Case Report

Encapsulating Peritoneal Sclerosis after kidney Transplantation: Success of Medical Treatment

Abstract

Encapsulating peritoneal sclerosis (EPS) is an infrequent but serious complication of long-term peritoneal dialysis (PD). EPS may become clinically apparent when patients are on PD (classical EPS) or after undergoing kidney transplantation (post-transplantation EPS). This presentation of EPS seems to occur shortly after kidney transplantation in former PD patients. In this report, we present our experience in our first case of patient diagnosed with EPS after kidney transplantation.

Keywords: *Encapsulating peritoneal sclerosis, kidney transplantation, peritoneal dialysis*

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Introduction

Encapsulating peritoneal sclerosis (EPS) is an infrequent but serious complication of long-term peritoneal dialysis (PD). It consists of a progressive inflammatory process involving both visceral and parietal peritoneum leading to encapsulation of the adhered intestinal tract.

EPS is associated with high morbidity related to bowel obstruction and malnutrition. The prevalence of EPS is 0.9% in patients who have been on PD for less than 5 years, but it is 11.5% in patients on PD for more than 10 years.[1] After 1 year EPS diagnosis, the mortality rate is 42%.[2] EPS may become clinically apparent when patients are on PD (classical EPS) or after undergoing kidney transplantation (post-transplant EPS).[3] This presentation of EPS seems to occur shortly after kidney transplantation. We report our experience with the first case of EPS diagnosed after kidney transplantation.

Case Report

A 56-year-old female patient who had a family history of nephropathy was on PD for 6 years. During this period, peritoneal transport remained high, and ultrafiltration decreased over time. In addition, she had severe hypertension, but she never developed peritonitis.

She underwent a kidney transplantation from a living donor (her niece). The patient had received induction immunosuppressant with antithymocyte globulin and methylprednisolone, followed by maintenance immunosuppressant with mycophenolate mofetil, tacrolimus, and steroids. Five months after kidney transplantation, she developed an abdominal pain and distension. Blood test showed an elevated C-reactive protein, anemia, and hypoalbuminia.

An abdominal ultrasonography (USG) revealed pelvic fluid collection with left pleural effusion without organomegaly.

The analysis of the ascetic fluid revealed a bloody and exudative fluid. The culture was negative. The pathological exam of the fluid did not find malignant cells. A computed tomography (CT) scan revealed pelvic fluid collection with septa inside, but it did not describe the peritoneum aspect. Tumor markers were normal. The diagnosis of tuberculosis (TB) was ruled out. Therefore, we decided to perform a peritoneal biopsy that showed encased loops because of fibrosis, mesothelial detachment, and thickened peritoneum with fibrin deposition, inflammatory infiltrates from the sub-mesothelial layer toward the inside layer. The patient had a transient increase in steroid therapy, and we started with tamoxifen 20 mg twice a day initially than 20 mg/day for 6 months. In the

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following 6 months of follow-up, the patient's symptoms ameliorated, and her condition improved.

Discussion

When exposed to PD solutions, the peritoneal membrane undergoes many changes characterized by a loss of normal mesothelial cell morphology, expansion of the sub-mesothelial subcompact zone, and neovascularization of the peritoneal membrane. Calcifications are rare; therefore, they are the signs of a significant inflammatory state.[4] This condition is known as the simple peritoneal sclerosis (SPS); however, a minority of patients on PD develop a more serious complication, which is the EPS.

EPS is not exclusive to PD and has been associated with a range of conditions, including systemic autoimmune disease, diseases of the gastrointestinal (GI) tract, peritoneal and intraabdominal malignancies, exposure to talc or particulate matter, or the use of intraperitoneal disinfectant for peritoneal lavage and blocker administration.[5] For PD, the major risk factor for EPS is the PD duration. A history of abdominal surgery, recurrent peritonitis, high-glucose concentration and Icodextrin-based PD regimens, and high or high-moderate transport in peritoneal equilibrium test (PET) are other important factors for the development of EPS.[4,6,7] Paradoxically, the interruption by the washout increases the risk of EPS.[8] EPS presents after withdrawal from PD in the majority (70–90% in some series) of the patients,[5,9] and the time from cessation of PD until the development of EPS has been reported as up to 5 years. That could explain partly the onset of EPS after kidney transplantation. EPS occurs after transplantation only in patients who have been exposed to PD for several years. There appears to be no risk if the patients have been on PD for a short time.[10] Ideally, therefore, patients should be transplanted within 3–4 years of starting PD. The critical phase for post-transplantation EPS is during the first year after transplantation.

Our patient was on PD for 6 years, but she did not have a history of abdominal surgery and did not have peritonitis while on PD. She developed EPS 5 months after kidney transplantation. Second, the occurrence after kidney transplantation is explained by the buildup of profibrotic factors,[8] as well as the use of calcineurin inhibitor drugs, which give rise to increased fibrosis.[11] The calcineurin inhibitors (CNIs) can lead to enhanced expression of the transforming growth factor-b (TGF-b).[12] Third, clinical signs of EPS could be insidious and the diagnosis can be late after kidney transplantation. EPS usually presents with mechanical intestinal obstruction.[13,14] Symptoms include early satiety, anorexia, nausea, vomiting, constipation, diarrhea, weight loss, abdominal fullness, and pain.[15]

Signs of inflammation may be pyrexia, raised C-reactive protein (CRP), anemia, bloody dialysate, and ascites. Abdominal masses and pain may indicate peritoneal adhesions and/or cocooning, and there may be features of acute or subacute intestinal obstruction.[16] In this case report, the patient had raised CRP, anemia, and bloody fluid.

Current imaging techniques are insufficiently sensitive to detect the early stages of EPS. Screening with CT is of limited value as up to 50% of the patients with established EPS have had a normal abdominal CT within 2 years prior to diagnosis.[17] In this observation, the CT scan did not help for the EPS diagnosis.

The differential diagnosis includes tuberculosis peritonitis, peritoneal mesothelioma, carcinomatosis, or posttransplant small bowel lymphoma, and it is important to obtain histology where there is doubt.[16] Due to the immunosuppressant state and endemic state for TB in our country state, our patient had biological and histological exploration to rule out TB and abdominal malignancy. Only a fibrous cocoon wrapped around the bowel confirms the diagnosis. A thickened peritoneal membrane and intraabdominal adhesions are common in long-term PD and after peritonitis, particularly tuberculosis peritonitis, and are therefore not diagnostic.[18]

Reported therapy includes the use of immunosuppressant agents, predominantly, corticosteroids; antifibrotic agent—tamoxifen; changing CNIs with mTORi; reducing CNIs to initial dose; discontinuing mycophenolate mofetil (MMF); and nutritional support are medical treatment options.[12,19] Our patient had an increased steroid dose and tamoxifene. In fact, some studies showed that tamoxifen (20 mg twice a day) with steroids was useful for EPS and decreased EPS symptoms.[20,21] If medications fail to improve the patient's condition, surgery seems to be the best option. Surgical intervention consists of enterolysis/adhesiolysis is usually required in refractory cases, and in that cases, the mortality rate is high.[16,22]

There is no preventive strategy to reduce the risk of EPS, but there is some evidence to support the following[10]; low glucose exposure, preserved residual renal function, little or no peritoneal infection and low small solute transport status, which may confer a lower individual risk. Japanese data has suggested a dramatic increase in the incidence of EPS after 8 years of therapy, and recommendations have been made to preemptively discontinue PD at that stage.[19]

Conclusion

As EPS is one of the most dangerous complications of PD, patients on PD must be evaluated closely and informed about the complications of PD before transplantation. It should be kept in mind that EPS has an insidious nature and the overt clinical signs could appear even after kidney transplantation. Immunosuppressant, particularly corticosteroids and tamoxifen, are the treatment of choice. In advanced stages, adhesions and encased loops can be released by surgery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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