

## Report of a Documented Case of Encapsulating Peritoneal Sclerosis in Iran

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### ABSTRACT

Encapsulating peritoneal sclerosis (EPS) is a rare, acquired condition that has been described interchangeably with peritoneal encapsulation (PE) as well as abdominal cocoon. EPS causes complications for peritoneal dialysis (PD) patients. In EPS, the small intestine is covered by a fibrotic and inflammatory peritoneal capsule that either partially or completely encases the bowel. For EPS, there is no agreement in the literature as to whether the treatment of choice should be surgery or conservative therapy. However in the obstructive phase the treatment of choice is surgery due to the impossibility of conservative treatment.

Here, we report the case of a 64-year-old woman who presented with several episodes of intestinal obstruction and peritonitis. Computed tomography (CT) of the abdomen did not show significant characteristic features of EPS. Exploratory laparotomy was performed, freeing the small bowel of adhesions and encapsulation.

For EPS, adequate clinical knowledge and a high degree of suspicion are crucial for a correct diagnosis and appropriate management.

**Keywords:** Encapsulating peritoneal sclerosis (EPS); Peritoneal dialysis; Intestinal obstruction; Peritonitis

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#### INTRODUCTION

Encapsulating peritoneal sclerosis (EPS) is a rare, but serious condition that has been described interchangeably with peritoneal encapsulation (PE) as well as abdominal cocoon(1). EPS that complicates peritoneal dialysis (PD) is a disease of the peritoneum. It is characterized by the presence of a fibrotic and inflammatory peritoneal capsule that either partially or completely encases the bowel,(2) resulting in fibrosis and adhesions of the peritoneum to loops of the small intestine leading to intestinal obstruction(3,4).

EPS was first described in 1868 as a pathologic report(1). In 1980, it was reported as one of the possible

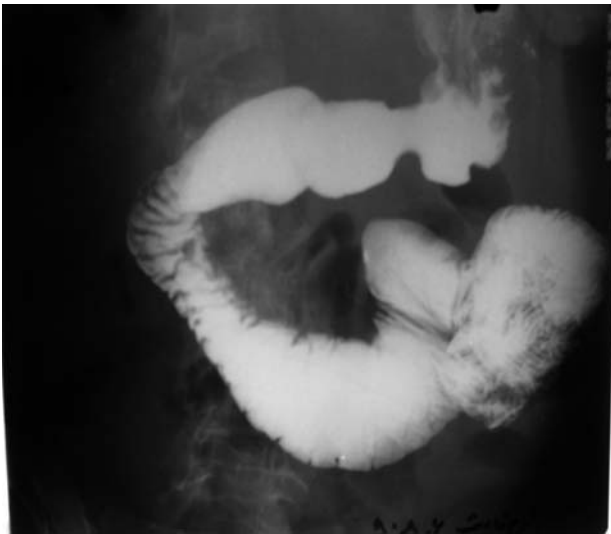


Figure 2: Small bowel transit study with normal results.



Figure 3: All visceral organs were covered by a thick, firm layer.

PD patients, where the small intestine is covered by a fibrotic and inflammatory peritoneal capsule that either partially or completely encases the bowel. There is no agreement in the literature whether the treatment of choice is surgery or conservative therapy. However in the obstructive phase, all are agree with surgery due to conservative treatment is impossible.

## DISCUSSION

EPS, also known as “abdominal cocoon” and PE, is a rare condition(1,4). For the first time, Owtschinnikow(1)

described EPS in 1907 by the name of “peritonitis chronic fibrosa incapsulata”. It was characterized by a thick grayish-white fibrous membrane that covered the small bowel wall. Abdominal cocoon was first described by Foo et al. in 1978,(2) as causing partial or complete mechanical obstruction of the small intestinal loop(1). PE was first described in 1868 by Cleland(10) through which the small bowel lies behind an accessory peritoneal membrane. PE is a congenital occurrence, in contrast to EPS which is known to have an acquired pathology(11). This entity has a various rate of occurrence, between 0.7% to 7.3%(4,8). The highest frequencies are observed in Japanese reports, while a very low prevalence is seen in the United States, Canada, and Europe(12). Its annual incidence in chronic ambulatory CAPD patients is 0.37 per 1000 patient-years with a male to female ratio of 5:2(6).

This condition has been attributed to chronic ambulatory PD, mainly its duration, acetate in the dialysate, and antiseptics used during bag exchanges. The intake of the beta-blocker, practolol, and recurrent peritonitis are additional causative factors(1). Abdominal surgery, ventricle peritoneal shunts, retrograde menstruation, tuberculosis peritonitis, sarcoidosis, systemic lupus erythematosus, carcinoid tumors or patients with liver cirrhosis(13) are other rare causes, all of which were absent in our patient(14,15).

Timely diagnosis and treatment of EPS is of importance as it may offer the chance for resolving bowel obstruction at an early stage, prior to complete encapsulation(8).Morbidity and mortality of this condition is extremely high with a rate close to 60%, and is associated with various surgical complications such as intestinal necrosis and frequent anastomosis breakdowns(1).

Unfortunately, there is much uncertainty, lack of specificity, and delay in the diagnosis of EPS. Clinically, presentation of EPS is non-specific, as in our patient, who presented with abdominal pain, distension, nausea, and vomiting. Its features are acute, subacute or chronic intestinal obstruction, colicky abdominal pain, and weight loss. In 54% of patients an abdominal mass may be palpable(4,6). Patients often have reduced peritoneal clearance. A definitive diagnosis of EPS is performed at laparotomy or laparoscopy and biopsy, however, clinical and radiological features may strongly support the diagnosis(4,6,8,12).

EPS patients may not show characteristic radiological features. Normal to non-specific features of intestinal obstruction are usually seen. Findings on CT, the technique of choice for diagnosis, sometimes show

thickened adhered bowel loops, loculated ascites, and peritoneal enhancement(1,6). The characteristic pattern in most cases is a dilated small bowel in the middle of the abdomen encased by a thick membrane sac. Other radiologic findings include fixation of the intestinal loops, fluid collection, ascites, thickened bowel walls, peritoneal or mural calcification, adenopathy, and peritoneal enhancement. These findings are usually associated with advanced stages of the disease. These are less effective in demonstrating the changes in the early stages of the disease in the peritoneum and may be suggestive of PE when the small bowel is enveloped in a thin membrane, as seen with our patient(4). While many of the techniques mentioned above are helpful in the diagnosis of established cases, there are no investigations that have examined these tools to be predictive. Early radiological screening for detection of EPS in high risk patients requires further study.

There is no agreement in the literature as to whether the treatment of choice should be surgery or conservative therapy. Initial management would be the cessation of PD treatment,(16) transfer to hemodialysis,(12) removal of the dialysis catheter, initiation of total parenteral nutrition (TPN),(16) and sustained rest for the bowel and peritoneum. Medical treatment with immunosuppressive, corticosteroids, and tamoxifen has been shown to be promising(7). Surgical treatment (including removal of adhesive

lesions and complete release of the small bowel when it involves non-viable resection of the loop) should be proposed if the patient does not improve with conservative or medical therapies, and whenever irreversible bowel obstruction is established. Some post-operative complications are recurrent intestinal obstruction, fistula formation, and anastomotic leakage or sepsis due to a perforated intestinal wall(17) as seen with our case(1,4,6,9,12). Our patient underwent laparotomy, with simple freeing of the adhesions as well as release of the trapped intestines. There was no resection of bowel required. She presented with suggestive signs as well as characteristic radiological features, which enabled decisive management.

The best clinical knowledge (including risk factors) of EPS along with a high degree of suspicion is crucial for a correct preoperative diagnosis and appropriate management of intestinal obstruction. EPS is a lethal, severe complication of PD with an increasing incidence that is affected by the duration of dialysis. So, although rare, clinicians should suspect this condition in patients with recurrent intestinal obstruction if no etiological factors can be found. Given the current published data and our experience with an EPS case, there is a rationale for a multi-disciplinary approach to these patients, necessitating the involvement of a nephrologist, dietician, and surgeon.

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