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Case Report

Encapsulating peritoneal sclerosis: A case report of a diagnostic dilemma

Nimeshana Bandara[1,2], Kamal Jayasuriya[2]

[1]Postgraduate institute of Medicine, University of Colombo, [2]District General Hospital, Kegalle

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Corresponding Author: Nimeshana Bandara, E-mail< nimeshanab@gmail.com >

https://orcid.org/0000-0003-1534-7923

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Introduction

One of the long-term complications of peritoneal dialysis (PD) is encapsulating peritoneal sclerosis (EPS). It is associated with high morbidity and mortality. Understanding regarding EPS is still evolving since its first description by Owtschinnikow in 1907.

The incidence of EPS in patients undergoing PD varies worldwide, ranging from 0.5%-7.3%, increasing with the duration of PD. Clinical presentation is non-specific. Imaging modalities aid the diagnosis. Majority are diagnosed during laparotomy. Surgery is indicated in severe stages of EPS. Despite advanced diagnosis modalities and the development of treatment strategies, EPS is associated with a high mortality of around 32%-35%.

We report a patient with a history of PD, presenting with features of intestinal obstruction with EPS diagnosed following laparotomy. This is the only reported case in Sri Lanka.

Case report

A 66-year-old male with a history of end-stage kidney disease on regular haemodialysis two times a week presented with vomiting and loose stools for one day associated with abdominal pain. He had had a similar event in the past which did not require hospital care. The current episode was associated with more severe symptoms. He had type 2 diabetes mellitus and hypertension. He had been on PD and changed to haemodialysis 2 years previously.

On examination, his haemodynamic parameters were stable and he was found to have a large abdominal mass. X-ray abdomen showed multiple fluid levels in the small intestine and the small intestine was gathered centrally (Figure 1).

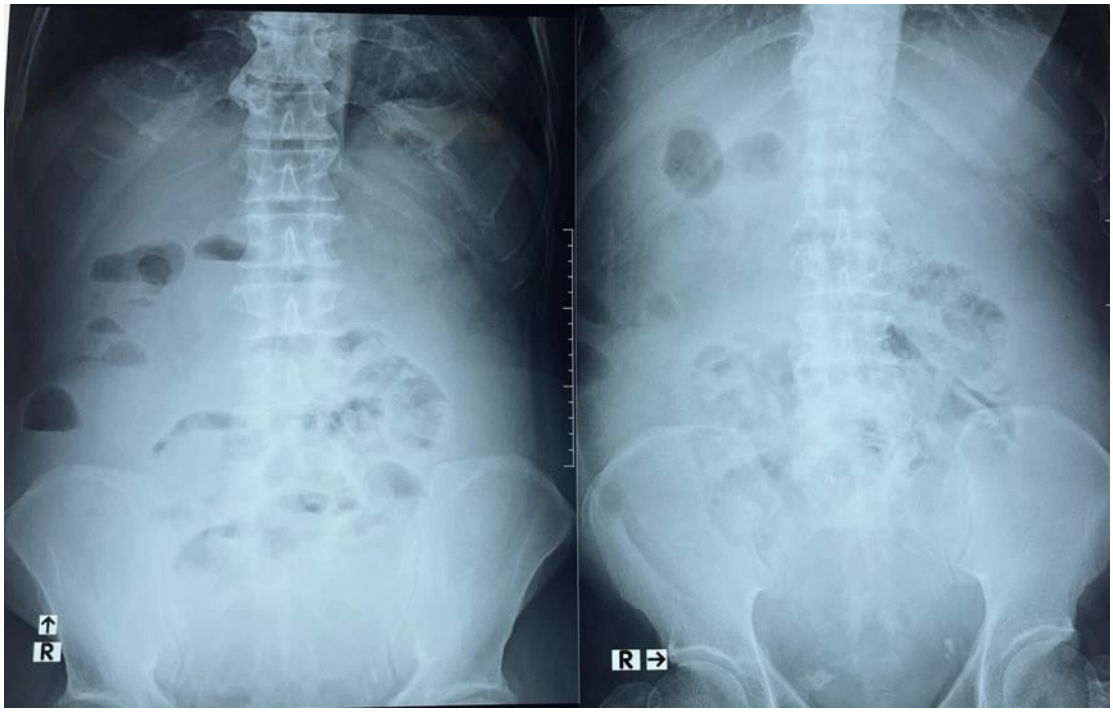

Figure 1: X-ray abdomen supine(right) and erect(left) of the patient

Ultrasound abdomen showed a large fluid-containing sac in the central abdomen and small intestine loops incorporated within the sac. Uncertainty of diagnosis leads to urgent CT-abdomen and pelvis (Figure 2).

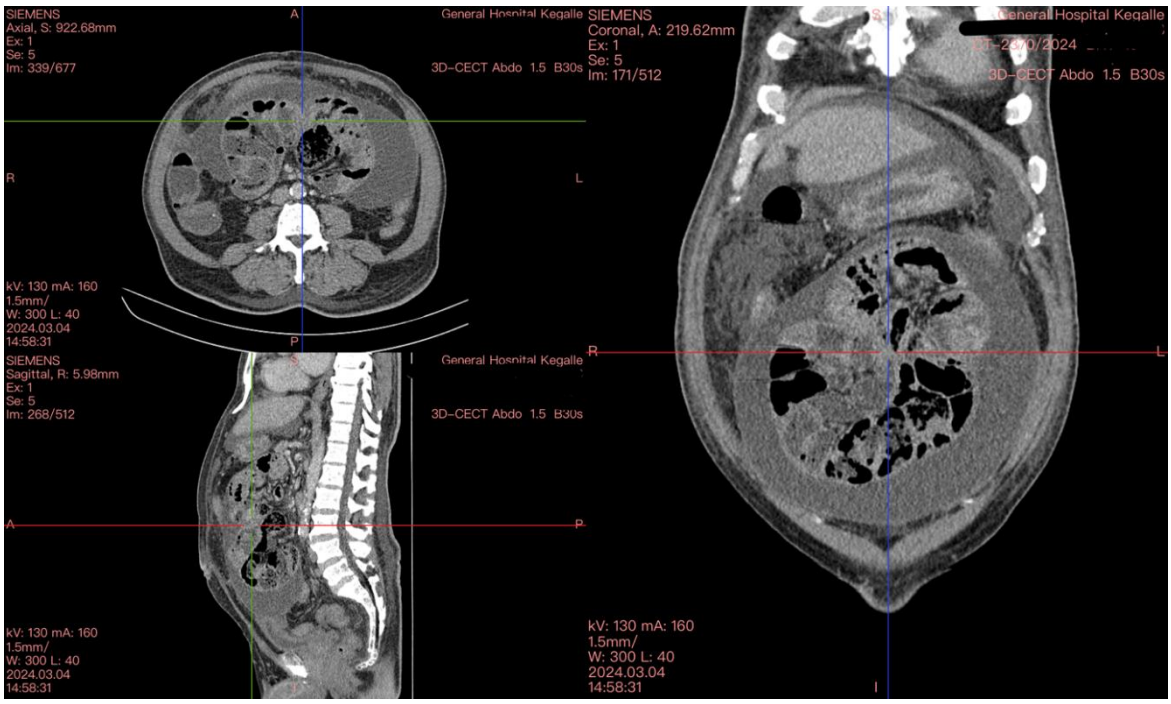

Figure 2: CT abdomen and pelvis of the patient

CT reported a large fluid-filled sac with the small intestine inside, most likely an internal hernia. Suspicion of an internal hernia led to the patient undergoing emergency laparotomy. During laparotomy, the fluid-filled sac was found to be a thickened peritoneal sac – an abdominal cocoon containing the small intestine (Figure 3). The small intestine was densely adhered and packed inside the cocoon. The cocoon was filled with blood-stained fluid. The sac was excised and all the small intestinal adhesions excised during the laparotomy and the patient was transferred to the ICU for post-op care.

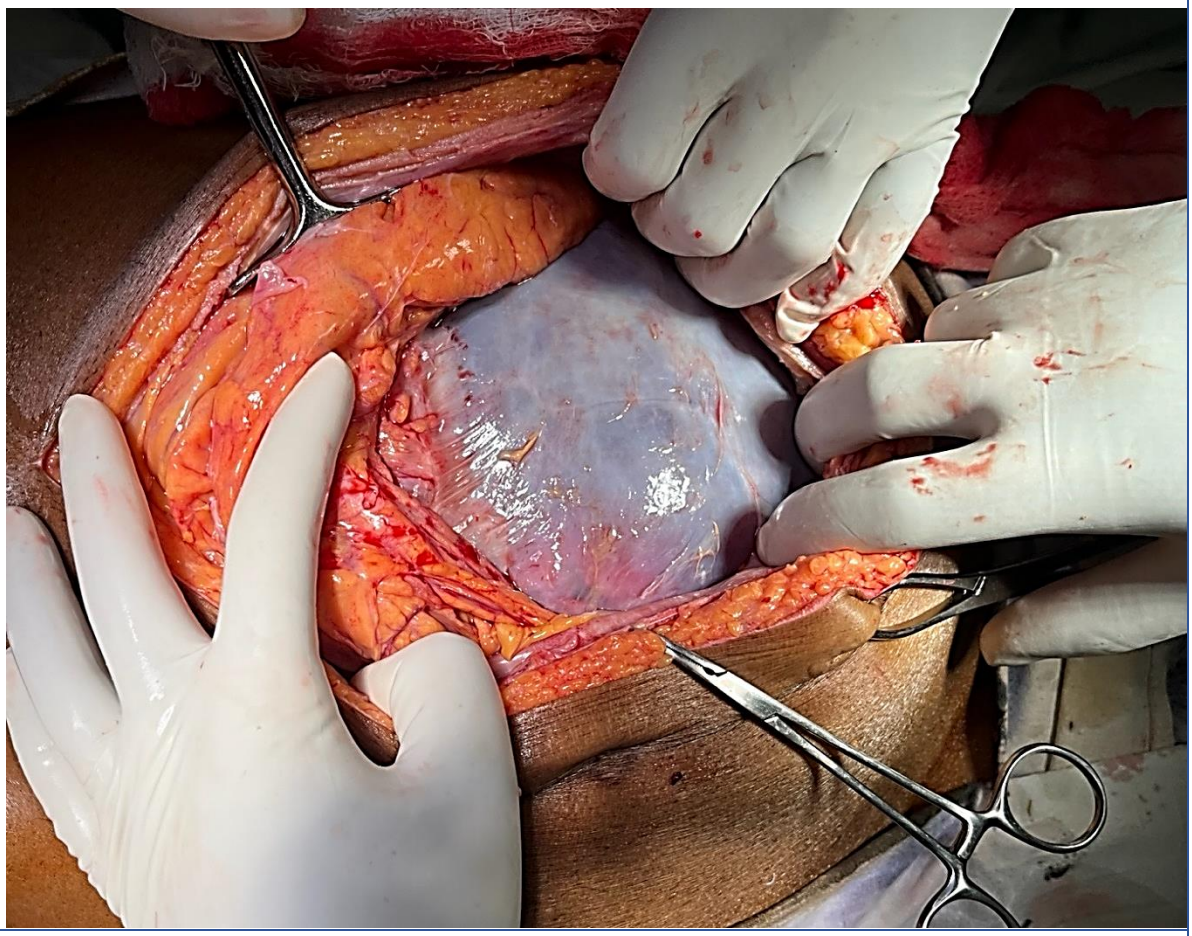

Figure 3: Abdominal cocoon encountered upon entering the peritoneum

The immediate post-op period was uneventful. He underwent regular haemodialysis and was started on TPN. He was transferred to the ward on post-op D3. The patient developed respiratory symptoms on D5 which were managed with IV antibiotics and chest physiotherapy. The patient passed away on D6 post-op. Post-mortem revealed no enterotomy or evidence of intra-abdominal sepsis and only revealed evidence of respiratory tract sepsis.

Discussion

PD is an established way of dialysis in end-stage kidney disease patients. Survival of patients undergoing PD is similar to that of haemodialysis[1]. One of the long-term complications of PD is EPS. It is associated with high morbidity and mortality[1,2]. EPD was first described in 1907 by Owtschinnikow and he named it “peritonitis chronica fibrosa incapsulata”[3]. Subsequently, with better understanding of the nature and pathophysiology of the disease the nomenclature changed to encapsulating peritoneal sclerosis [3]. EPS is not exclusively associated with PD and is found in systemic autoimmune diseases, peritoneal and gastrointestinal malignancies and peritoneal infections[4].

The youngest patient reported to have EPS is a 2-day-old neonate following meconium peritonitis[6] and the oldest, an 82-year-old man who was incidentally diagnosed during laparotomy for small bowel obstruction[7].

There are 4 stages of EPS described by Nakamoto in 2005[8].
Stage 1 - Pre-EPS - Asymptomatic with mild ascites and no inflammation
Stage 2 - Inflammatory phase - Patients are symptomatic with nausea and diarrhoea correlated with partial encapsulation of bowel and intestinal swelling, mild inflammation and fibrin exudate present
Stage 3 - Encapsulation - Symptoms of bowel obstruction due to the formation of a fibrous cocoon causing encapsulation
Stage 4 - Chronic stage of ileus - Absolute bowel obstruction caused by thickening of the encapsulating fibrous cocoon

Development of EPS is associated with the duration of PD[1–4,8], acute cessation of PD[1,2,8], organ transplantation[2,8], recurrent episodes of bacterial peritonitis[2–4,8], composition of dialysate (low pH, high glucose)[2–4], young age[8], ultrafiltration failure[8] and exposure to cleaning agents used to clean peritoneal dialysis, such as chlorhexidine/povidone iodine[2,8].

Presenting symptoms and signs of EPS are vague and non-specific[3]. EPS frequently presents with severe abdominal pain (86%), abdominal distension (82%), nausea and vomiting (54%), inability to maintain nutrition and weight loss (75% with a mean BMI of 17.5kg/m^2)[5]. In addition, stepwise symptom progression is an important feature with progression of the disease stage[1]. This progression manifests early as bloody ascites, appetite loss, nausea, diarrhoea, abdominal pain and progresses to much more severe symptoms of intestinal obstruction such as constipation, severe malnutrition, weight loss and abdominal mass[1].

Current imaging modalities are not sensitive enough to diagnose early stages of EPS[4]. Abdominal X-rays are usually normal due to the subacute/chronic nature of the disease unless complications such as complete obstruction or perforation occur[3,5]. Diffuse peritoneal calcifications can be seen in abdominal X-rays in advanced disease[9]. Ultrasound scans show clumping of the small intestine in the centre of the abdomen, resembling a cauliflower[3]. CT is the mainstay for the diagnosis of EPS [3,5]. Enhancement of the thickened peritoneum which progresses to encapsulate the small intestine (cocooning) and loculated ascites are seen in the contrast-enhanced CT[9]. Intravenous contrast is sufficient to make the diagnosis of EPS. In addition, oral contrast helps to delineate the bowel loop but it is not always indicated[3]. CECT also helps to identify complications associated with EPS such as bowel gangrene and perforation[3,9].

The level of evidence available on the management of EPS is weak and no randomized trials have been done so far[4]. After the diagnosis of EPS, PD should be stopped[1,5,8,10]. Early intervention in nutrition optimization is required to prevent disease-related and surgery-related mortality and morbidity. Most patients require total parental nutrition[1,5,8].

Numerous medications including immunosuppressants were tried, targeting the inflammatory components involved in EPS development. Commonly corticosteroids[1,2,4,5,10], tamoxifen[1,2,5,10] and immunosuppressants (azathioprine, mycophenolate mofetil)[2,5,8,10] are used to manage EPS. Corticosteroids are considered the drug of choice, specifically in the early stages of the EPS[8]. Even though there are many drugs used to treat EPS, only corticosteroids and tamoxifen have showed promising results but the effects are not conclusive[1,5,8,10].

In the past, most patients with EPS were treated with surgery because the diagnoses were made intraoperatively following laparotomy for unexplained recurrent intestinal obstruction[3]. Surgery is indicated in severe stages of EPS[2] and it is associated with high mortality around 32-35 %[1,5]. Surgical options include stripping of thickened parietal and visceral peritoneum and release of the small intestine off adhesions[2–5,8,10] which demand more careful dissections to avoid inadvertent enterotomy that leads to fatal complications[4]. The best results are achieved when surgery is performed early in the course of EPS[3].

Despite the advanced diagnosis modalities and development of treatment strategies, EPS is still associated with high mortality[2,3]. The most common causes of death are malnutrition and sepsis[2]. In more than 60% of patients with more severe disease, death occurs within 4 months from the diagnosis either due to complications of bowel obstruction or iatrogenic complications[3].

Even though there are possible mechanisms that explain the development of EPS, there are still no strategies to prevent EPS[8]. However, associated risk factors suggest some preventive approaches that might be helpful to prevent EPS[1,4]. The strong association with PD and duration of PD suggest an expiry date for PD, but no recommendation can be given with current evidence. Japanese data suggest discontinuing PD after 8 years to reduce the development of EPS[4,8]. Suggestions can be made for the minimization of dialysate glucose exposure and the prevention of peritonitis[1,4.]

Conclusion

Despite advancements in diagnostic techniques and management choices in the medical field, EPS remains without established management guidelines. There is little awareness and knowledge among clinicians. It is important to seek and diagnose EPS in high-risk patients due to its association with high mortality. All clinicians should be encouraged to report cases of EPS and a national registry should be maintained.

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