# A Rare Presentation of Encapsulating Peritoneal Sclerosis

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## BACKGROUND

Encapsulating peritoneal sclerosis is a rare condition in which the intra abdominal contents are encapsulated by a fibrotic material. This condition was first described by Owtschinnikow (1). The incidence of EPS is reported to vary between 0.3 to 3.3% (2). The most common risk factor for developing EPS is reported to be peritoneal dialysis. The diagnosis is usually made clincially. Many patients presenting with symptoms or exam findings such as abdominal pain, nausea vomiting, anorexia, abdominal fullness, bowel obstruction or abdominal mass. CT or ultrasound may aid in diagnosis. The diagnosis is confrimed by lapartomy or laparoscopy (3). EPS has traditionally been treated surgically by extensive lysis of adhesions. Recently, there has been a shift toward conservative management, generally with cessation of factors that could cause EPS. This can only be done in patients without signs of ischemia. THe prognosis of EPS is grave. The average mortatlity rate is about 35%.

## CASE DESCRIPTION

A 53 year old female with past medical history significant for hepatitis C, cirrhosis, end stage renal disease on hemodialysis presented to the hospital after recent admission for spontaneous bacterial peritonitis with complaint of abdominal pain, nausea and vomiting for several weeks since her last discharge. On exam the patient was diffusely tender to palpation and positive for a fluid wave. A CT scan was obtained showing a significant amount of pneumoperitoneum. At this time surgical intervention was offered to the patient and after a discussion with family, the patient decided to proceed. The patient was taken for an exploratory laparotomy. Upon entering the abdomen, thick murky fluid was evacuated, along with ascetic fluid. After further inspection, a yellow gelantenous material was noted to be mixed in with ascetic fluid, most likely secondary to chronic infection from spontaneous bacterial peritonitis. After clearing all the fluid, no abdominal contents were visualed, but a thick fibrous cocoon like structure was seen, consistent with Encapsulating Peritoneal Sclerosis. The cocoon was then entered and bowel was found to be denslely adhered to the fibrous tissue. An extensive lysis of adehesions was done to free the small bowel. Once peristalsis was noted the patient was irrigated and closed. On post operative day one the patient was noted to have frank blood from JP drains and require pressor support,the patient was taken back to the operating room for a reexploration, but no area of significant bleeding was found. The patient died of disseminated intravascular coagulation later that day.

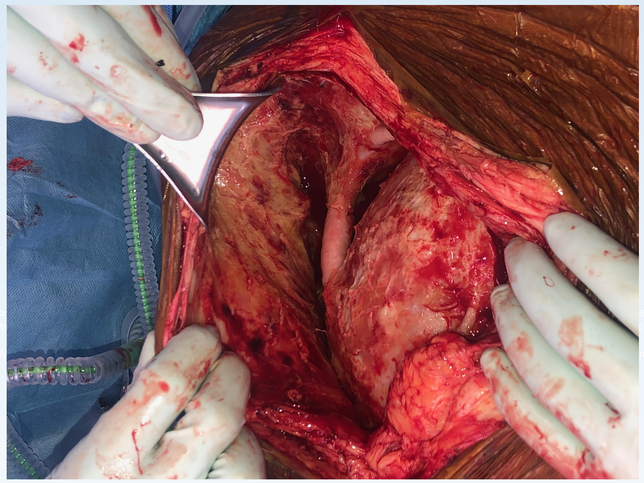

FIgure 1: Fibrionous caccoon found upon entering the abdomen

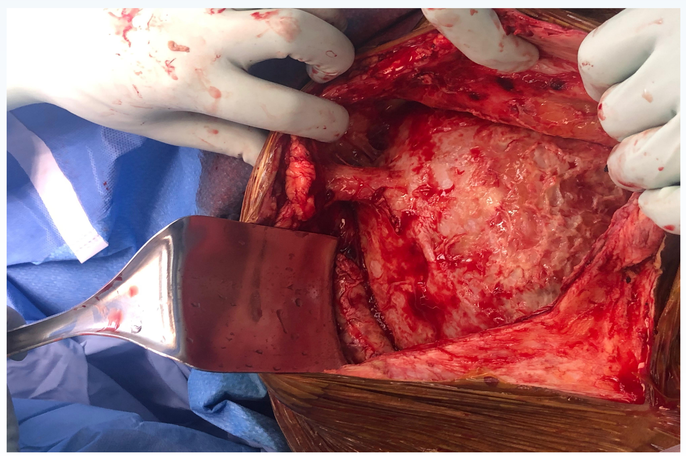

Figure 2: Fibrinous cacoon found upon entering the abdomen, second view

## DISCUSSION

Encapsulating Peritoneal Sclerosis was first descrbed in 1907. It has beome a growing concern, especially for those undergoing peritoneal dialysis. Our patient had no history of peritoneal dialysis, making this case urnique. There are primary causes which are idopathic and secondary which can further be divided into associated with peritoneal dialysis or not (4). The exact mechanism of how peritoneal dialysis causes EPS is unknown but it is thought to be due to duration of dialysis, recurrent episodes of bacterial peritonitis, use of glucose based dialysate or hypertonic solution, use of acetate dialysis solution, plasticizers, chlorhexidine and endotoxins from bacterial fillers (4). There are rare reports of patients with idiopathic EPS. These are usually younger patients. Many initially were prepubescent females. The incidence varies from 0.3 to 35%. (2). The mortality reate ranges from 17 to 55% (3). Most patients in dcomented cases range from ages twenty to fourty, which this patient was in her fifties. Many patients present with GI symptoms (3). The developement of these symptoms is usually insideous, as with our patient. Many cases of EPS do have a history of peritonitis (5). Imaging may be of little use for diagnosis in EPS. CT is the best modality to diagnose EPS. The most specific findings on CT including teh bowel at the center of the abdomen and peritoneal calcifications, how these are only reported in 35% of CT scans in those with EPS (3). The patietns CT scan had peritoneal thickening, but no peritoneal calcifications, a CT scan done one month prior had no report of peritoneal thickening. The gold standard of diagnosis is surgery, which is also theraputic. Some patients are placed on bowel rest and given TPN, however with the pneumoperitoneum on CT the patient had clear indiaction for the operating room. Many patients require re operation after the first surgery..

## CONCLUSION

EPS is a grave disease. The development is almost always associated with peritoneal dialysis. This patient had no history of peritoneal dialysis. She most likely developed EPS from multiple paracentesis and spontaneous bacterial peritonitis. Due to the concern for CT findings of pneumoperitonium and abdominal exam consititant with possible bowel perforation the patient was taken for a laparotomy. This would have been the definitive treatment of her EPS had it been diagnosed beforehand. Her prognosis may have been better had she been optimized with TPN and bowel rest. In these patients early diagnosis and treatment is key.

## REFERENCES

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