Encapsulating peritoneal sclerosis: case report

Esclerose peritoneal encapsulada: relato de caso

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Abstract

Patients with chronic kidney disease (CDK) can develop several diseases caused by the renal replacement therapy. Here we report a rare complication of peritoneal dialysis, the encapsulating peritoneal sclerosis (EPS) in which the peritoneal tissue is gradually replaced by fibrous tissue. The patient in question, after late loss of renal graft and conversion to peritoneal dialysis, evolved with multiple hospitalizations for spontaneous bacterial infections, in recent admission, he was diagnosed with sub-occlusive abdomen secondary to the EPS. Five days after, presented with intestinal obstruction requiring surgical approach by laparotomy, being performed with right colectomy, enterectomy, enteroraphy and ileostomy with drainage. The patient progressed well and follows on prednisone and tamoxifen-associated with intermittent hemodialysis.

Keywords: peritoneal dialysis; peritoneal fibrosis; renal insufficiency, chronic.

Resumo

No contexto da insuficiência renal crônica (IRC), os pacientes estão sujeitos a diversas patologias advindas da própria terapêutica de substituição renal. Relatamos aqui uma complicação rara da diálise peritoneal, a peritonite esclerosante encapsulante (PEE), na qual o tecido peritoneal é progressivamente substituído por tecido fibroso. O paciente em questão, após perda tardia de enxerto renal e conversão para terapêutica dialítica peritoneal evoluiu com múltiplas internações por infecções bacterianas espontâneas, em último internamento, foi diagnosticado com abdome sub-oclusivo secundário à PEE. Após 5 dias apresentou quadro de abdome obstrutivo com necessidade de abordagem cirúrgica por laparotomia exploradora, sendo realizado colectomia direita, enterectomia, enterorrafia e ileostomia com drenagem. O paciente evolui bem e segue em tratamento com prednisona e tamoxifeno associado à hemodiálise intermitente.

Palavras-chave: diálise peritoneal; fibrose peritoneal; insuficiência renal crônica.

Introduction

Encapsulating peritoneal sclerosis (EPS) is a rare complication with high morbidity and mortality secondary to the peritoneal dialysis (PD). The incidence of this complication in PD varies among studies, but they all report low levels, ranging between 0.7% to 3.3%.[1-3] However, despite the small number of cases, approximately 5 of 10 of these patients die as a result, directly or indirectly, of the EPS.[1]

According to the Brazilian Census of Dialysis in 2014, by the Brazilian Society of Nephrology, it was estimated that from the 100,000 dialysis patients per year in Brazil, 8.7% of these patients are on peritoneal dialysis, more often in the automated peritoneal dialysis modality (5.6%).[4] Worldwide, approximately 196,000 patients are on peritoneal dialysis, representing about 11% of the entire population on renal replacement therapy (RRT), with a tendency to increase in developing countries.[5]

The EPS was first described in 1907 by Owtschinnikow[6] and related to PD only in 1978 by Gandhi *et al.*[7] In its final stage, the syndrome is marked by repeated intestinal obstruction, which is clinically characterized by nausea, vomiting, bloating, anorexia, weight loss and eventually ascites. Macroscopically the bowel is involved with a coarse fibrous tissue capsule.

Radiographic findings include thickening associated with peritoneal calcification, with points of obstruction and dilatation of the bowel.[8] Because the symptoms of EPS are nonspecific, the diagnosis in

Submitted on: 03/07/2017.
Approved on: 03/20/2017.

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DOI: 10.5935/0101-2800.20170083

most cases is slow and usually happens after discontinuation of PD.[2]

Therefore, because of the rarity, high mortality and difficulty in diagnostic investigation, we describe a case of encapsulating peritoneal sclerosis in the a patient with chronic kidney disease under dialysis with a brief review of the literature about the pathophysiology involved in the development of this disease, its risk factors, diagnostic methods and treatment possibilities.

Case report

Patient with 45 years, male, caucasian, with a prior diagnosis of a longstanding hypertension, heart failure, chronic kidney disease for 16 years, renal transplant of a living donor for 15 years, which has evolved with graft dysfunction and graft loss after 3 years, in continuous ambulatory PD for 12 years with multiple hospitalizations for bacterial peritonitis. In all the course, hyperosmolar glucose solution was used in the PD. One year ago the patient needed to convert the PD to hemodialysis.

He was admitted to hospital in February 2016 by distension and diffuse abdominal pain, low food intake, nausea and vomiting for 15 days. He remained afebrile during evolution and also reported weight loss of about 20 kilograms in the past year.

At admission, he was in good condition, lucid and oriented in time and space, eupneic, afebrile, with semi-globose distended abdomen, bowel sounds were present, painful to superficial and deep palpation without peritoneal irritation signs.

During the hospitalization period, it was performed abdominal CT scan for investigation, showing diffuse extensive vascular calcification, bowel distension in the mesogastrium with some fluid level images. Blurring of the outlines of the handles suggested that is attached to the anterior abdominal wall (Figure 1).

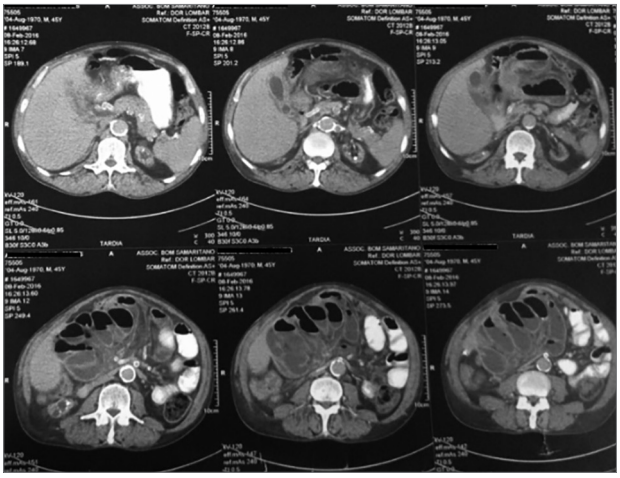
Without surgical indication at the time, he was submitted to clinical treatment of sub-occlusive acute abdomen secondary to the EPS with fasting and open nasogastric tube, total parenteral nutrition, prednisone and tamoxifen. After 5 days, evolves with obstructive acute abdomen requiring laparotomy, being conducted right colectomy, enterectomy, enteroraphy and ileostomy with drainage.

The patient progressed well and follows on prednisone and tamoxifen. Kept on intermittent hemodialysis.

DISCUSSION

The peritoneal sclerosis is an inflammatory process in which the peritoneum is diffusely transformed into fibrous tissue. The clinical picture is variable, the EPS is its most severe presentation. In EPS, the gut is surrounded by a dense fibrous layer with blockage of peristalsis, leading to sub-occlusive or occlusive abdominal frame.[9]

Figure 1. CT of abdomen showing extensive vascular calcification, distension of intestinal loops in the mesogastrium with air-fluid levels.



The pathophysiology is still poorly understood, but some risk factors such as the use of more than five years of PD, a high concentration of glucose in the dialysis solute, repeated peritonitis, which causes structural and functional damage to the membrane of the peritoneum are associated factors well documented in the literature.

Among all these, the duration of PD is a single important factor, occurring almost exclusively on PD of patients with 5 years or more of therapy, and the prevalence of this condition in therapies of 2 years or lower is very low.[10] An important observational study that included 4290 PD patients observed from 1981 to 2002, had only 34 patients who developed EPS, less than 1% of cases. The average age of affected was 44.5 years, 68% were on PD for over than 5 years and 79% with a prior history of peritonitis.[11]

A multi-center, randomized, prospective study from Hong Kong selected 80 patients with ambulatory PD from 2006 to 2008 and randomized to a low glucose solution and conventional glucose-based solution for PD. The data from the study showed increased levels of adiponectin and anti-fibrotic markers in the low glucose group, with a possibly a delay or reduction of peritoneal sclerosis.[12]

The cases presented corroborates with the data cited above on the correlation of the development of EPS with time on PD and the type of fluid used, occurring 12 years after the onset of PD, including other associated factors such as the repetitive peritonitis and the age of the patient.[12]

The diagnosis of EPS is made by combining clinical and radiological findings, and should be suspected in patients with long-standing PD evolving with

sub-occlusion or intestinal occlusion. Signs such as nausea, vomiting and weight loss are non-specific but suggest EPS, which makes diagnosis difficult, considering the rarity of this condition.[10] Patients undergoing kidney transplantation are possible patients at risk of developing the disease, despite the very low prevalence, the absolute mortality rate was zero.[13]

Radiological findings were recently described and evaluated. The use of plain abdominal radiography, as easy as its access may be, has lower sensitivity and specificity than computed tomography (CT) for the diagnosis. Ultrasound has its clinical value to prevent patient exposure to ionizing radiation and can also identify other conditions such as the presence of collections in abdominal or intra-abdominal wall, peritoneal thickening, peritoneal calcification, and loops of small thickened and dilated bowel, but It has limitations such as its operator dependency.[14]

However, CT remains the best diagnostic tool, which can detect early cases that would be hardly viewed on radiographs or ultrasound. Belatedly, there is an encapsulation of the wall of the small intestine by peritoneal fibrosis leading to a thickening and contraction of the mesentery, which includes the intestinal loops in the center.[14]

The use of magnetic resonance imaging (RMI) as well as being more costly and with more difficult access in some centers, has a smaller resoluteness then CT and still an uncertain risk of nephrogenic systemic fibrosis induced by the use of gadolinium. Therefore it is not routinely indicated.[14]

About the treatment, there is little evidence for pharmacological therapy such as corticosteroids, tamoxifen and immunosupression while compared to the surgical treatment of EPS.[15] Although, little experience with the pathology and the inability to indicate a specific treatment, it is suggested that the early surgery associated with corticosteroid treatment may improve the prognosis[16] while another cohort study shows that the success of surgery in this disease is determined by the technique used. The technique used is implied to free adhesions and extirpate the capsule as much as possible. Drilling, resection and intestinal anastomosis, significantly increased mortality.[17]

Conclusion

The EPS is a rare complication of PD but has high mortality. The pathophysiology of EPS remains uncertain. The radiological diagnosis, especially CT, should be used and mainly applied in patients with a long-standing of peritoneal dialysis and development of sub-occlusive and occlusive intestinal symptoms. Besides the lack of data to guide the clinical treatment with corticosteroids, immunosuppressants and tamoxifen or surgical treatment, with enterolysis and peritonectomy, the actual treatment is based on low-level evidence studies.

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