

## Encapsulating peritoneal sclerosis: do not be too late for the right diagnosis! Case report and short literature review

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**SUMMARY: Encapsulating peritoneal sclerosis: do not be too late for the right diagnosis! Case report and short literature review.**

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*Encapsulating peritoneal sclerosis (EPS) is a rare clinical syndrome characterized by an acquired, inflammatory fibrocollagenous membrane encasing the small intestine, resulting in symptoms of bowel obstruction. It is still unclear whether early surgical intervention has*

*an advantage over conservative management, but, in most reviewed case reports, it is preferred to preserve the surgical management in patients not responding to conservative measures, or when bowel ischaemia is occurring.*

*We report a case of a 58-year old patients, affected by chronic renal failure, on treatment with peritoneal dialysis, in which a late diagnosis of encapsulating peritoneal sclerosis was made, and where surgical intervention was not sufficient to guarantee survival due to the late diagnosis.*

KEY WORDS: Abdominal cocoon - Encapsulating peritoneal sclerosis - Sclerosing encapsulating peritonitis.

### Introduction

Encapsulating peritoneal sclerosis (EPS), also described as “abdominal cocoon” and “sclerosing encapsulating peritonitis” is a rare clinical syndrome characterized by an acquired, inflammatory fibrocollagenous membrane encasing the small intestine, resulting in symptoms of bowel obstruction (1). Given its rarity and heterogeneity of etiologies, the incidence and prevalence of EPS as a whole is unknown. In a review of idiopathic EPS, cases were more commonly reported from tropical and subtropical countries [China (54%), India (18%), Turkey (9%), and Nigeria (3%)] (2). The mean age was 34.7 (range 7-87 years) with a 2:1 male predominance (1). Most of

patients would resolve with only conservative medical treatments, but some may require surgical intervention. It is still unclear whether early surgical intervention has an advantage over conservative management, but, in most reviewed case reports, it is preferred to preserve the surgical management in patients not responding to conservative measures, or when bowel ischaemia is occurring.

### Case report

A 58-year old patient was admitted, in October 2018, to our surgical department with a longstanding history of weight loss, diffuse abdominal pain and distention, and absolute constipation. He was affected by the birth by polycystic kidney disease, evolving in end-stage chronic renal failure. Due to this reason in 2016 he had undergone renal transplantation, however with subsequent graft rejection, therefore he started peritoneal dialysis. A computed tomography (CT) of the abdomen and pelvis with and without intravenous contrast injection was per-

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formed. It showed an abundant fluid density collection, of probable ascitic nature, with edematous and swollen aspect of the mesentery, and fluid distension (with associated hydroaire levels) of loops of the small intestine, conglobed in mid-abdominal site, partly floating, with small parietal enhancement; the conglobate aspect of these loops did not allow a secure image referable to a mechanical stop (Figure 1 A, B).

The clinical presentation and imaging were suggestive of severe small bowel obstruction. After several days of unsuccessful conservative management with a nasogastric tube and intravenous hydration, she was taken to the operating room where an exploratory laparotomy was done. Laparotomy revealed, together with an abundant ascitic collection, a hick adherent fibrous sheath that was encapsulating most of the small bowel, making not possible every attempt of viscerolysis; therefore, a lateral ileostomy was performed (Figure 2).

The postoperative period was complicated by multi-organ failure due to sepsis secondary to *P. aeruginosa*, and unfortunately he died on six day after the surgical intervention.

## Discussion

EPS can be divided into primary (idiopathic) or secondary in which a trigger for the inflammatory process can be identified (1). The primary EPS usually manifests itself in young adolescent girls in trop-

ical and subtropical countries such as China, India, Turkey, Nigeria, Malaysia, Singapore, Pakistan, Kenya, and Saudi Arabia, although cases in temperate zones also have been reported (3, 4). Although the real cause is still fully unknown, the geographical distribution of this disease lead to theories of retrograde menstruation or gynecologic infection as the cause (1).

In the secondary EPS, a local or systemic factor can be identified as triggering peritoneal inflammation. Implicated triggers include medications (5-8), infection (9-15), mechanical or chemical intraperitoneal irritants (16-26), cirrhosis (27), organ transplantation (28-30) endometriosis (31), gynecologic neoplasms (32, 33), dermoid cyst rupture (34), and systemic rheumatologic and inflammatory disorders (35-37).

Preoperative diagnosis requires a high index of clinical suspicion (38), considering that the early clinical features of EPS are nonspecific and are often not recognized (38). Clinically, it presents with recurrent abdominal pain, nausea, vomiting, anorexia, weight loss, malnutrition, recurrent episodes of acute, subacute or chronic small bowel incomplete or complete obstruction, and at times with a palpable soft non tender abdominal mass, but some patients may be asymptomatic. In some cases, abdominal distension was secondary to ascites (38). A high index of clinical suspicion may be generated by the recurrent attacks of non-strangulating obstruction in the same individual combined with relevant im-

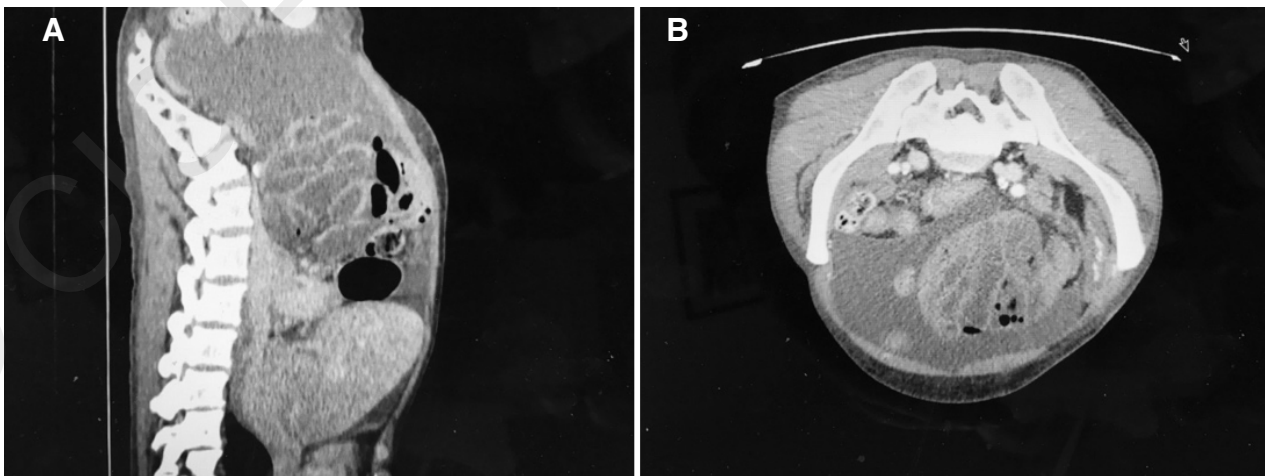


Figure 1 A, B - CT scan showing an abundant fluid density collection, of probable ascitic nature, with edematous and swollen aspect of the mesentery, and fluid distension (with associated hydroaire levels) of loops of the small intestine, conglobed in mid-abdominal site, partly floating, with small parietal enhancement.



**Figure 2 - Laparotomy revealed a hick adherent fibrous sheath that was encapsulating most of the small bowel, making not possible every attempt of viscerolysis.**

aging findings and lack of other etiology (38).

Less than 1% of PD patients develop overt EPS as manifested by combinations of weight loss, ultrafiltration failure, and intestinal obstruction (39-41), like in the case here reported, in which the secondary form of EPS, due to peritoneal dialysis, was the final diagnosis.

With regard to the medical treatments of EPS, the treatment of the underlying cause, the nutritional support, and the use of immunosuppressive (corticosteroids, colchicine, azathioprine, cyclosporine, mycophenolate mofetil (MMF), and mammalian target of rapamycin (mTOR) inhibitors) (42-47) or antifibrotic agents (Tamoxifen) (48, 49) have been employed with alternating results.

With regard to the surgical treatment of EPS, dissection (open or laparoscopic) of fibrotic membrane and adhesiolysis is usually sufficient to treat abdominal cocoon (50, 51). Complications after surgery were reported; these include intra-abdominal infections (like in our patient), enterocutaneous fistula and perforated bowel (52).

Danford et al. recommend that treatment strategies should be tailored to each patient, depending on the extent and stage of EPS [according to the classification of Nakayama (53) and Nakamoto (54)]. In all stages, especially during the early stages, the underlying cause should be identified and treated or removed. During the earlier stages (pre-EPS and inflammatory stages), the pathophysiology tends to be more inflammatory and the degree of sclerosis tends to be minimal (1). Thus, after infection has been ruled out, corticosteroids may be of benefit (1). In the later stages (encapsulating and ileus stages), more advanced sclerosis may be present, and patients may start exhibiting signs and symptoms of partial or complete bowel obstruction (1). In treating abdominal pain, opioids should be avoided. However, this may not always be possible and thus opiate antagonists are recommended in this setting (1). Tamoxifen plays an increasing role in the later stages (1). If poor oral intake or malnutrition is present, total parenteral nutrition may be required (1). If symptoms are severe and there has been no response to medical therapy, surgical intervention may be considered.

In conclusion, as previously highlighted, a high index of clinical suspicion of EPS may be generated by the recurrent attacks of non-strangulating obstruction in the same individual combined with relevant imaging findings and lack of other etiologies (38). The preoperative diagnosis of this entity may be helpful for proper treatment of these patients (38). Most cases are diagnosed incidentally at laparotomy, although a preoperative diagnosis is purported feasible by CT scan (small bowel loops congregated to the center of the abdomen encased by a soft-tissue density mantle) (38, 55). However, a better awareness of this entity and the imaging techniques may facilitate preoperatively diagnosis, thus avoiding a late recourse to the surgical approach that in “difficult” patients may be insufficient, as in the case here reported.

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*Conflict of interests*

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