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Jaime Manuel Justo-Janeiro (fics-facs)[1, 2]*, Elizabeth Huerta-Calixto[2], de la Rosa Paredes René[2], Fernando Vázquez de Lara[3] and Luis G Vázquez de Lara[1]

[1]Faculty of Medicine, Meritorius Autonomous University of Puebla, Mexico
[2]Department of Organ Transplantation, General Hospital of Puebla "Dr. Eduardo Vázquez Navarro", Puebla, Puebla, Mexico
[3]Department of Internal Medicine, Icahn School of Medicine at Mount Sinai West and St. Luke's, New York, NY, USA

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***Corresponding author:** Jaime Manuel Justo-Janeiro, Department of Organ Transplantation, General Hospital of Puebla "Dr. Eduardo Vázquez Navarro", Puebla, Puebla, Mexico, Tel: 52 (222) 303-83-91; E-mail: jaime_justo@hotmail.com

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Research Article

Encapsulating Peritoneal Sclerosis: Different clinical presentations and their management

Abstract

Introduction: One of most common treatments of end-stage renal disease is peritoneal dialysis. Encapsulating peritoneal sclerosis is a complication in which the osmotic capacity of the peritoneal barrier is lost, due to infections or the irritating effect of dialysis solutions. This pathology has different clinical presentations, hence the need of different diagnostic and therapeutic methods.

Objective: To present a series of three cases with encapsulating peritoneal sclerosis and different clinical pictures, using laparoscopy as the mainstay for diagnosis and treatment.

Patients and methods: This work describes the clinical and imaging features of three patients. A systematic approach was utilized which included the delimitation of the affected zone, sterilization, collapse of the cavity, and change of dialysis mode until renal transplantation.

Conclusions: Our work suggests that a laparoscopic approach to encapsulating peritoneal sclerosis can be very valuable for the diagnosis and treatment of this condition and controlled clinical trials are warranted to validate this observation.

Introduction

According to The National Kidney Foundation Kidney Disease Outcomes Quality Initiative (NKF KDOQI) guidelines for chronic kidney diseases, end-stage renal disease (ESRD) is defined as an irreversible decline in kidney function, characterized by a level of glomerular filtration rate (GFR) below 15 mL/min/1.73 m2 or by the need of dialysis or transplantation, regardless of the level of GFR [1]. The etiology is diverse and age-related. In adults, diabetes (30-40%) and arterial hypertension (25-30%) are the most common causes, while in children, primary glomerulopathies and congenital abnormalities are the usual culprits [2].

Currently, patients with ESRD are offered peritoneal dialysis, hemodialysis or renal transplantation as renal replacement therapies [2,3]. Peritoneal dialysis is the most widely used, because it is inexpensive and can be performed in an ambulatory basis [4]. The first description of this therapy was made by Ganter in 1923, but it was until 1976 when Popovich et al. [5], described the basic concepts of continuous ambulatory peritoneal dialysis (CAPD) as they are known today. Current techniques allow peritoneal fluid instillation and drainage with minimum disruption in patient's lifestyle [6]. This therapy utilizes the peritoneal membrane's properties for diffusion and ultrafiltration and its effectiveness depends on the functional and structural integrity of the peritoneum. The peritoneal membrane is a complex and functional structure which is formed by a layer of mesothelial cells, connective tissue, blood vessels, lymphatic vessels and innate immune cells which can be affected by peritoneal dialysis in the long term [3,7].

Encapsulating peritoneal sclerosis (EPS) is defined by clinical and pathologic criteria, such as macroscopic honeycomb appearance, fibrin deposits, edema, mononuclear infiltration, fibroblast activation markers and fibroblast proliferation into the peritoneum [8,9]. EPS impairs the capability of the peritoneum for diffusion and dialysis and it must be suspected in any patient with evidence of ultrafiltration failure [10,11]. EPS was originally described by the International Society for Peritoneal Dialysis in patients with loss of the peritoneum osmotic capability and the diagnosis is confirmed by imaging studies. The images seen may be originated either by peritoneal abnormalities, such as liquid trilaminate appearance, organ adherence to the anterior abdominal wall, calcifications, peritoneal thickening and encapsulation [12]; or by small intestine abnormalities, such as abnormal peristalsis, intestinal obstruction, small intestine immobilization, liquid collections

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and a cocoon appearance. These findings can be seen with ultrasound, tomography or MRI. Clinically, patients may be asymptomatic or show evidence of intestinal obstruction with nausea, vomiting, abdominal distention, anorexia and weight loss, fever and an abdominal mass [13]. There is not a widely accepted biochemical marker to diagnose EPS; however, some authors have suggested that low levels of CA125 and high concentrations of IL6 in dialysis fluid have a 70% sensitivity and 89% specificity in diagnosing EPS. Honda has proposed that a persistently elevated C reactive protein may aid in the diagnosis of EPS [14].

The aim of this work is to describe three clinical presentation patterns found in EPS in a clinical case format along with laparoscopy-based therapeutic options.

Methods

We present the clinical and imaging features of three different patients with EPS. A systematic decision-making process for the resolution of each case was used, which comprised of the delimitation of the affected zone, sterilization, collapse of the cavity, and change of dialysis mode until renal transplantation.

Patient 1

An 18 year-old male with ESRD treated with CAPD for 42 months, was considered for renal transplantation. During the physical exam, a solid and fixed 20 x 30 cm epigastric mass was found, and laparoscopic exploration was offered. Intraoperatively a sclerotic peritoneum was observed in the inferior middle part of the abdomen with the peritoneal pigtail catheter in place (Figure 1). In the upper part of the abdomen a transparent membrane with small intestine inside (Figure 2) was also found. No further treatment was necessary.

Patient 2

A 41-year-old male with ESRD treated with CAPD developed ultrafiltration failure and began hemodialysis. A mesogastric 30 x 30 cm mass was discovered during the physical exam. Ultrasound was performed which showed liquid content, and computerized tomography (CT) imaging evidenced a mass occupying the supra and inframesocolic compartments (Figure 3). An abdominal paracentesis was performed. Culture of the fluid obtained was positive for *E. coli*. A laparoscopic exploration was performed which showed the presence of thrombi and fibrinopurulent debris in the peritoneal cavity. These were subsequently removed and curettage of the cavity was performed, leaving a drainage catheter inside (Figure 4). Follow-up was done on an outpatient basis until the fluid obtained from the drainage was serous and clean. The cavity was then occluded with 2% polydocanol and external compression. Three months later, the patient received a renal transplantation from a living donor without complications.

Patient 3

A 23-year-old male with ESRD treated with CAPD for 36 months, developed ultrafiltration failure, and was switched to hemodialysis. A mass covering the meso and hypogastric regions was apparent during physical examination (Figure 5). Ultrasound and CT imaging showed a liquid content within the mass which measured 110x173x154 mm. (Figure 6). Abdominal paracentesis was performed under spinal anesthesia and 2.5 liters of cloudy green liquid were obtained leaving a drainage catheter in place. Culture of the fluid showed *E. coli*, and antibiotic treatment was initiated with daily lavages for three months. The closure of the cavity was performed with polydocanol instillation and removal of the catheter. Renal transplantation was subsequently offered to the patient.

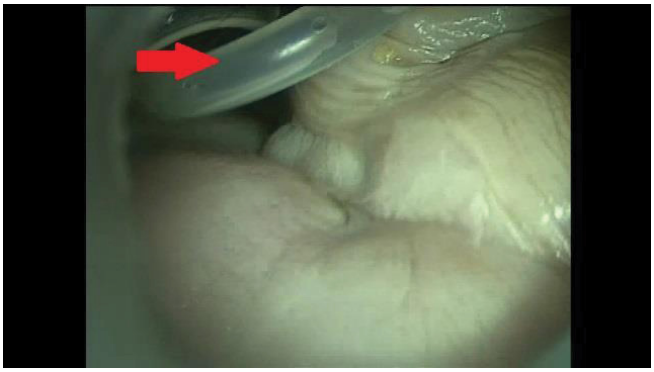

Figure 1: Peritoneal catheter (red arrow) and the fibrotic membrane covering the small intestine.

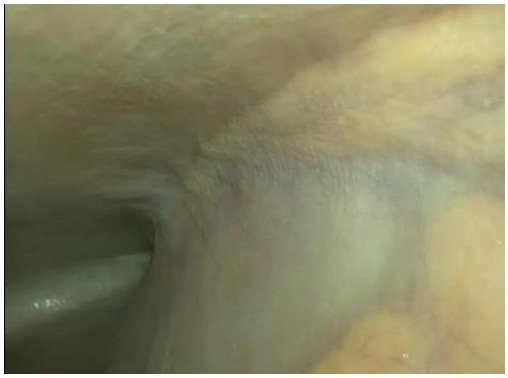

Figure 2: The peritoneal view shows the adherence from the fibrotic membrane covering the small intestine to the anterior abdominal wall.

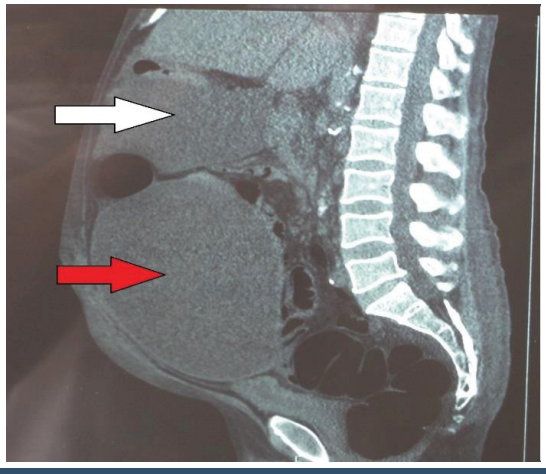

Figure 3: In this sagittal abdominal tomography we can observe the supramesocolic collection (white arrow) and the inframesocolic collection (red arrow).

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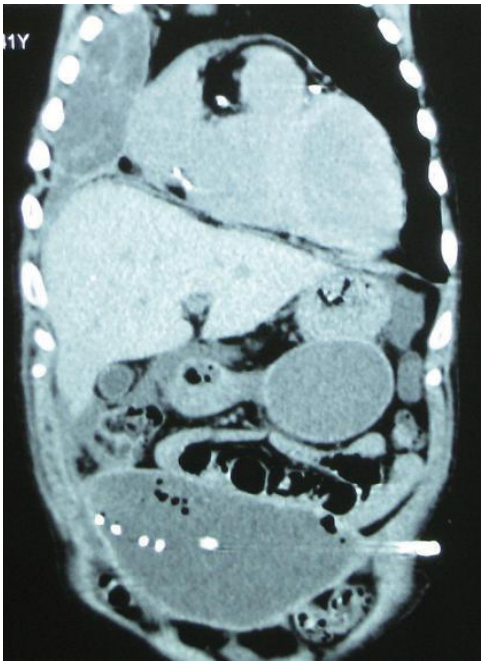

Figure 4: In this coronal view we can see the catheter in the abdominal collection to drain all the debris.

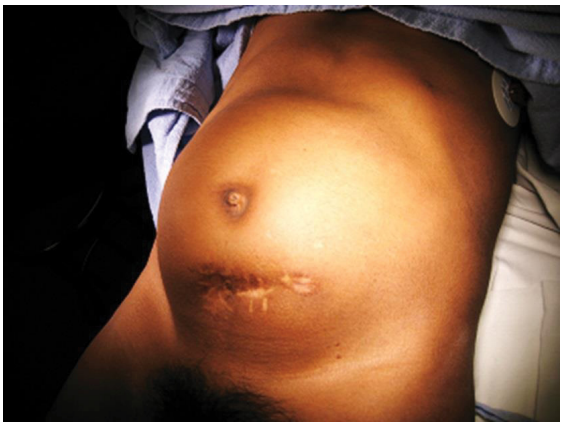

Figure 5: Meso and hypogastric macroscopic abdominal mass.

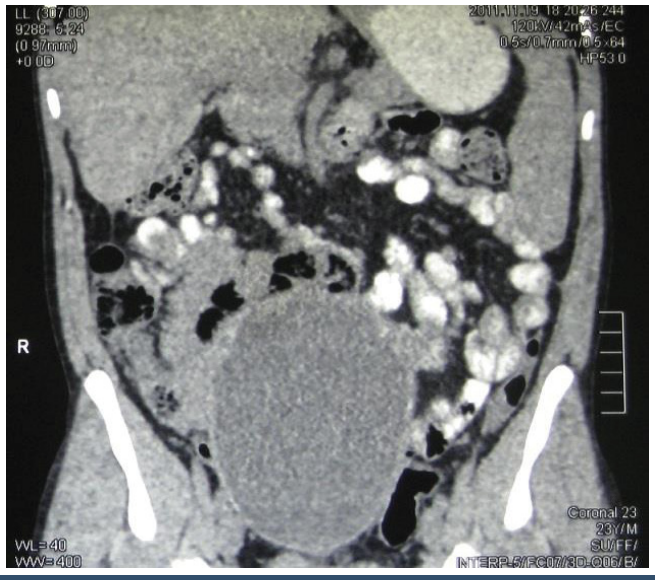

Figure 6: Abdominal´s tomography coronal view from the Figure 5.

Discussion

Long term CAPD diminishes solute transport, changes the vascularity and increases blood flow to the peritoneum, resulting in a conductance decrease that leads to progressive fibrosis [15]. This mode of renal replacement therapy is not exempt from morbidity. During CAPD the peritoneum is continually exposed to hyperosmotic fluids with high glucose concentration. The acidity of the fluids and the degradation products that are formed during CAPD favor a chronic inflammatory state, which modifies the peritoneum's immunologic response and predisposes to infections and membrane loss in the long term, even in the absence of episodes of peritonitis [4]. Efforts have been made to develop new biocompatible dialysis fluids [16] such as Dianeal™ (Baxter Healthcare, Deerfield, Il, USA), which increases pH resulting in an alkaline fluid and 7.5% Icodextrin (Extraneal™, Baxter Healthcare, Deerfield, Il, USA) which decreases the glucose degradation products; other fluid types contain amino acids in place of glucose. Other factors that influence peritoneum remodeling are peritonitis, uremia and the presence of a catheter. In addition, fibrosis and sclerosis are favored by the chronic release of proinflammatory mediators and cytokines such as: vascular endothelial growth factor, interleukin 1, 1-β, 2, 6 and 8, tumor necrosis factor, prostaglandin E2 and vascular and cell adhesion molecules [3, 17].

Morphologic changes observed include loss of mesothelial cells, submesothelial fibrosis, granulation tissue, macrophage infiltration and neoangiogenesis [18]. Uremia and chronic exposure to these molecules lead to changes in proteoglycan production and alteration of the extracellular matrix. Jiménez-Heffernan et al. [4], observed that myofibroblasts involved in the peritoneum fibrogenesis, originate from local fibroblasts and from transformation of mesothelial cells which acquire contractile ability and produce extracellular collagen, glycosaminoglycans and fibronectin matrix [19].

Furthermore, it has recently been observed that chronic dialysis promotes chronic inflammation and adiposity. Adipocytes affect the metabolism of mesothelial cells through the synthesis of a number of mediators including adipokines (leptin, resistin and other growth factors) and their receptors, forming an autocrine, paracrine and endocrine local web which contribute to systemic inflammation and decreasing survival [20,21].

After one year with peritoneal dialysis all patients develop peritoneal sclerosis, but only a few will have encapsulating peritoneal sclerosis (EPS). Even though it is a rare complication it carries a high morbidity and mortality. Some authors have observed a 20% death rate after 10 years with dialysis. In children the incidence is approximately 6.6% at 5 years and 22% after 10 years [22].

The clinical picture of EPS has a wide range of symptoms. We presented three cases which required an individualized diagnostic and therapeutic approach, from simple observation to direct intervention for sealing the sclerotic cavity. We identified four phases for the diagnosis and treatment: 1)

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clinical suspicion, 2) image confirmation, 3) microbiological work up with antibiotic therapy when necessary and 4) cavity collapse.

In the presented cases, clinical suspicion began with the visualization or palpation of an abdominal mass, followed by image confirmation to rule out a solid mass. Hence, the first study should be an abdominal ultrasound and later a tomography to define size, location and organ involvement [23]. In addition, EPS should be ruled out when there is evidence of ultrafiltration failure.

Diagnostic abdominal paracentesis is mandatory in order to obtain a culture sample to rule out infection. When culture is positive, the treatment must include specific antimicrobial therapy and repeated peritoneal lavages through the dialysis catheter with dialysis solutions until the culture is negative. In case 1, once the culture became negative, the cavity was collapsed with 2% polydocanol trough the dialysis catheter and external compression to avoid further fluid collection, without complications or adverse reactions.

The use of laparoscopy is helpful to determine the nature of the cavities, allowing the cleaning and curettage of the inner wall under direct vision to remove detritus that may complicate the resolution of infection, while minimally invading the patient [24].

Laparoscopy also aids in the differential diagnosis. Other diseases such as pseudomyxoma peritonei, peritoneal tuberculosis, mesothelioma and peritoneal carcinomatosis may show similar imaging features, but the distinction can be made with laparoscopic exploration, because of their characteristic visual appearances and the ability of performing a biopsy of the lesion [25].

When the diagnosis is made, the peritoneal dialysis must be suspended and the patient should be switch to hemodialysis. Nutritional support must be considered because of the risk of protein loss.

Treatment modalities include the administration of drugs to decrease the fibrotic process, which include prednisolone, sirolimus and everolimus [26] or renin-angiotensin blockers. Other drugs that have shown some benefit, especially in diabetic patients, are azathioprine, mycophenolate mofetil, acetyl cysteine, colchicine, thalidomide and rosiglitazone. Tamoxifen, an antifibrinolytic agent, has been used intraperitoneally with positive results [27].

Surgical treatment includes peritoneum cleavage, lysis of adhesions and intestinal resection when nonviable tissue is found. Reported mortality ranges from 24 to 53% in some series [28].

Conclusion

EPS is a rare complication in patients with ESRD who undergo peritoneal dialysis. It must be suspected in patients with evidence of ultrafiltration failure, especially if an abdominal mass is present. The early recognition and adequate treatment of this complication gives the opportunity to switch to other renal replacement therapies, especially renal transplantation. Our work suggests that the laparoscopic approach to this disease can be of substantial benefit and controlled clinical trials are warranted to validate this observation.

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