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Intestinal obstruction due to idiopathic sclerosing encapsulating peritonitis. Clinical report and review of literature

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SUMMARY: **Intestinal obstruction due to idiopathic sclerosing encapsulating peritonitis. Clinical report and review of literature.**

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The sclerosing peritonitis (SP) is a rare illness secondary to the peritoneal dialysis or due to intraperitoneal chemotherapy or the positioning of a peritoneal-jugular shunt in cirrhotic patient with refractory ascites or due to unkown other factors (idiophatic form) like in our patient.

The clinical pattern is various and insidious, but when an intestinal occlusive symptomatology is presents an urgent operation is mandatory. The surgical operation is often not easy and asks for a lot of attention especially in the dialyzed subject or in patients with cirrhosis, due to the possibility of postoperative bleeding and other serious complications that can result fatal.

In this report we describe surgical treatment, pathologic pattern and clinical findings of this rare disease.

RIASSUNTO: **Occlusione intestinale da peritonite sclerosante idiopatica incapsulante. Caso clinico e rassegna della letteratura.**

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La peritonite sclerosante è una condizione patologica rara, solitamente secondaria a dialisi peritoneale o a chemioterapia intraperitoneale o ancora dovuta al posizionamento di uno shunt peritoneo-giugulare in pazienti cirrotici con ascite refrattaria al trattamento; può essere causata anche da altri e ignoti fattori (forma idiopatica), come nel caso qui descritto per il quale si può pensare ad una predisposizione genetica.

Il quadro clinico è variabile e insidioso, ma quando compare una sintomatologia occlusiva si impone l'intervento, che spesso non è semplice e anzi richiede una speciale attenzione nei pazienti dializzati o cirrotici, per la possibilità di emorragie e altre gravi complicanze che possono condurre all'exitus.

Descriviamo il trattamento chirurgico e il quadro istologico e clinico di un caso giunto alla nostra osservazione.

KEY WORDS: Idiophatic sclerosing peritonitis - Intestinal obstruction - Surgery.
Peritonite idiopatica sclerosante - Occlusione intestinale - Chirurgia.

Introduction

Peritonitis, both primitive and secondary, still presents high elevated morbility and mortality despite the notable progress about the physiopathologic knowledges. Encapsulating Sclerosing Peritonitis (SP) is a severe kind of peritonitis, relatively frequent in subjects submitted to peritoneal dialysis during at least 5 years or, rarely, in subjects that received chemohypertermic intraperitoneal perfusion (1-18).

Its physiopathologic mechanisms are today still unknown even if recently some Authors suggest that the Connective Tissue Growth Factor (CTGF) can be responsible of the development of peritoneal fibrosclerosis, through the mediation of the Transforming Growth Factor-Beta 1 (TGF-β1) (1).

The evolution of post-dialythic peritonitis is characterized by the onset of fibrosis and abdominal adherences; these play an important role in the pathogenesis of chronic abdominal-pelvic pain and they constitute the most frequent cause of intestinal obstruction.

We observed a patient suffering from intestinal obstruction due to idiopathic encapsulating SP. The patient had'nt undergone dialysis neither submitted to

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intraperitoneal chemotherapy and therefore we hypothesize to other unknown etiologic factors.

Case report

C. B., 45 years old male, operated for Fallot's tetralogy twenty years before and for left inguinal hernia three years before. From four years he suffered colic pain, diffused to all abdominal quadrants and abdominal tension. Four months before he was admitted to a Department of Cardiology for cardiac failure and ascitis.

At admission the patient presents symptoms of intestinal obstruction. The rectum was empty at the rectal touch. Laboratory findings showed neutrophilia, alteration of serum electrolytes (Na 131; Cl 94; Ca 8.7). Direct Rx abdominal examination showed relevant liquid and gaseous levels. Ultrasound abdominal examinations revealed the presence of free liquid into Douglas' space and thickened walls of the ileum. Chest x-Rays showed a diffused congestive bronchial reinforcement, congestion of pulmonary hila and a very increased cardiac shade.

Laparotomy is planned and performed without a precise diagnosis. The peritoneal wall appeared thickened and duodenum, ileum, right colon and a part of the transverse colon were enormously dilated and covered and englobed by a thickened membrane. At the parietocolic grooves and among some intestinal loops serous policystic formations were present. A partial resection of the tickened layer has been performed (with dissection of all involved intestinal loops) and debulking of the cystis lesion; no intestinal resection has been necessary.

In the postoperative period the patient presented no problems; canalization to gas happened after 48 hours and to faeces after 4 days. The patient has been discharged after 9 days.

The histological examination revealed laminar fragments of fibrosclerotic tissue and cystic angiomatous-like formations of mesothelial derivation in zone areas associated with fibro-sclero-productive chronic inflammation.

Discussion

Incapsulated idiopathic SP was described for the first time by Owtschinnikov (1907); it's a rare and dangerous pathology of the peritoneum. It's characterized by the presence of a fibrous tissue, thickened and with white-grey tinges, that covers and englobes the intestinal loops, determining serious complications as obstruction, necrosis, gut-skin fistulas, malnutrition, ascites (19-23) with possibility of generalized sclerosis (24).

Mortality rate is relevant, up to 60% within 4 months from the diagnosis (25). Rarely, a fulminant SP is immediately following an acute bacterial peritonitis (26) or SP presents itself in liver transplant patients (27) or during familial mediterraneal fever (28) or after intestinal transplant in child (29) or during cirrhosis (30), but also during fatal mycotic peritonitis in humans (6) or post-leishmaniasis in a dog-model (31). In another animal model (rats) activation of matrix metalloproteinase-2 causes peritoneal injury during peritoneal dialysis (18). Other clinical factors can be the immission in the peritoneum of sterilizing chemical mediums or tampons or catheters or drugs into dialyzing liquids and others substances (32).

Among the not dialysis-dependent factors that allow the onset of the SP, there are betablocker drugs, tumors, genetic predisposition, umbilical hernias, previous abdominal operations, gallbladder disorders (33, 34). Other factors or pathologic mechanisms can be epithelial-mesenchimal transition (9) or hypertrophic mesothelium (35) or advanced (hyper-)glication end-products (36) or intracellular pH changes (induced by sclerosing drugs) in peritoneal fibroblasts (37).

The encapsulating SP, characterized by a tickening of the peritoneal membrane, is complicated by occlusion and in the dialytic patient it presents an incidence ranging from 0,7% up to 0,9% (32). After insertion of Le Veen's valve in patients with cirrhosis and ascites the incidence is up to 38% but to 17% if we consider the obstructed ones (38). Gerunda and coll. (33) reported an incidence of 8% for symptomatic patients with patterns of intestinal obstruction.

From the pathologic point of view the intestinal loops appear covered by a thickened membrane, shining and whitish, that determines stiffness in the organs involved forming tumor-like masses containing ileal loops and ascites. Microscopically the principal pattern is a homogeneously distributed sclerosis (39). There is a compression of the muscular layers with fibroblastic and mesoblastic cell infiltrates and it's often presents a cellular infiltration of leukocytes, red blood cells, macrophags and giant cells also, like in the our case.

An important question is if fibrosis and sclerosis are different disorders or different stages (39); for this kind of pathophysiologic study, PET-imaging becomes very useful (40).

The symptoms associated with the incapsulated SP are several: anorexia, nausea, vomiting, diarrhea, constipation, abdominal tension, fever, loss of muscular mass, abdominal pain, abdominal palpable mass, intestinal obstruction.

The diagnosis is obtained by abdominal direct Rx examination with the presence of the levels or by ultrasound and CT or MRI (41), that show the tickening of the intestinal wall, the evidence of picked and forced ileal loops into abdominal central region and the presence of pluricystic ascites. These signs confirm the diagnosis of encapsulated SP. It's very interesting the observation of 'peritoneal/mesenteric ossification' by CT-scans (42, 43).

Nevertheless some experiences with medical therapy (44-47), the treatment of advanced forms and especially of complications is surgical and consists in dissecting the fibrous layers from intestinal loops. In the cirrhotic or dialyzed patients mortality is elevated rather than in

patient without such pathological conditions as in our case (resolved without any complications).

Conclusions

The incapsulating SP is a rare illness secondary to the peritoneal dialysis or due to the positioning of a peritoneal-jugular shunt in cirrhotic patient with refractory ascites or due to unkown other factors like in our patient. In the case described, the etiology can be attributed to a genetic predisposition.

The clinical pattern is various and insidious, but when an occlusive symptomatology is presents an urgent operation is mandatory; the surgical operation is often not easy, especially in the dialyzed subjecty or in patients with cirrhosis, due to the possibility of bleeding and serious complications that can result fatal.

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