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Case report

Encapsulating peritoneal sclerosis with intestinal serosal sclerosis: A rare cause of intestinal obstruction in children on continuous ambulatory peritoneal dialysis: A case report

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ABSTRACT

Encapsulating peritoneal sclerosis (EPS) is a rare cause of intestinal obstruction in children. A 13-year-old boy on peritoneal dialysis (PD) for 2 years underwent laparotomy for intestinal obstruction which revealed extensive peritoneal thickening, encapsulation, and cocooning. The child got relief after creating ventral hernia. Obstruction recurred, and he died of sepsis. An active surveillance for EPS in children on PD should be carried out.

1. Introduction

Encapsulating peritoneal sclerosis (EPS) was first reported in adults in 1980 in the patients undergoing peritoneal dialysis (PD).[1] EPS is rare in children and presents clinically as intestinal obstruction. Histologically, it is characterized by severe submesothelial sclerosis >400 μm in thickness.[2] We report a rare case of EPS with intestinal serosal sclerosis in a child with chronic kidney disease (CKD) on continuous ambulatory peritoneal dialysis (CAPD).

2. Case report

A thirteen-year-old boy, who had been on CAPD for two and half years as a consequence of CKD secondary to posterior urethral valves with reflux nephropathy, presented with abdominal pain and multiple episodes of bilious vomiting, for 7 days. Initially, PD was started with 1.5% glucose Dianeal PD fluid. But as the time passed on, the dextrose concentration needed to be increased to 2.5% to get the required ultrafiltrate. Two months before presentation, ultrafiltration failure was noted (even no response to usage of 4.5% dextrose PD fluid) which was also documented on peritoneal equilibrium test (PET). After this, an option to switch over to hemodialysis was given but refused by the parents. During his last two and half years of CAPD, he had three episodes of culture-positive peritonitis which were treated with appropriate antibiotics. Supportive treatment of CKD was continued in the form of iron supplements, recombinant erythropoietin, calcium, vitamin D, bicarbonate supplementation, and antihypertensives including the beta blockers.

Plain abdominal radiograph showed multiple air fluid levels in the central portion, suggestive of small bowel obstruction. Contrast-enhanced computed tomography (CECT) scan of the abdomen showed a cluster of contrast-filled mildly dilated jejunal loops in the left hypochondrium. Multisegmental areas of circumferential thickening of distal jejunal and ileal loops were noted with adjacent mesenteric stranding. The child was taken up for an exploratory laparotomy. It revealed a large cocoon formed by the thickened omentum, which was excised. (Fig. 1a). The parietal peritoneum was covered by a thick hyperpigmented coating, which looked like skin. It was peeled off at multiple sites. There was no mechanical obstruction; therefore, cause of intestinal obstruction was adjudged to be compartment syndrome due to the peritoneal sclerosis. Hence, a ventral laparostomy was created to relieve the compartment syndrome secondary to intraperitoneal encasement (Fig. 1 c). Histopathology of the peritoneal specimen showed a denuded mesothelium. The submesothelial interstitial tissue

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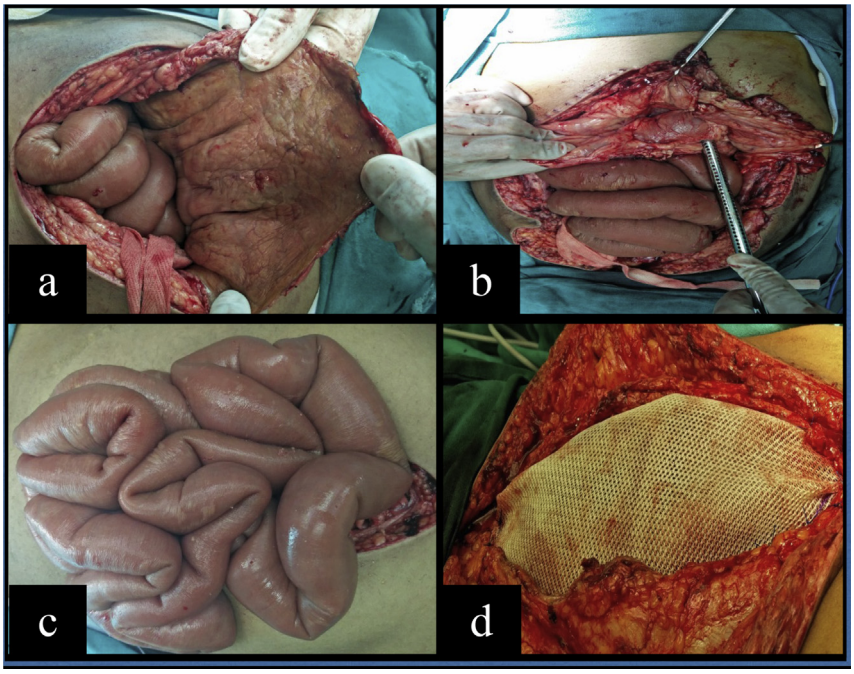

Fig. 1. Findings at laparotomy: (a, b) omental cocoon, (c) sclerosed bowel loops, and (d) mesh for ventral hernia.

showed plump fibroblasts along with inflammatory cells, fibrosis, and proliferating capillaries (Fig. 1 a). Extensive fibrin deposition was seen over the peritoneal surface (Fig. 2 b).

Ventral hernia was closed by a prolene mesh (Fig. 1 d). However, he developed infective complications later, requiring two surgical interventions to drain collections. However, despite all efforts, the child could not be saved and died of multiorgan failure and fungal sepsis.

3. Discussion

EPS is a clinical syndrome characterized by signs and symptoms of an obstructive ileus with peritoneal thickening and encapsulation, cocooning, and/or peritoneal calcification.[3] Duration of PD is implicated as the single most important factor for EPS; however, other factors such as drugs (beta blockers) and recurrent bacterial and fungal peritonitis are also recognized. The incidence of EPS in

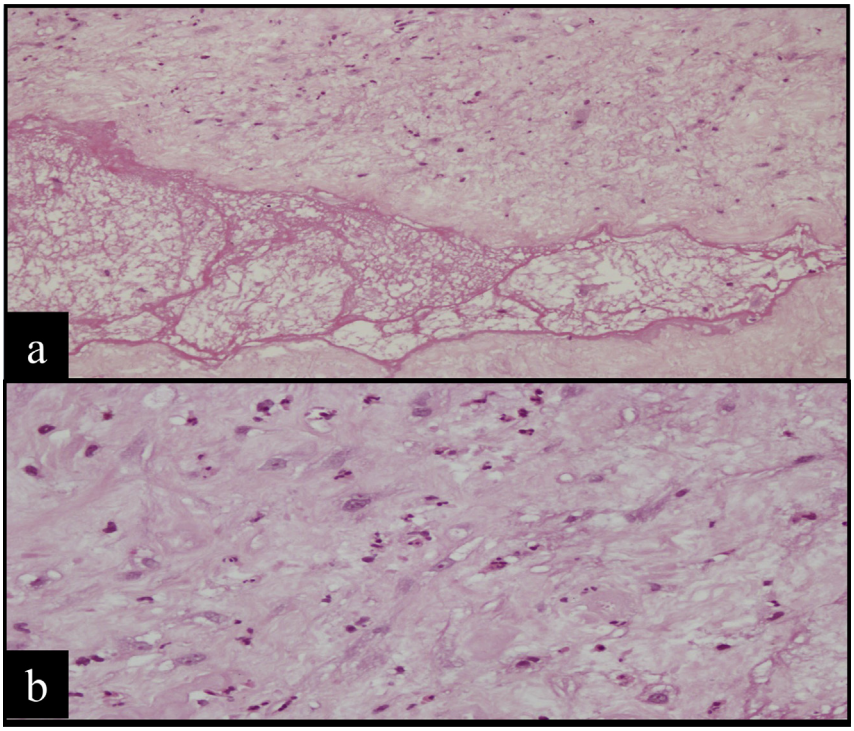

Fig. 2. HPE of the peritoneum: (a) denuded mesothelium and (b) hyalinization of collagen in epithelium. HPE, histopathological examination.

children on PD < 5 years is 0.45%; however, with PD > 5 years, incidence is 21%. However, in our case, the EPS occurred only within two and half years of PD.

The main pathophysiology is due to nonenzymatic glycosylation by dialysate glucose causing fibrin deposition and hyalinization of the superficial stream collagen leading to excessive fibrogenesis. Histology shows loss of the mesothelial cell layer and fibrin deposition on the peritoneal surface.

The usual presentation in children is with clinical triad of abdominal pain, vomiting, and weight loss. Diagnosis is mainly based on imaging and histological examination. Imaging may reveal frank intestinal obstruction or delayed transit time. Ultrasound is highly sensitive, showing a typically thickened bowel with a trilaminar appearance and adhesion of bowel loops to the anterior abdominal wall. Peritoneal calcification and clouding of mesenteric fat are the usual findings on the CECT scan abdomen.

Various treatment modalities have been suggested including immunosuppressants, tamoxifen, and surgical debridement. Surgical management plays a key role in patients presenting with intestinal obstruction. Meticulous lysis of the serosal membrane is performed. EPS carries a poor prognosis with high mortality ranging between 56% and 93%. EPS typically affects the peritoneal membrane; however, in our case, it extended to the entire serosal surface of the gut in the infracolic compartment. The supracolic compartment was relatively free.

EPS is a severe complication of PD, often presenting with intestinal obstruction. Surgical treatment may require debridement and creation of ventral hernia to address the compartment syndrome. It is difficult to treat and has high mortality. Not only is the composition of the dialysate fluid a causative factor, it also affects the incidence of EPS. The severity of irritation is directly related to the concentration of glucose solution used in PD fluid, and this could have been responsible for earlier presentation in this case.

The cause of death in our case was sepsis which could be related to the mesh. The immunocompromised status in view of CKD and its prolonged treatment may have been also contributory.

Conflict of interest

None.

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