Hong Kong College of Physicians
Case report for Interim Assessment
Specialty Board of Advanced Internal Medicine (AIM)

For AIM Training, case reports should be submitted in the prescribed format together with the application form for Interim Assessment at least EIGHT Weeks before the date of Interim Assessment

Name of candidate (print and sign):
Hospital and Unit: Specialty:
Name of supervisor (print and sign)
Date(s) and place (hospital) of patient encounter: 14/4/2022 – 3/5/2022
Date of report submission: 8/9/2022

Case report

Note: Failure to follow the prescribed format (including the number of words) results in a FAILURE mark (score between 0 and 4) for the Case Report.

Title: A case of Encapsulating peritoneal sclerosis

Case history:

Mr. Lee was a 64-year-old gentleman with a history of hypertension, obesity, diabetes mellitus with diabetic nephropathy. He developed end stage renal failure and started continuous ambulatory peritoneal dialysis (CAPD) since October 2015. He had an episode of eosinophilic peritonitis in August 2018, an episode of serratia peritonitis in April 2019 and a severe episode of tuberculosis (TB) peritonitis in January 2020. His mode of renal replacement therapy was changed to haemodialysis since October 2020 in view of recurrent episodes of peritonitis and CAPD ultrafiltration failure. He had been receiving twice-weekly in-centre haemodialysis regularly prior to the index admission.

Upon admission, he presented with absence of bowel opening with no flatus, colicky abdominal pain, abdominal distension, and repeated vomiting up to three times a day. His last bowel opening was one week before the admission. There was no per-rectal bleeding, no passage of tarry stool or mucus. Systemic review of the respiratory, cardiac, neurological or urinary systems were all unremarkable.

His vital signs were stable and he was afebrile. There was no pallor or dependent edema. Abdominal examination revealed a moderately distended abdomen with mild central tenderness without guarding or rigidity. Per rectal examination revealed an

empty rectum with no mass. Erect chest radiograph showed clear bilateral lung fields with no free gas under diaphragm. Abdominal radiograph revealed faecal loaded bowels but no dilated bowel loops. Initial bloods tests were unremarkable.

The provisional diagnosis was constipation. He was put on oral and rectal laxatives. On the first night after admission, he complained of worsening and excruciating abdominal pain with increasing abdominal distension. Surgical team was urgently consulted and subsequently a contrast computed tomography (CT) of abdomen and pelvis was performed and showed close loop small bowel obstruction with a cystic mass compressing on the small bowel without pneumoperitoneum.

Emergency laparotomy was performed under general anesthesia. Intraoperative finding showed a large abdominal cyst near the abdominal wall, cocoon abdomen with no identifiable plane to approach the bowel. The operative diagnosis of encapsulating peritoneal sclerosis (EPS) was made. Surgical team suggested that the abdomen was not feasible for safe adhesiolysis and decided not to proceed to adhesiolysis. The abdomen was closed after a tubal drain was inserted for cystic fluid drainage.

The patient was transferred to intensive care unit for postoperative care. He was put on continuous veno-venous hemofiltration as the renal replacement therapy. The follow-up CT scan one week later showed an increased size of the large intra-abdominal cystic fluid collection and an increased amount of fluid and air in the collection despite previous continuous drainage. The radiological differential diagnosis was worsening air-forming infection versus leakage of air from the bowel in the collection. Surgical team suggested that the patient was no longer a surgical candidate for re-operation. Unfortunately, he was complicated with hospital-acquired pneumonia, septic shock with increasing respiratory and inotropic support. After discussion with the patient and his family members, conservative care was offered in view of grave prognosis of late-stage EPS. He eventually succumbed three weeks after admission.

Discussion and literature review

Encapsulating peritoneal sclerosis (EPS) is an uncommon but one of the most devastating complications of long-term peritoneal dialysis (PD). It is characterized by extensive peritoneal inflammation, fibrosis and thickening, and formation of fibrous cocoon encapsulating the bowel. It is a rare condition with the incidence varies between 0.7 and 13.6 per 1000 patient-years.[1-2] The morality was high, ranging from 35 to 50 percent in some studies.[1-2]

The duration of PD therapy is the single most important risk factor for the development of EPS. The incidence of EPS was shown to be increased with the duration of PD in an Australian study in 1998.[3] Thereafter, a Japanese prospective study in 2004 also reported similar findings.[2] However, most cases of EPS, just like our case, are diagnosed after discontinuation of PD therapy. The history of multiple episodes of severe peritonitis appears to be the second most important risk factor. In a UK study, patients with three or more episodes of peritonitis made up the largest proportion of the study population.[4]

Interestingly, some studies suggest that EPS is associated with other conditions such as autoimmune diseases, sarcoidosis, peritoneal and intra-abdominal malignancies, chronic peritoneal ascites, intra-peritoneal chemotherapy, intra-peritoneal exposure to particulate matter or disinfectant, abdominal surgery, endometriosis, intra-peritoneal TB infection, beta-blocker and calcineurin inhibitor administration.[5]

In our case, Mr. Lee developed TB peritonitis in January 2020 within his five-year's of PD therapy from 2015 to 2020. It is postulated that his history of TB peritonitis and the prolonged duration of PD therapy made him more susceptible to the development of EPS.

Typically, EPS is a slowly progressive clinical disorder that occurs after many years of PD therapy (generally over 5 years), It usually develops in a stepwise manner, with early (inflammatory) and late (ileus) stages. Unfortunately, symptoms are invariably non-specific in the early stage. Patients may report intermittent anorexia, nausea, early satiety, vomiting, abdominal pain and diarrhoea. Similarly, the physical examination is often unrevealing in this stage. Patients who are on PD therapy may show symptoms of fluid overload despite a history of adjusting of the peritoneal dialysis prescription to increase ultrafiltration. Occasionally, blood-stained dialysate may be observed on exchanges. Symptoms would become more prominent with disease progression into the late stage. The encapsulation of bowel from the peritoneal adhesions gives rise to the classical features of intestinal obstruction including severe

cramping, colicky abdominal pain, constipation and repeated vomiting. Physical examination may show an abdominal mass or abdominal distension by this stage.[6]

Given that the symptoms in the early stage are usually vague and non-specific, EPS is often not recognized in its early stages and patients usually, just like in our case, present late in the disease course. Therefore, a high index of clinical suspicion is crucial for the diagnosis EPS.

To date, there is no laboratory finding specific for the diagnosis of EPS. The peritoneal dialysis fluid may show white blood cells, but these are usually non-specific. Radiologically, CT scan may reveal peritoneal enhancement and calcification, bowel thickening, tethering, and dilatation with bowel tethering and peritoneal calcification being the most specific findings. An unremarkable CT finding is unusual in late EPS but does not completely exclude early EPS.[7]

A presumptive diagnosis of EPS can be made on the basis of characteristic CT findings together with the compatible clinical presentation. In many occasions, the only way to confirm the diagnosis of EPS is laparotomy and/or laparoscopy.[6]

At surgery, advanced cases of EPS typically show a thickened brownish peritoneum with a cocoon-like encapsulation of the entire bowel by the visceral peritoneum. The bowel loops are adherent to one another. The visceral peritoneum is fibrosed and thickened. Macroscopic peritoneal calcification may be present. In severe cases, adhesions between the visceral and parietal peritoneum may be present as well.[8]

Treatment of EPS consists of medical therapy in the early stage and surgery in the late stage. Before starting treatment, patients are recommended to temporarily stop PD therapy (if the patient is still on PD therapy) to rest the peritoneum. Such patients must be maintained on HD via a temporary central venous catheter.[9]

In the early phase of EPS when the inflammation is evident, corticosteroids are the drug of choice since steroids act by suppressing inflammation, preventing fibrin deposition and collagen synthesis and maturation. They may also possibly prevent ascites formation and accumulation. The favourable clinical outcome and improved survival with corticosteroids have been shown in various case reports and series. Nonetheless, the clinical effect of corticosteroids tends to reduce in the later stage of EPS owing to increased fibrosis and bowel obstruction. Other forms of immunosuppressants including azathioprine, mycophenolate mofetil have been used alone or combined with corticosteroids to treat EPS. However, the efficacy of these

drugs was only studied in small series or case reports.[9]

Tamoxifen is a selective estrogen receptor modulator (SERM) with antifibrotic property used in fibrotic disorders like retroperitoneal fibrosis, fibrosing mediastinitis and fibrosing cervicitis. Various case reports and small series have reported satisfactory outcome with tamoxifen in the treatment of EPS. Tamoxifen may exert its effect through its inhibition and modulation of transforming growth factor β (TGF-β), leading to reduced peritoneal fibrosis and angiogenesis. However, clinical use of tamoxifen is hindered by its potential adverse effects of venous thromboembolism, endometrial and uterine malignancies. As such, further studies are needed to address the clinical efficacy and safety profile of tamoxifen in treating EPS.[10]

Surgery involving peritonectomy, careful enterolysis to release the bowel is usually indicated in patients with advanced EPS presenting with bowel obstruction or acute abdomen. Although surgery is the most successful form of treatment in advanced EPS, it carries a high mortality (ranges from 19 to 34.5 percent) and high recurrence rates at around 25 percent. Therefore, surgery is usually performed in selected centres only where experienced surgical experts are available.[9]

In summary, this case illustrates the clinical presentation, diagnosis, and possible treatment strategies for EPS. Given the rarity and the devastating nature of this condition, a high index of clinical suspicion in at risk groups remains crucial for clinicians to make an early and definite diagnosis of EPS.

Tables and figures (where applicable) (no more than two figures)

Reference (not more than 10)

1. Brown EA, Van Biesen W, Finkelstein FO, Hurst H, Johnson DW, Kawanishi H, Pecoits-Filho R, Woodrow G; ISPD Working Party. Length of time on peritoneal dialysis and encapsulating peritoneal sclerosis: position paper for ISPD. Perit Dial Int. 2009 Nov-Dec;29(6):595-600.
2. Kawanishi H, Kawaguchi Y, Fukui H, Hara S, Imada A, Kubo H, Kin M, Nakamoto M, Ohira S, Shoji T. Encapsulating peritoneal sclerosis in Japan: a prospective, controlled, multicenter study. Am J Kidney Dis. 2004 Oct;44(4):729-37.
3. Rigby RJ, Hawley CM. Sclerosing peritonitis: the experience in Australia. Nephrol Dial Transplant. 1998 Jan;13(1):154-9.
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5. Kawanishi H, Moriishi M. Epidemiology of encapsulating peritoneal sclerosis in Japan. Perit Dial Int. 2005 Apr;25 Suppl 4:S14-8.
6. Kawaguchi Y, Kawanishi H, Mujais S, Topley N, Oreopoulos DG. Encapsulating peritoneal sclerosis: definition, etiology, diagnosis, and treatment. International Society for Peritoneal Dialysis Ad Hoc Committee on Ultrafiltration Management in Peritoneal Dialysis. Perit Dial Int. 2000;20 Suppl 4:S43-55.
7. Kawaguchi Y, Saito A, Kawanishi H, Nakayama M, Miyazaki M, Nakamoto H, Tranaeus A. Recommendations on the management of encapsulating peritoneal sclerosis in Japan, 2005: diagnosis, predictive markers, treatment, and preventive measures. Perit Dial Int. 2005 Apr;25 Suppl 4:S83-95.
8. Honda K, Oda H. Pathology of encapsulating peritoneal sclerosis. Perit Dial Int. 2005 Apr;25 Suppl 4:S19-29.
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10. Cornelis T, Oreopoulos DG. Update on potential medical treatments for encapsulating peritoneal sclerosis; human and experimental data. Int Urol Nephrol. 2011 Mar;43(1):147-56.

No of words in Case History and Discussion (excluding references): 1508

(should be between 1000-2000)

Declaration

I hereby declare that the case report submitted represents my own work and adheres to the prescribed format. I have been in clinical contact with the case selected. The case report has not been submitted to any assessment board or publication and it is NOT related to my second specialty(ies), if any. My consent is hereby given to the College to keep a copy of my case report, in written and/or electronic, at the College Secretariat and allow the public to have free access to the work for reference.

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(signature of Trainee)

Endorsed by Supervisor *

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