been published to have a correlation between clinical presentation and histopathological anomalies.

The first stage usually is the asymptomatic stage. It is considered the debut of the inflammatory process. At the second stage, there is the progression of adhesion and encapsulation. It is the encapsulating process. At this stage, usually the patient complains about recurrent non-specific chronic abdominal pain and this stage may last several years. The third stage is the stage of bowel obstruction, which is the main complication of this pathology.[13]

The diagnosis of sclerosing encapsulating peritonitis should be considered for patients without any surgical history and admitted for intestinal obstruction, especially for patients having peritoneal dialysis.

The abdominal CT scan has an important contribution to making the diagnosis. It shows the digestive tract encased within a thick fibro-collagenous membrane, as it was in a cocoon. It may show the signs of small intestinal obstruction, peritoneal, or mural calcification. However, it may be normal in the early stages.[11,14]

There is no consensus on the treatment strategy. Conservative or non-surgical management is indicated for patients with moderate symptoms or multi-operated patients. It is treated like an intestinal obstruction with nasogastric tube decompression.

Several drugs have been used to inhibit fibroblastic production by inhibiting collagen syntheses, such as tamoxifen, colchicine, steroids, and azathioprine. However, their effectiveness requires further investigation.[1,11]

It seems that surgical management is the best treatment. It consists of total or subtotal excision of the encapsulating membrane with adhesiolysis of the small intestine. Resection-anastomosis is indicated for non-viable digestive segments. Manipulation of the bowel should be done very carefully and gently; the dissection and excision of the membrane should be preferably done by sharp dissection using scissors avoiding usage of electrocautery to avoid iatrogenic bowel injury. Complete excision of the membrane decreases the recurrence rate of intestinal obstruction. However, total excision increases morbidity and mortality.[15]

Some selective case has been operated on with the laparoscopic approach.

The postoperative morbidity rate is about 9%.[1] The postoperative complications are intra-abdominal abscess, entero-cutaneous fistula, or recurrent intestinal obstruction.

The postoperative mortality rate is 40% and occurs within a few weeks of the intervention. However, Kawanishi et al, reported a good outcome following surgical procedure with a mortality rate of 4%.[9,11]

4 | CONCLUSION

Sclerosing and encapsulating peritonitis is a rare etiology of intestinal obstruction. Clinical presentations are not specific but radiological images may evoke this diagnosis before surgery. The diagnosis is made generally during surgical procedures. There is no consensus for the treatment strategy but surgery retains an important place. The resection of membrane and adhesiolysis are the principal surgical strategy to avoid ischemia, perforation, and recurrences.

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CONFLICTS OF INTEREST

None.

AUTHOR CONTRIBUTION

Asma Sghair conceived the idea for the document and contributed to the writing and editing of the manuscript. Mehdi Debaibi contributed to the writing and editing of the manuscript. Majdi Kchaou contributed to the acquisition and conception of the manuscript. Skander Talbi and Azza Sridi reviewed and edited the manuscript. Adnen Chouchen contributed to the literature review, manuscript writing, editing, and review of the manuscript. All authors read and approved the final manuscript.

ETHICAL APPROVAL

Personal data have been respected. Published with the consent of the patient.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

The personal data of the patient were respected. No data are available for this submission.

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