East African Scholars Journal of Medicine and Surgery
Abbreviated Key Title: EAS J Med Surg
ISSN: 2663-1857 (Print) & ISSN: 2663-7332 (Online)
Published By East African Scholars Publisher, Kenya

Volume-5 | Issue-7 | Aug-2023 | DOI: 10.36349/easjms.2023.v05i07.001

OPEN ACCESS

Case Report

Idiopathic Sclerosing Encapsulating Peritonitis: A Case Reports

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Article History
Received: 18.06.2023
Accepted: 25.07.2023
Published: 02.08.2023

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: Sclerosing encapsulating peritonitis is a disease characterized by a chronicle inflammatory process in which part or all of the digestive tract is enveloped by a dense fibro collagen membrane. The clinical picture is not very telling and nonspecific, however imaging, in particular injected computed tomography, easily allows the diagnosis, but the idiopathic form remains very rare. In this article, we report the case of a 44-years-old man with no particular history, who presents to the emergency room for stopping materials and gases, an abdomino-pelvic calculated tomography injected confirms the diagnosis of an intestinal obstruction consecutive to idiopathic sclerosing encapsulating peritonitis.

Keywords: Encapsulating peritonitis, idiopathic, diagnosis, surgery, a case report.

INTRODUCTION

Encapsulating peritonitis is a chronic attack of the visceral peritoneum leading to the formation of a thick fibrous membrane, sheathing the digestive tract either by leaving it free, or by wrapping it in a bag like a cocoon [Tagnaouti M *et al.*, 2009; Dutranoy JC *et al.*, 2009].

In 1907, Owtschinnikow PJ [PYa Ovchinnikov,1907] described sclerosing encapsulating peritonitis "Peritonitis chronica fibrosis incapsulata", followed in 1978 by Foo KT who described the primary forms "the abdominal cocoon" [K.T. Foo *et al.*,1978] and the secondary forms.

We report the case of a 44-years-old young man with no particular history in whom the diagnosis of encapsulating peritonitis was revealed by an occlusive syndrome and we discuss the clinical, etiopathogenic and therapeutic aspects of this affection.

CASE REPORT

44-years-old man, with no particular medical history, operated on for appendicitis at the age of 20, who consults for vomiting, cessation of substances and gases with the notion of diffuse abdominal pain. The history of the disease seems to go back 20 days, marked by scattered episodes of early post-prandial vomiting on a background of diffuse abdominal pain relieved by self-medication.

Faced with the exacerbation of vomiting and the cessation of materials and gases, the patient presents to the emergency room.

The clinical examination found a patient in good general condition, afebrile, with an appendectomy scar at the level of the right iliac fossa, palpation revealed at the level of the left flank an abdominal mass extending over 7cm with a tender abdomen as a whole, digital rectal examination noted an empty rectal ampulla, the hernial orifices were free, the rest of the examination being unremarkable.

Before this clinical examination, a pre-therapeutic and iconographic assessment was requested.

Abdominal X-rays without preparation objective an air-fluid level of the small intestine type and the injected abdomino-pelvic computed tomography (Figure 1) reports the presence of a distension of the 2nd duodenum (55mm) upstream of a disparity of abrupt caliber projecting at the level of the 3rd duodenum and downstream a cluster of jejunal loops grouped at the level of the left flank and the umbilical region cer born by a thin membrane slightly enhanced by the contrast product surrounded by a colonic frame without anomaly (Figure 2) .

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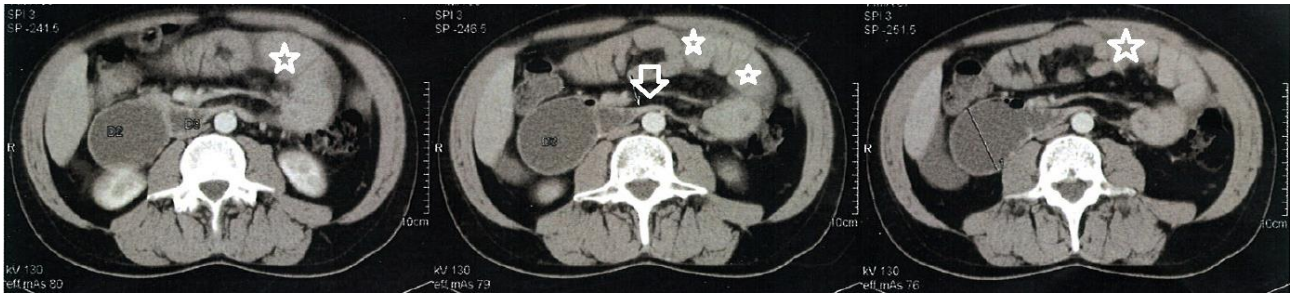

Figure 1: axial sections of the CT scan of our patient, acquisition after digestive marking and after injection of iodinated contrast product; we demonstrate the presence of a pseudo sac made of agglutinate of hail loops (white star).

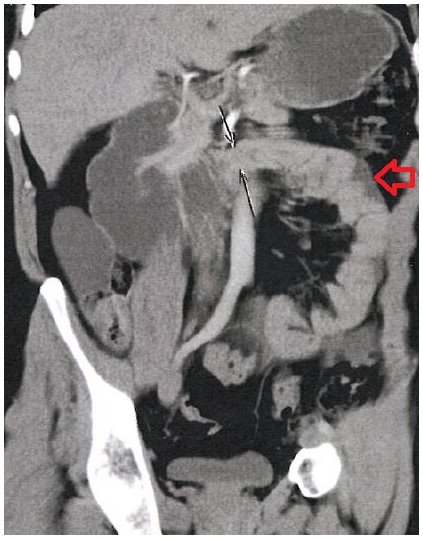

Figure 2: coronal section of the CT scan of our patient, acquisition after digestive marking and after injection of iodinated contrast product; the presence of a transition zone (white arrow) and the presence of a thick peritoneal capsule (fibrous shell) (red arrow) are demonstrated.

The biological assessment shows a slight hyperleukocytosis with hypokalemia at 2.8 meq /l.

After resuscitation, the patient is admitted to operating room for an exploratory laparotomy which revealed sheathing by a fibrous membrane of the 3rd duodenum and the proximal small intestine with distension upstream of the 2nd duodenum and a completely closed inter-hepato-diaphragmatic space.

Multiple incisions were made on the capsule that surrounded the 3rd duodenum as well as laborious adhesiolysis on the proximal small intestine with multiple biopsies of the capsule and peritoneum.

The simple postoperative course allowed the resumption of food on the 2nd day and the discharge of the patient on the 3rd day.

The histological study showed lamellar deposits of fibrin with abundant extracellular matrix, proliferation and swelling of peritoneal fibroblasts, infiltration by mononuclear cells and total loss of mesothelium on fragments labeled as capsule biopsies.

The patient was followed for three years and presented no complications or recurrences.

DISCUSSION

Encapsulating peritonitis is a chronic attack of the visceral peritoneum resulting in the formation of a thick whitish and pearly fibrous membrane totally or partially sheathing the digestive tract [Bediouia H *et al.,* 2004].

Its incidence varies between 0.7% and 7% according to the literature. Its low incidence explains the absence of a clear consensus.

In general, it occurs around the fourth decade, with a clear male predominance [Akbulut S ,2015].

The most plausible pathophysiological mechanism would be an excessive inflammatory response resulting in loss of the mesothelial cell layer and proliferation of fibroconnective tissue [Dutranoy JC *et al.,* 2009].

This chronic condition of the peritoneum can be exceptionally idiopathic [Danford CJ *et al.,* 2018] whose pathophysiology is obscure or secondary:

- **Infectious Origin:** Peritoneal tuberculosis, gynecological and digestive infections.

- **Chronic Inflammatory States:** Granulomatosis, chronic inflammatory rheumatism.
- **Iatrogenic:** Intraperitoneal chemotherapy, prolonged use of beta blockers.

The diagnosis of encapsulating peritonitis is often late or even per operative [Tagnaouti M *et al.*, 2009].

The clinical symptoms are varied and nonspecific. Combining in 86% of cases a deterioration in general condition, vague and chronic abdominal pain, episodes of sub occlusive syndromes with spontaneously resolving abdominal bloating or only nausea and vomiting.

Sometimes the Sclerosing encapsulating peritonitis can manifest itself acutely, in this case an occlusive syndrome, ischemia or digestive perforation [Li N *et al.*, 2014].

Physical examination quite often finds a hard mass with clear outlines corresponding to agglutinated loops associated with fluid effusion, as was the case in our patient.

Biologically, there is no biological criterion likely to help in the diagnosis of idiopathic encapsulating peritonitis.

The unprepared abdomen shows an air-fluid level evoking small bowel obstruction with no specific character [Tagnaouti M *et al.*, 2009; Dutranoy JC *et al.*, 2009].

Computed tomography is the cornerstone allowing both to make the positive diagnosis and to establish an adequate pre-therapeutic assessment finding specific signs:

- Agglutination of digestive loops enveloped by a thin thick membrane forming a pseudo-bag,
- Associated with dilation of the intestinal loops upstream of a stenosis.

Magnetic resonance imaging remains little used for diagnosis [Nizar El Bouardi *et al.*, 2021].

Surgical treatment is indicated in repetitive stages of sub occlusion or in the presence of an acute abdomen, as was the case in our patient.

Two surgical approaches are possible:

- A total decortication of the capsule, causing iatrogenic wounds and also an almost constant recurrence.
- The multiple incisions on the capsule associated with adhesiolysis thus allowing the release of the stenotic loops.

Postoperative mortality varies between 19 and 34% and is higher in patients with an acute abdomen [Dutranoy JC *et al.*, 2009].

The pathological aspect of encapsulating peritonitis is not specific and can be confused with simple peritoneal sclerosis or infectious peritonitis; the search for podoplanin which is a transmembrane glycoprotein on the peritoneal mesothelial cells which binds to inflammatory cytokines, helps to differentiate sclerosing encapsulating peritonitis from peritoneal sclerosis and peritonitis [Braun N *et al.*, 2011].

CONCLUSION

Encapsulating peritonitis is a rare chronic disease, its low incidence explains the lack of clear consensus.

Computed tomography constitutes the cornerstone of the diagnosis and its treatment remains essentially surgical in a conservative perspective.

ACKNOWLEDGEMENTS

To Doctors Soualili Faysal and Soualili Zinedine for their editorial assistance.

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Cite This Article: Nadim Abdelhak Soualili, Meriem El Batoul Bourouba, Abdelghani Lahcene, Kamel Bouchenak (2023). Idiopathic Sclerosing Encapsulating Peritonitis: A Case Reports. *East African Scholars J Med Surg*, *5*(7), 107-110