WORLD JOURNAL OF PHARMACEUTICAL RESEARCH
SJIF Impact Factor 8.074
Volume 7, Issue 05, 181-190. Case Report ISSN 2277–7105

SCLEROSING ENCAPSULATING PERITONITIS COMPLICATING CONTINUOUS AMBULATORY PERITONEAL DIALYSIS SUCCESSFULLY TREATED WITH IMMUNOSUPPRESSION: CASE REPORT AND LITERATURE REVIEW

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Article Received on
10 Jan. 2018,
Revised on 01 Feb. 2018,
Accepted on 22 Feb. 2018
DOI: 10.20959/wjpr20185-11349

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ABSTRACT

Sclerosing encapsulating peritonitis (EPS) is well recognized serious complication of continuous ambulatory peritoneal dialysis (CAPD). We report a case of ESP complicating CAPD in a young female who presented with recurrent attacks of subacute small bowel obstruction and was treated successfully with immunosuppressive therapy**.**

KEYWORDS: Sclerosing encapsulating peritonitis, continuous ambulatory peritoneal Dialysis, immunosuppression.

INTRODUCTION

Sclerosing encapsulating peritonitis (EPS) is one the most serious complication of continuous peritoneal dialysis (CAPD) with reported high mortality and morbidity. We report a case of ESP in a 42 years old female presented 3 years after removal of the (CAPD) catheter with recurrent subacute intestinal obstruction. We also review the literature for pathogenesis, diagnosis and management of this serious complication.

CASE REPORT

A 42-year-old female was presented to our surgical unit with abdominal pain, constipation and frequent vomiting for 2 days. She is a known case of chronic renal failure on hemodialysis. She had been on continuous ambulatory peritoneal dialysis (CAPD) for 4 years, switched to hemodialysis for 8 years because of recurrent catheter related skin infections. She gave history of recurrent abdominal pain and vomiting and she was admitted before four times with subacute intestinal obstruction in the last 5 years. She was treated conservatively in all the occasions. On admission she was slightly dehydrated, not febrile. Her pulse was 92/minute, BP110 over 70 mm of Hg. her abdominal examination revealed slightly distended abdomen without tenderness or muscle rigidity. Her all blood tests including CBC, liver function test and coagulation screen were within normal values. Her plain abdominal x ray showed dilatation of small bowel loops at the center of abdomen and toward the left side with multiple air fluid levels (figure 1).

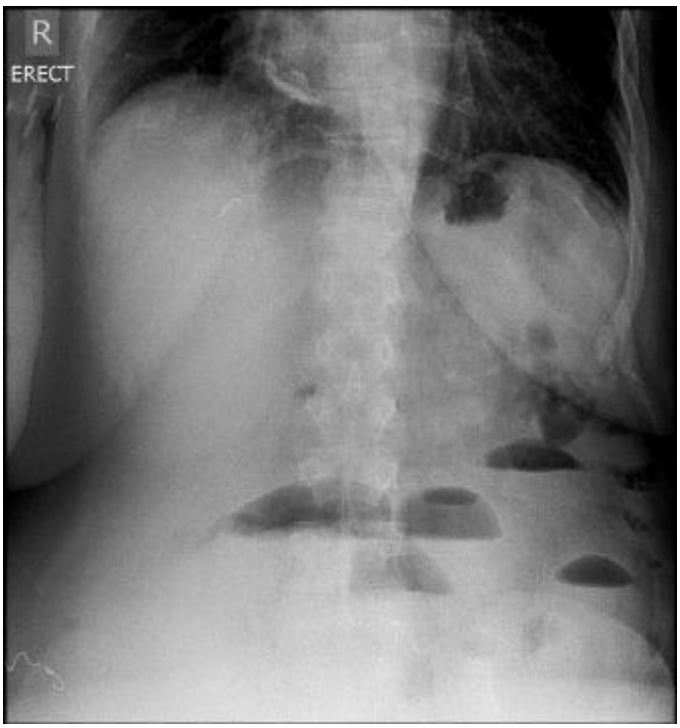

Figure 1: The plain x ray abdomen showing dilatation of small bowel loops at the center of abdomen and toward the left side, with multiple air fluid levels. There were no signs of pneumoperitoneum.

The chest x ray showed the hemodialysis catheter in situ with slight cardiomegaly and pulmonary congestion (figure 2).

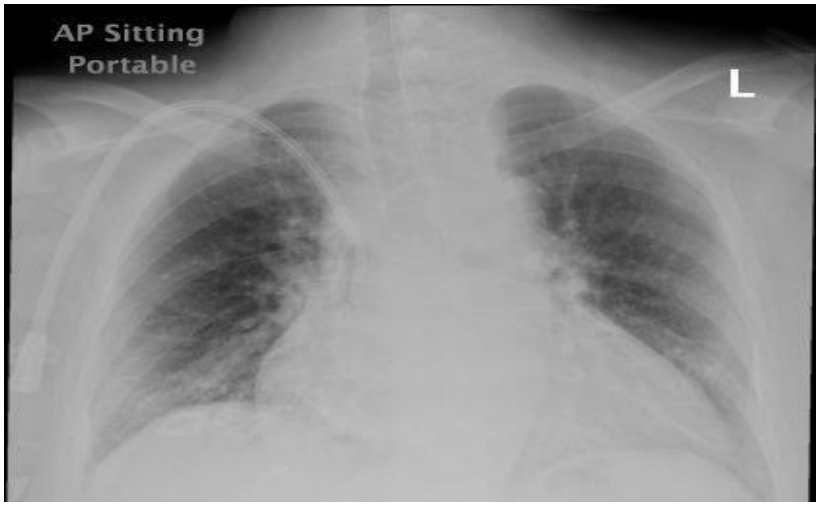

Figure 2: The chest x ray showing right-sided permcath in situ. the heart size is enlarged with evidence of pulmonary congestion.

Her CT scan showed cluster of collected dilated small bowel loops seen in the center of abdominal cavity, which are apparently enclosed within a sac like membrane that showed foci of calcification in its anterior wall with multiple mesenteric and retroperitoneal sub centimetric lymph nodes. There was no evidence of free fluid or air intra-peritoneally. Findings are highly suggestive of sclerosing encapsulating peritonitis (abdominal cocoon). Both kidneys showed evidences of chronic renal parenchymal disease (figure 3&4).

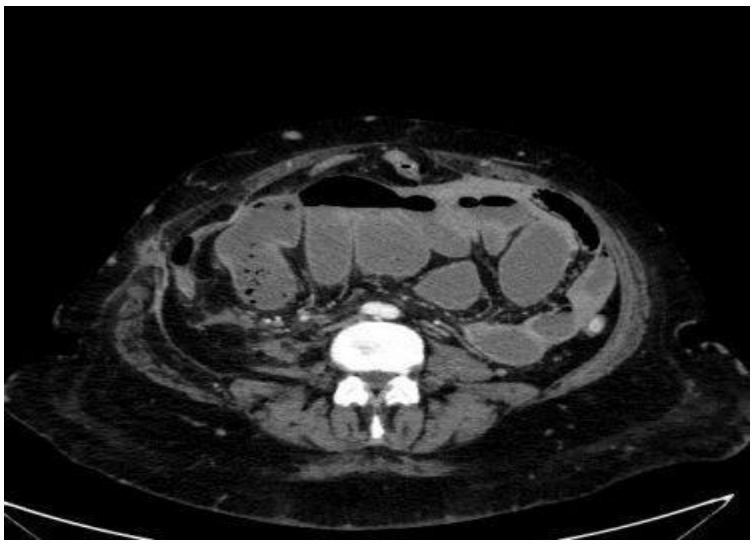

Figure 3: The abdominal CT scan showing cluster of collected dilated small bowel loops seen in the center of abdominal cavity, which are apparently enclosed within a sac like membrane that showed foci of calcification in its anterior wall with multiple mesenteric and retroperitoneal sub centimetric lymph nodes. There was no evidence of free fluid or air intra peritoneally.

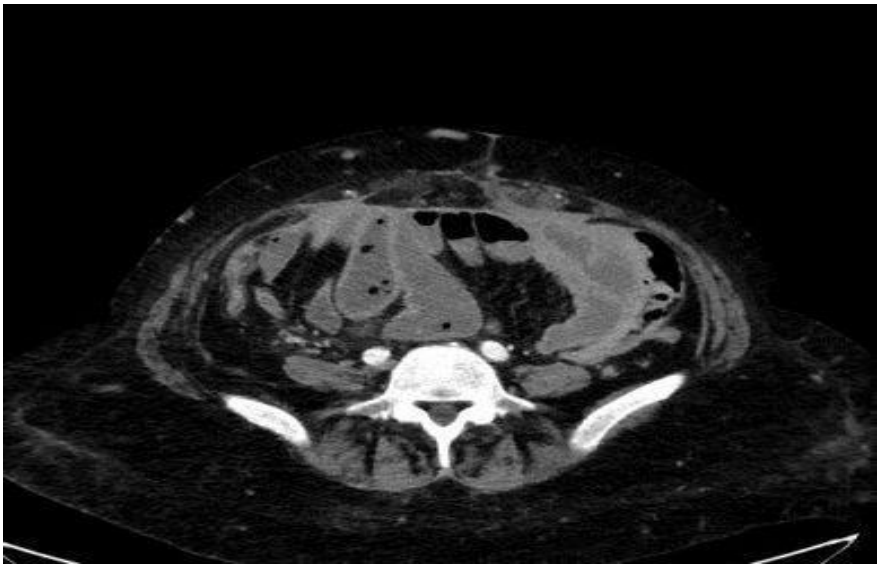

Figure 4: The abdominal CT scan showing the same findings of figure 3.

The patient was treated conservatively initially by nothing per mouth, nasogastric suction and intravenous fluids. Subsequently she was started on tamoxifen 40 mg daily and prednisolone 1 mg/kg body weight orally for the first months reduced to o.5 mg/kg body weight for 3 months and 0.25mg/kg body weight for another 3-month tapered down to 10mg /day for a year. She showed good response to the combined steroid /tamoxifen therapy and she remained symptoms free on follow up for 2 years.

DISCUSSION

Encapsulating peritoneal sclerosis (ESP) or (abdominal cocoon) is a condition characterized by total or partial encasement of the small bowel by a fibro collagenous cocoon-like sac.[1] it was first described by Owtschinnikow in 1907 as "peritonitis chronica fibrosa incapsulata"[2] and was named by Deeb et al, in 1998 as sclerosing encapsulating peritonitis.[3]

The condition has been classified as primary and secondary based on whether it is idiopathic or has a definite cause. Secondary ESP is commonly referred to as being multifactorial, genetic susceptibility may also be an etiological agent.[4]

Secondary ESP was described in association with sarcoidosis, systemic lupus erythematosus, indwelling abdominal catheters (specifically Le-Veen shunts), orthotopic liver transplantation, and tuberculous pelvic inflammatory disease.[2,3,5] It has also been also described in association with Some drugs, especially the beta-adrenergic blocker 'practolol, and as serious complication of continuous ambulatory peritoneal dialysis (CAPD).[6,7,8,9]

The association of ESP with long-term use of (CAPD) is well documented in the literature with few reported cases. It is estimated incidence varies between 0.7 and 3.3% in patients on peritoneal dialysis.[10]

The pathogenesis of SEP in relation to continuous ambulatory peritoneal dialysis (CAPD) remains uncertain. However, recurrent peritonitis has been reported to be the main cause.[11,12]

Nakamura et al. reported that SEP pathogenesis in long-term CAPD patients might be related to advanced glycation end-product accumulation in the peritoneal membrane and alteration in peritoneal cell function, accompanied by increased expression of various growth factors and peritoneal sclerosis, and finally followed by SEP.[1,13]

Recently, features of peritoneal cell infiltrate of EPS were studied. A characteristic mononuclear cell infiltrate consisting of CD4+ and CD163+ cells dominates the peritoneum of EPS patients. These findings suggest a role for both CD4+ T cells and M2 macrophages in the pathogenesis of EPS.[14,15] Increased serum sCD25 concentrations and peritoneal lymphocytosis in EPS patients indicate the involvement of activated T cells in the pathophysiology of excessive fibrosis.[14,16]

Levine et al[17] produced an animal model of sclerosing encapsulating peritonitis, for studies of the pathogenesis and prevention of complication of peritoneal dialysis by injecting rats with a chemical irritant and fresh whole rat blood intraperitoneally.

Similarly, Nakamoto et al[18] produced an animal model by injecting rats with different PH solutions (acidic dialysis solution PH 3.8) and (neutral dialysis solutions PH 7). They proved that long-term intraperitoneal injection of acidic dialysis solution produced features typical of EPS while neutral dialysis solutions protect against the development of EPS during peritoneal dialysis in rats.

It is becoming clearly evident now that ESP associated with diffuse inflammatory process affecting the peritoneum and constitutes a rare but life-threatening serious complication in patients on long-term (CAPD).[19,20] with a mortality that exceeds 30%.[21]

Diagnosis depend on high index of suspicious. The condition should be suspected in all patients who are on or had been on long term peritoneal dialysis presenting with partial or

complete intestinal obstruction. In the past diagnosis only made at laparotomy for intestinal obstruction, however with the increased use of CT scan for investigation of patients with intestinal obstruction, preoperative diagnosis became more feasible. Preoperative diagnosis is essential for avoiding intra operative surprise and allow proper planning of management.

Radiological examinations play key role in diagnosis of the disease. Plain x-rays may show dilated loops of small bowel at the center of the abdomen. Abdominal ultrasound may show clumping of bowel loops with the bowel surrounded by a thick rim of hypo-echoic tissue. Tethering of the bowel posteriorly or the presence of a membrane anterior to the small bowel may be seen.[22]

The classic barium study findings are a serpentine or concertina-like configuration of dilated small-bowel loops in a fixed U-shaped cluster. Some authors have described a cauliflower-like appearance on barium study.[23,24]

CT scan is considered the gold standard in diagnosis of ESP. The typical finding of ESP on CT is a concentration of the whole small-bowel to the center of the abdomen encased by a soft-tissue-density mantle.[25] Other CT features of abdominal cocoon include signs of obstruction, agglutination and fixation of intestinal loops, mural thickening, ascites and localized fluid collections, peritoneal thickening and enhancement, peritoneal or mural calcifications, and reactive adenopathy.[26]

Management usually depends on patient's presentation. Patients with mild recurrent abdominal pain and subacute intestinal obstruction may benefit from removal of peritoneal dialysis catheter together with trial of steroid and tamoxifen therapy.

Tamoxifen is a selective estrogen receptor modulator (SERM) that inhibits the production of TGF-beta by fibroblasts. It has been used in the past in several fibrosing syndromes such as retroperitoneal fibrosis and fibrosing mediastinitis.[27]

Allaria et al[28] were the first to describe the successful use of tamoxifen in an EPS patient, since then there was increasing trend of its use either alone or in combination of steroids in management of ESP. However, the reported experience with the use of tamoxifen for ERS is limited due to the low number of patients with EPS, and lack of large, randomized, controlled trials.

Cornelis T et al[29] reviewed the literature for updating the medical treatment of encapsulating peritoneal sclerosis. He found that between 1992 and 2007, 14 different groups have reported on their experience with tamoxifen therapy for EPS. The number of patients varied between 1 and 14 in each group. In total, 36 patients were studied. In 2 patients, the outcome was reported as 'resolved', 20 patients 'improved', 14 patients remained 'stable', 4 patients died and in 1 patient the outcome was not reported. He concluded by supporting the use of tamoxifen and prednisone for the treatment of EPS and also for prophylaxis in the early inflammatory phase of EPS.

The therapeutic potential of tamoxifen therapy is also confirmed in a significant proportion of other reported cases.[30] The Dutch EPS study which is the largest controlled series showed a decreased mortality in a group of EPS patients treated with tamoxifen (45.8 vs 74.4%, p=0.03) compared with a group who were not.[31] The recommended dose of tamoxifen is 40 mg/day.

In 1997, Mori and colleagues reported for the first time on the successful use of steroids alone in one patient with EPS.[32] The pharmacological mode of corticosteroid action on EPS is still unknown. However, the speculation is that it may be via both anti-inflammatory effect and the immunosuppressive effect.[29] In spite the uncertainty of their action, corticosteroids are the most reported successfully used drugs in treating EPS. Steroids are thought to be effective in suppressing the inflammatory process of the peritoneal membrane and inhibiting collagen synthesis and maturation.[33]

Although the optimum dose and duration of steroid therapy have not been established by a controlled trial, most publications support a regimen of prednisolone 0.5 to 1.0 mg/kg/day during the first month, 0.25 to 0.5 mg at months 2 and 3 and thereafter tapered to 10 mg at six months. Treatment with steroids must be continued for at least one year.[30]

Surgery is indicated in patients present with complete intestinal obstructing and in presence of signs of peritonitis. The typical finding at surgery is a conglomeration of small bowel loops encased in a dense white membrane.[34, 35] Treatment consists of excision of the accessory peritoneal sac with lysis of the inter-loop adhesions. Bowel resection is unnecessary[34], unless a non-viable segment is found.[35, 36]

Conflict of Interest: None declared

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