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# Sclerosing Encapsulated Peritonitis: A devastating and infrequent disease complicating kidney transplantation, case report and literature review

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ABSTRACT

*INTRODUCTION:* Sclerosing Encapsulating Peritonitis (SEP) is a rare condition with an incidence of up to 3% and a mortality of up to 51% among peritoneal dialysis (PD) patients (Brown et al., Korte et al. and Kawanishi et al.). In the last ten years, the incidence of SEP in kidney transplant recipients has increased (Nakamoto, de Sousa et al. and Korte et al.).
*PRESENTATION OF CASE:* A 31-year old male with a 15 years history of PD and later kidney retransplantation was admitted to the emergency service after experiencing several weeks of diffuse abdominal pain which had escalated to include vomiting and diarrhea during the 24 h previous to admission.

The patient underwent an exploratory laparotomy where severe peritoneal thickening was found, in addition to signs of chronic inflammation and blocked intestinal loops. Histopathologic findings were suggestive of sclerosing peritonitis. After two months of treatment in hospital, the patient presented an obstructed intestine, with a rigid and thickened peritoneum compromising all the intestinal loops.
*DISCUSSION:* Despite being rare, SEP, represents a significant complication due to its high mortality and recurrence. It is insidious in its early stages and culminates in an intestinal obstruction (Fieren). Risk factors for its development in kidney transplant recipients include a history of prolonged treatment with PD and the use of calcineurin inhibitors as an immunosuppressive treatment (Korte et al.).
*CONCLUSION:* Given the increase in the incidence of SEP in kidney transplant recipients, the clinician should be alert to the presence of this complication. A greater number of multi-centre studies are required to identify the risk factors for SEP that are inherent in renal transplant recipients.


## 1. Introduction

SEP is an uncommon complication of PD with an incidence of 0.7-3.3% and a mortality approaching 51% in the first year [1–3]. It is characterized by a diffuse fibrosis of the peritoneal membrane with an encapsulation of the intestinal loops, causing intestinal obstruction [4]. Although its etiology continues to be non-specific, it is believed that it originates from an injured peritoneal membrane, which, after a second inflammatory stimulus, such as peritonitis, hemoperitoneum or surgery, SEP is triggered [5].

Few cases have been reported involving kidney transplanted patients [6,7], and given the rare nature of this pathology, we decided to publish this case report in line with the SCARE criteria [8] and with emphasis on a review of the literature. To our knowledge, this is the first case report of SEP in a kidney retransplanted patient.

## 2. Presentation of case

A 31-year-old male patient, with Alport Syndrome, diagnosed at the age of 9, with kidney disease and hypertension who received a kidney transplant at ten years old, from a related living donor. Ten years after the kidney transplant, the patient presented an episode of rejection, and lost the kidney graft; for which he received PD for

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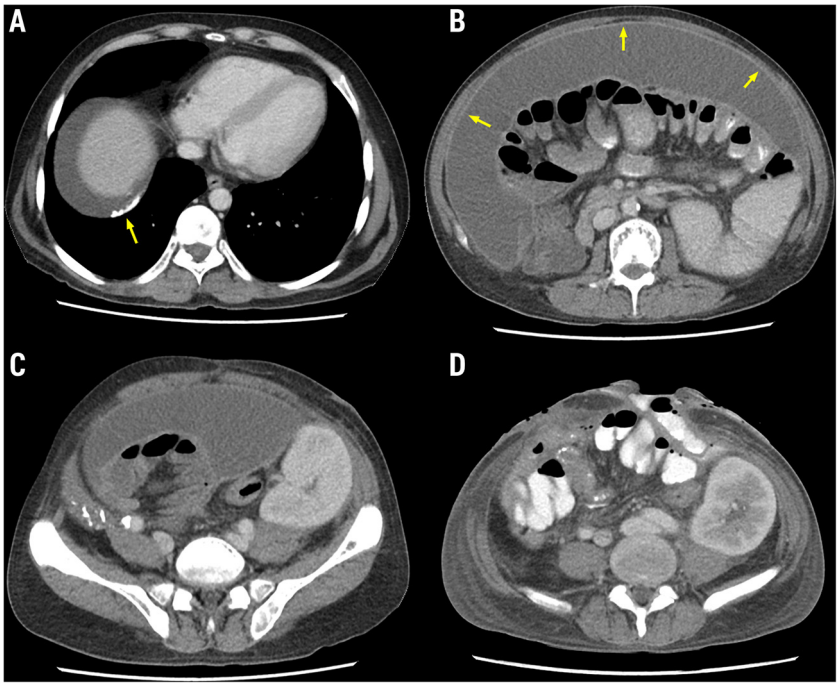


**Fig. 1.** Axial view of an abdominal enhanced CT. Panel A, right subdiaphragmatic calcified peritoneal plaque. Panel B, progression in the thickening of the peritoneum with contrast enhancement, which englobes the intestinal loops and conditions a discrete small bowel distention with the presence of air-fluid levels, compatible with an initial obstructive process. Panel C, inferior view of the pelvis, with left kidney graft without alteration and atrophic changes of the first right kidney graft. Panel D, postsurgical changes of parietal peritoneum resection with an abdominal wall defect and oral contrast leak compatible with enterocutaneous fistula.

**Table 1**
Laboratory test results taken in admission.

| | |
|---|---|
| Glycemia (mg/dL) | 92.8 |
| Blood Urea Nitrogen (mg/dL) | 26.7 |
| Serum Creatinine (mg/dL) | 1,28 |
| Alanine Aminotransferase | 6 |
| Aspartate Aminotransferase | 9,8 |
| Serum Sodium (mEq/L) | 138 |
| Serum Potassium (mEq/L) | 4,9 |
| Serum Chloride (mEq/L) | 97,4 |
| Serum Calcium (mEq/L) | 10 |
| Serum Phosphorus (mEq/L) | 3,83 |
| Serum Lipase | 28,7 |
| Blood Count | |
| Leukocytes | 12910 |
| Neutrophils (%) | 84% |
| Lymphocytes (%) | 3,40% |
| Hemoglobin (g/dL) | 6,9 |
| Hematocrit (%) | 24 |
| Platelets | 589000 |

15 years until a deceased kidney transplant donor was found, with 1dr-1a compatibility.

One year after the kidney re-transplant, the patient was admitted after experiencing several days of crampy abdominal pain localized in the epigastrium and mesogastrium, associated with vomiting bile and diarrheic bowel movements. At the moment he was receiving immunosuppressive management with Tacrolimus 7 mg/day, Mycophenolate Sodium 1080 mg/day and prednisolone 5 mg/day.

Physical examination revealed a globose abdomen, painful upon deep palpation, with no signs of peritoneal irritation and with no pain in the area of the transplant. Paraclinicals were taken upon admission (Table 1) and due to suspicion of an intra-abdominal infection, an enhanced abdominal Computed Tomography (CT)) (Fig. 1A and B) was taken which showed evidence of intra-abdominal collections in the right paracolic gutter. These findings

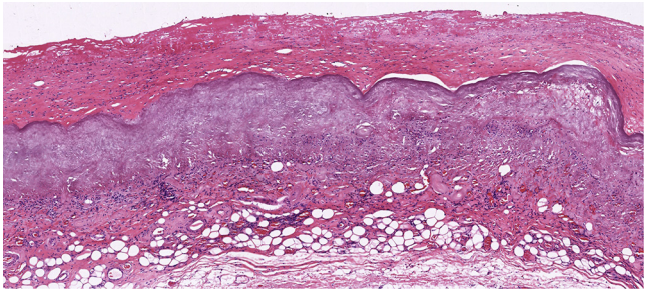

**Fig. 2.** Colouring of H&E at 200X. In the histological section, there is mesothelial replacement due to a proliferation of fibroblasts and capillaries, accompanied by a chronic and acute inflammatory response with exudation. Subsequently, an acellular layer with calcium deposits is formed and ends with a layer of fibrin.

were suggestive of peritonitis, with indirect signs of intestinal obstruction due to adhesions. It was therefore decided that an exploratory laparotomy should be conducted, which found three liters of cloudy brown liquid, severe thickening of the parietal peritoneum with chronic inflammation, abundant membranes and blocked loops; drainage of the peritonitis was performed, enabling partial release of the adhesions, and a biopsy was taken off the peritoneum. Subsequently, the pathology report revealed chronic peritonitis with histologic findings suggestive of a sclerosing peritonitis (Fig. 2).

Two months later, while still hospitalised, the patient developed a clinical picture of intestinal obstruction and underwent a laparotomy which showed a 100% blocked abdomen, a thickened and stoned peritoneum, compromising the coating of all the intraabdominal organs, with multiple collections. Associated with fibrinopurulent membranes and intestinal obstruction at the level of the distal ileum, involving multiple segments (Figs. 3 and 4). He required an Ileostomy but due to its high output, compromising the patient's hydroelectric balance and nutrition, it was later closed

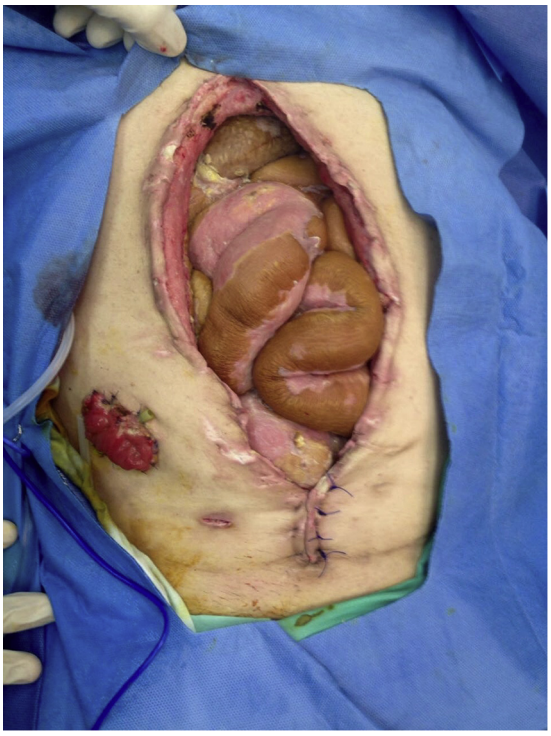

**Fig. 3.** Observed surgical image of stoniness and thickened peritoneum.

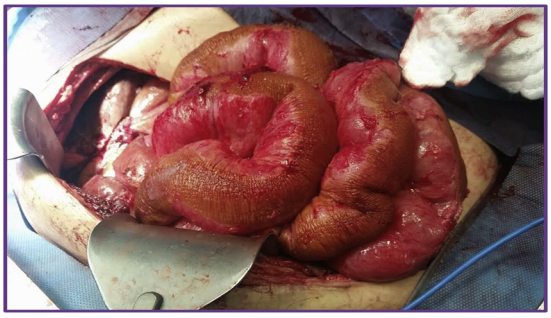

**Fig. 4.** Observed surgical image of stoniness and thickened peritoneum.

with subsequent fistulas originating from the enterorraphy (Fig. 1C and D).

The patient remained hospitalized with a torpid evolution, with the continuing presence of intraabdominal collections evidenced via images taken over subsequent days. These were percutaneously drained, and multiple revisions of the abdominal cavity were undertaken with the need for enterorraphy in the distal ileum. Despite intense antibiotic treatment and drainage of intrabdominal collections, the patient developed septic shock and died.

## 3. Discussion

"Chronic fibrous encapsulating peritonitis" was first described in 1907, a term that encapsulated alterations and macroscopic findings, in the peritoneum causing intestinal obstruction. In 1980, a relationship was established between SEP and continuous peritoneal dialysis [9].

Its etiology is still unknown, although an association with a history of abdominal surgery and prolonged treatment with PD has been demonstrated [1,3,10]. Specifically, chronic exposure to PD solutions containing glucose degradation products (GDP) is thought to be the leading cause of peritoneal injury [11,12], given its acidic nature and its high non-physiological osmolality which contributes to damage of the peritoneal membrane [13]. Leading the search for new biocompatible PD solutions, which contain a lower amount of GDP, to preserve the peritoneal membrane and enhance the viability of the mesothelial cells, contributing to a lower incidence of SEP [14]. Not all patients with PD develop SEP, but it has been observed, as in our case, that the disease may manifest subsequently to PD suggesting that a second stimulus must act as a triggering factor [15].

Some case reports have demonstrated an increase in the incidence of SEP in kidney transplant patients, suggesting the presence of a risk factor different from the prolonged use of PD [6,7]. Among the suggested possible risk factors is the use of calcineurin inhibitors, such as Tacrolimus and cyclosporine, which are capable of increasing the expression of transforming growth factor β (TGF-Î²) and which are, therefore, capable of inducing an increase in peritoneal fibrosis [16].

The clinical presentation of SEP in the early stages is characterized by insidious symptoms of hyporexia, nausea, and vomiting, with intermittent periods. In later stages, as can be seen in our patient, constipation, abdominal mass and abdominal pain may appear [5].

SEP is characterized histologically by a loss of mesothelium along with a significant thickening of the peritoneal membrane with dense layers of fibro-connective tissue; inflammatory infiltrates, increases in fibrin, exudates, ossification, calcification and interstitial fibrosis [5]. These can be seen in the biopsy obtained from our patient (Fig. 1).

The best imaging method is computerized axial tomography, which can show loop dilation, loculated ascites, intestinal calcifications, peritoneal and intestinal thickening and intestinal encapsulation [5] as can be seen in our case report (Fig. 1). However, in some cases, a diagnosis can only be obtained via laparoscopy [3].

Surgical treatment of SEP is highly complex, since the behavior of SEP consists of an adhesive process throughout the small intestine, and the goal of therapy is to release the gut. In the majority of cases, given the absence of a plane of dissection, there is a risk of multiple perforations [3,17], as seen in our case (Figs. 3 and 4). Adhesiolysis can reverse the intestinal obstruction but does not stop the deterioration process in the peritoneum, which continues progressing with a high probability of recurrence of symptoms in 6–12 months [3]. The use of Tamoxifen has been studied, due to its effect as a selective inhibitor of estrogen receptors and their ability to inhibit the production of TGF-β by fibroblasts. Its use has been described in alone or in conjunction with corticosteroids with satisfactory results regarding a decline in mortality in the early stages of the disease [5].

To our knowledge this is the first case report of SEP in a kidney re-transplanted patient, and although management did not vary from those described in the literature, we believe our case report alerts transplant centers and contributes in the search of risk factor among potential kidney graft receptors that will culminate in SEP.

## 4. Conclusion

Sclerosing Encapsulating Peritonitis is a rare but critical condition that mainly affects patients who have been exposed to PD for extended periods of time. An increase in its incidence has been noted in kidney transplant patients, given their history of PD, and possibly, also, due to the growing use of calcineurin inhibitors as an immunosuppressive treatment. To our knowledge, this is the first report of SEP in a kidney transplanted patient in Colombia. Further multicentre studies are required to establish the risk factors associated with this population group.

## Conflicts of interest

The authors declare that there are no potential conflicts of interest to declare.

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## Ethical approval

Consent from the patient was secured for this case report and is available upon request.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

## Author contributions

Liliana Caicedo; Alejandro Delgado; Luis A. Caicedo; Gabriel J. Echeverri: Study concept and design.

Alejandro Delgado; Laura S. Thomas, Liliana Caicedo; Juan Carlos Bravo; Martin Rengifo: Adquisition of data.

Juan Carlos Bravo; Martin Rengifo: Drafting of the manuscript and edition.

Alejandro Delgado; Liliana Caicedo; Gabriel J. Echeverri; Oscar Serrano, Jorge I. Villegas, Gabriel J. Echeverri, Luis A. Caicedo: Critical Revision of the manuscript for important intelectual content.

## Registration of research studies

Does not apply, since it is not a “first to man” case report.

## Guarantor

The guarantor for this manuscript is Gabriel J. Echeverri.

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