

Peritoneal sclerosis and massive hemoperitoneum: case report and short review

Abstract

Secondary Peritoneal Sclerosis has been reported in several cases but is especially frequent among chronic peritoneal dialysis users, being its most serious complication. Clinical suspicion in chronic PD users is no challenge as intestinal symptoms and hypoalbuminemia appear and radiological confirmation is usually achieved before the need for surgery and intra abdominal findings prove confirmatory. A case report of a 37-year-old male patient with a 13 year long peritoneal dialysis in whom laparotomy findings were a massive hemoperitoneum, parietal/visceral peritoneum, small/large bowel and mesentery with chronic inflammatory changes, thickening and dark-brown coloration. As a distinctive feature a gastroepiploic artery branch in the gastric curvature was identified with persistent oozing and hemostasis was achieved. No intestinal obstruction was evident. Postoperative was uneventful. In patients undergoing peritoneal dialysis a hemorrhagic effluent from the catheter or elevated levels of inflammatory mediators should demand a thorough investigation.

Keywords: abdominal cocoon syndrome, peritonitis chronica fibrosa incapsulata, etiology, bacterial, eosinophilic, meconium, renal transplantation, Liver transplant, laparotomy, Thecomas

Volume 7 Issue 3 - 2019

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Received: June 30, 2019 | **Published:** July 18, 2019

Abbreviations: EPS, encapsulating peritoneal sclerosis; PS, peritoneal sclerosis; US, abdominal ultrasound; TGF-beta, transforming growth factor beta;

Introduction

Mainly due to the multiple possible primary and secondary etiologies Sclerosing Peritonitis has been referred to in different terms. These include Abdominal Cocoon syndrome, "peritonitis chronica fibrosa incapsulata" initially coined in 1907 and Encapsulating Peritoneal Sclerosis (EPS).^{1,2} Looking for a wider term that can group several etiologies and stages, Peritoneal Sclerosis (PS) has been suggested and will be appropriated for this review.³ The various terms, difficulties in diagnosis and heterogeneity of findings according to the stage in which it is described have made its study an ongoing project. Differentiation from peritoneal encapsulation is a must since this is a developmental condition that differs widely in pathogenesis/management and will not be addressed herein. Primary etiology refers to an idiopathic process with no diagnosis yet made (probably in the future understanding of the process will reduce this classification). Secondary SP has been reported in several cases but is especially frequent among chronic peritoneal dialysis users. Other causes include Tuberculosis, Malignancy (Neuroendocrine tumours, Thecomas, Ruptured tumours like GIST or dermoids), Post-operative (Renal transplantation, Liver transplant, laparotomy, use of Povidone iodine), Drugs (Beta-blockers, Ergot), Generalized peritonitis (bacterial, eosinophilic, meconium).⁴

Pathophysiology and classification

Basic pathophysiology of SP stands on chronic irritation of the peritoneum and intra abdominal viscera. Peritoneal stromal and mesothelial inflammation ultimately lead to peritoneal sclerosis or fibrosis and cocoon formation. Both chemical or infectious agents disrupt the mesothelial cell junction with destruction of subserosal

tissue, a process that ends in fibrosis.⁵ Recurrent peritonitis might explain the pathology findings in a "two hit" theory in which the first "hit" will be the irritating agent and subsequent inflammation and chronic peritonitis will represent the second "hit".⁶

Classification has been established into type I, II and III. For types I and II according to membrane involvement of part of, the whole of the small intestine or the whole of the small bowel and other organs, respectively.⁷ Several histologic criteria have been proposed for diagnosis after a judicious evaluation of a large series of cases and include fibrin deposition, fibroblast swelling (enlargement), capillary angiogenesis, mononuclear cell infiltration and the presence of several immunohistochemical markers for peritoneal fibroblast activation and proliferation.⁶ Classification in the clinical spectrum has not always been available and patients can dwell into and out of subtypes. Defined in Japan in response to the rise in cases after introduction of PD four subtypes were (I) Pre-EPS with only ascites, (II) Inflammatory stage with nausea/diarrhea, mild encapsulation and intestinal swelling, (III) Encapsulating stage with ileus, mild to severe inflammatory symptoms, encapsulation and adhesions, (IV) Chronic stage with prolonged ileus, mild inflammation, but encapsulation and loop shrinkage.⁸

Clinical presentation and diagnosis

Presentation varies according to each clinical stage with initial stages having ultrafiltration failure and solute transport, while in the final stages intestinal symptoms prevail, especially intestinal obstruction, weight loss and formation of cocoon. Hemoperitoneum has been reported in stages 1 and 2 in 7 to 50% of patients. Clinical suspicion in chronic PD users is no challenge as intestinal symptoms and hypoalbuminemia appear and radiological confirmation is usually achieved before the need for surgery and intra abdominal findings prove confirmatory. This is not the case for other etiologies as diagnosis can be elusive and other causes may have to be ruled out. In

patients undergoing peritoneal dialysis an hemorrhagic effluent from the PD catheter or elevated levels of inflammatory mediators should demand a thorough investigation.⁹

As imaging studies have become more accurate, different findings have been described that guide the clinician in the evaluation of SP, but pathognomonic findings are yet to be described. Plain abdominal radiography usually presents with diffuse peritoneal calcifications¹³ observed as diffuse radiopaque enhancements of intestinal loops occasionally associated with patterns of dilation and air-fluid levels in bowel obstruction.^{4,10} Barium contrast studies demonstrate a conglomeration of bowel loops at the midline level with enhancements of the intestinal wall due to peritoneal diffuse calcifications, associated with slowing of transit without evidence of endoluminal lesions or mucosa. A particular sign described as in *accordion pattern* has been described,^{4,10,12} although no specificity has been reported.

Abdominal Ultrasound (US) has often identified segmental dilation of intestinal loops encapsulated by a dense fibrous membrane, abnormal pattern of peristalsis, loculated free fluid and thickening of the intestinal walls, described in some cases as trilaminar thickening, however these findings are limited to the expertise and technique of the radiologist.⁴ There is no clinical evidence to support the sensitivity and specificity of each of the findings, although it is worth highlighting the usefulness in the evaluation of patients with sclerosing peritonitis given the easy access and low cost of ultrasonography.^{4,10,12} CT has become most useful with characteristic signs given by dense encapsulation at the midline level with segmental involvement of intestinal loops without presence of contrast or peripheral enhancement, collections or loculated ascites, as well as calcifications and diffuse peritoneal thickening. With these findings, a classification system has been proposed in which, having three of the present findings, the sensitivity and specificity can be close to 100% and 94%, respectively.¹⁰ It is especially useful in patients presenting with signs/symptoms of intestinal obstruction and as part of pre-surgical planning in which it has demonstrated a decrease in the rates of unnecessary intestinal resections.^{12,14} MRI has been proposed as an alternative, with similar findings to CT, however until now there are no studies that validate its diagnostic usefulness, nor comparative studies against the standard evaluation study.¹⁰

Treatment

So far there is no consensus on the optimal management of sclerosing peritonitis because its pathophysiology is not clearly elucidated, therefore the proposed treatments are directed against possible pathways of physiopathology. Medical management has been proposed with use of compounds that block and control the inflammatory cascade: antifibrotic, anti-inflammatory and immunomodulatory molecules.^{4,10,15} Corticosteroids are one of the medications up to now supported in different case reports, based on their ability to inhibit the inflammatory response and prevent the formation and maturation of the collagen present in SP.^{4,10} Tamoxifen is part of the group of antifibrotic drugs that, being a selective inhibitor of estrogen receptors, blocks the production of transforming growth factor beta (TGF-beta) by fibroblasts. A retrospective Dutch study in patients with peritoneal dialysis in which SP has developed showed that the use of tamoxifen reduces mortality.¹⁶ Colchicine is another of the proposed drugs since it blocks the mRNA of fibroblast TGF-beta which results in decreased formation of fibroid tissue.¹⁰

The natural course of the disease may involve recurrent episodes of

intestinal obstruction, with prior clinical loss of appetite, weight loss and malnutrition. It has been shown that in cases of mild symptoms, patients have a better prognosis and response to conservative than surgical management, where nutritional repletion plays a fundamental role in the prevention of postoperative complications. Nutritional repletion should preferably be enteral, however some patients might benefit from parenteral route, taking into account its associated complications.⁴

Surgical treatment is based on adhesiolysis proposed as the gold standard in patients with severe symptoms or recurrent episodes of intestinal obstruction. In patients in whom a complete adhesiolysis of fibrous tissue is not achieved, the concomitant use of anti-inflammatory and corticosteroids is proposed, however there is no clinical evidence demonstrating the postoperative benefit in the reduction of symptomatology or recurrence.¹⁰ The recommendation with high clinical evidence that has been determined so far is to avoid resections and anastomosis given the high rate of leaks, increasing morbidity and mortality.⁴ Complications related with surgical management are early intestinal obstruction secondary to extensive manipulation, edema of the intestinal wall and prolonged operative time, being greater in the first 30 days postoperatively.⁴ Laparoscopic approach is not currently recommended due to the high risk of intestinal perforation secondary to the insertion of trocars in the context of the adhesion syndrome and dilation of loops, with current clinical evidence being scarce.⁴

Associated bleeding/massive hemoperitoneum

In the context of limited clinical evidence, we present the first reported case of SP with clinical presentation of hemoperitoneum without intestinal obstruction, this case report contributes to the clinical evidence on the different forms of presentation of sclerosing peritonitis.

Case report

We present the case of a 37-year-old male patient with a history of arterial hypertension and end-stage renal disease in PD the thirteen previous years. At the ED he presented with bleeding through the peritoneal dialysis catheter after valsalva maneuvers due to sudden vomiting. Initial assessment revealed hypotension, tachycardia and pallor, without mental status alterations and a referred abdominal pain in the mesogastrium and epigastrium without peritoneal irritation. Initial blood workup showed mild leukocytosis and neutrophilia, anemia, high azotees, no electrolyte imbalance and a normal abdominal ultrasound Figure 1. Subsequent follow up anemization and abdominal CT findings compatible with perihepatic, perisplenic and pelvic fluid associated with hyperdense peripheral enhancement of bowels and mesentery/peritoneum urged surgical intervention. Laparotomy findings were a massive hemoperitoneum with no initial clear bleeding site, parietal/visceral peritoneum, small/large bowel and mesentery with chronic inflammatory changes, thickening and dark-brown coloration. After a thorough exploration a gastroepiploic artery branch in the gastric curvature presented persistent oozing with appropriate management. No intestinal obstruction was evident. Postoperative evolution was favourable, with renal replacement therapy by hemodialysis initiated without setbacks. A post-operative RBC transfusion was necessary with subsequent stabilization of hemoglobin, modulation of inflammatory response, decrease in leukocytosis, neutrophilia and CRP. Blood cultures, peritoneal fluid cultures and a negative adenosine deaminase in peritoneal fluid. Two weeks follow up was performed with no early surgical complications.

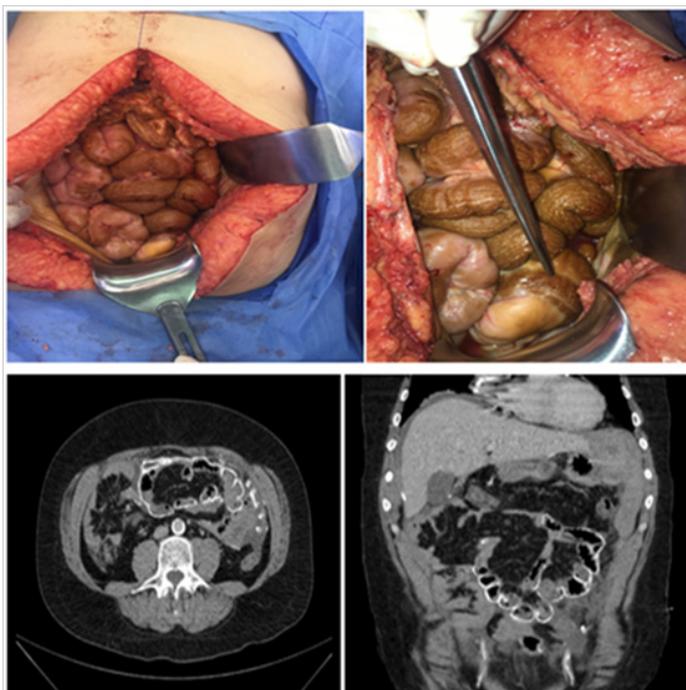


Figure 1 CT Scan images.

Acknowledgements

None.

Conflicts of interest

The author declares there is no conflict of interest.

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