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A rare case of pneumatosis cystoides intestinalis with bowel perforation and secondary sepsis

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A rare case of pneumatosis cystoides intestinalis with bowel perforation and secondary sepsis

MATERIALS AND METHODS: *We present here the case of an 83 y.o. male with intestinal perforation from pneumatosis cystoides intestinalis and consequent sepsis.*
RESULTS: *The patient underwent urgency intestinal resection in our institute, with complete restitution ad integrum*
Discussion: Pneumatosis cystoides intestinalis is a rare affection, which can be categorized as primary (15%) or idiopathic(85%). The clinical appearance can be very variable from patient to patient, since it can be completely asymptomatic or start with life-threatening clinical presentation of bowel perforation and sepsis. There are various theories about the formation of the gas bubbles trough the intestinal wall. The mechanical theory assumes that the gas, tearing trough the intestinal wall seeps trough it. The bacterial theory assumes that antibiotic treatment, such as with metronidazole, allows the creation of gas by microbiological elements like Clostridium Perfringens or Clostridium Difficile. The pulmonary theory, instead, assumes that air released from ruptured alveoli gets into the mediastinum and retro peritoneum, reaching the intestinal tract. The treatment is conservative most of the times, except for the cases of intestinal perforation and sepsis.
CONCLUSIONS: *Despite of the long history of the disease, with the first description in 1783, little is known nowadays about PCI, due to the rarity of symptomatic disease. Further studies are needed to better evaluate the aetiology of the condition, and the prognostic criteria, which may be very important for clinical decisions about conservative or surgical treatment.*

KEY WORDS: Diagnosis, Pneumatosis cystoides intestinalis, Peritonitis, Therapy

Introduction

Pneumatosis cystoides intestinalis is a rare affection, with a reported incidence of 0.03%, which may occur in any age group [1]. The first to describe the disease was Du Vernoi in 1783, while the first to subcategorize it in primary or idiopathic type (15%) and secondary type (85%) was Koss in 1952 [2]. The secondary type of the disease can be associated with ileal surgery [3], colonoscopies [4], chronic pulmonary disease [5], connective tissue disorders [6] or ingestion of sorbitol [7] or lactulose [8]. The clinical presentation of the disease is varying from patient to patient, some being completely asymptomatic, while in other cases abdominal pain, diarrhoea, abdominal distension, constipation, bloody stool, flatus, anorexia, weight loss and even life threatening clinical presentations with bowel perforation and sepsis can be observed [9,10]. We report here a case of an 83 year old male with a bowel perforation and consequent sepsis from PMC which came to our attention and was treated in our institution.

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Case Experience

The patient is an 83 y.o. caucasian male, affected by Alzheimer syndrome and COPD, who had prevoius laparoscopic cholecystectomy 15 years before. When he presented to emergency unit hyperpyrexia, abdominal pain, confusional status and dyspnoea were relieved. The abdomen was intensely painful, tense, with positive Blumberg sign. There was no leucocytosis (WBC 7100, neutrophils 71%), while an abundant quantity of free air, a moderate quantity of free fluids and some intramural bubbles of air in the 1st bowel loop were found with an abdominal CT scan (Fig. 1). The patient underwent urgency laparotomy, as a laparoscopic approach was contraindicated by the anaesthesiologist, due to poor respiratory functionality.

Results

At surgical exploration a moderate quantity of intestinal content was found in the peritoneum, and a bowel loop extending for 30 cm, 10 cm after the Treitz ligament, presented multiple intramural bubbles of air (Fig. 2). An intestinal resection, including the perforated bowel loop was performed. The intestinal continuity was re-established with a mechanic anisoperistaltic latero-lateral anastomosis and an abundant peritoneal washing with 10 liters of hot saline solution was carried out. Two surgical drainages were positioned near the anastomosis and in the Douglas cavity. The patient was then transferred to the ICU unit, where he was estubated after 24 hours, and retransferred to the surgery division after 48 hours. Cycles of non invasive ventilation were carried out up to the 3rd P.O. day, when the patient started oral intake and an intense program of physiokinesitherapy. The drainages were removed at 4th and 5th P.O. days. The vesical catheter was removed at 4th P.O. day. After complete recovery, the patient was discharged from hospital at 11th P.O. day. The histological examination of the specimen reported a bowel loop of the length of 38 cm with signs of pneumatosis cystoides intestinalis (Fig. 3). After an uneventful follow up of 22 months, the patient underwent a severe respiratory infection and died due to the complications of the latter disease.

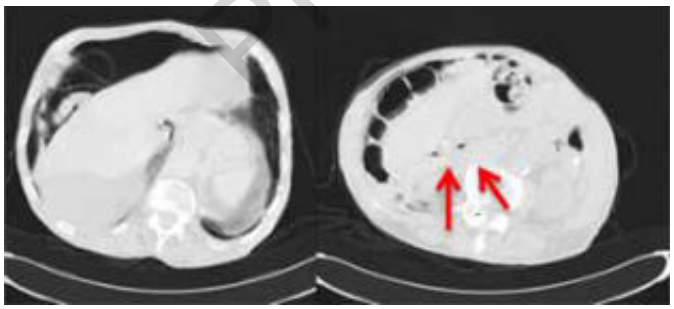

Fig. 1: Preoperative CT scan (red arrows on intraparietal bubbles of the 1st bowel loop).

Discussion

The physiopathologic mechanism which leads to PCI is mostly unknown. Inflammation, physical damage of intestinal mucosa, nutritional imbalance with disbacteriosis, gastrointestinal dysmotility and immune dysfunction have been proposed as possible causes of the disease [11] The gaseous cysts have been found to contain nitrogen, hydrogen and carbon dioxide [12]. The most common site of involvement is the ileum, followed by the colon. In 20% of the cases both of the intestinal tracts are affected [13]. As for the aetiology, there are various theories about the formation of the gas bubbles trough the intestinal wall. The mechanical theory assumes that the gas, tearing trough the intestinal wall seeps trough it [14]. The bacterial theory assumes that antibiotic treatment, such as with metronidazole, allows the creation of gas by microbiological elements like Clostridium Perfringens or Clostridium Difficile. The pulmonary theory, instead, assumes that air released from ruptured alveoli gets into the mediastinum and retro peritoneum, reaching the intestinal tract. The latter theory is sustained by the fact that PCI is often associated with COPD and asthma [5,15]. PCI affects the three layers of the intestinal wall. The cysts have no real wall and in 50% cases they

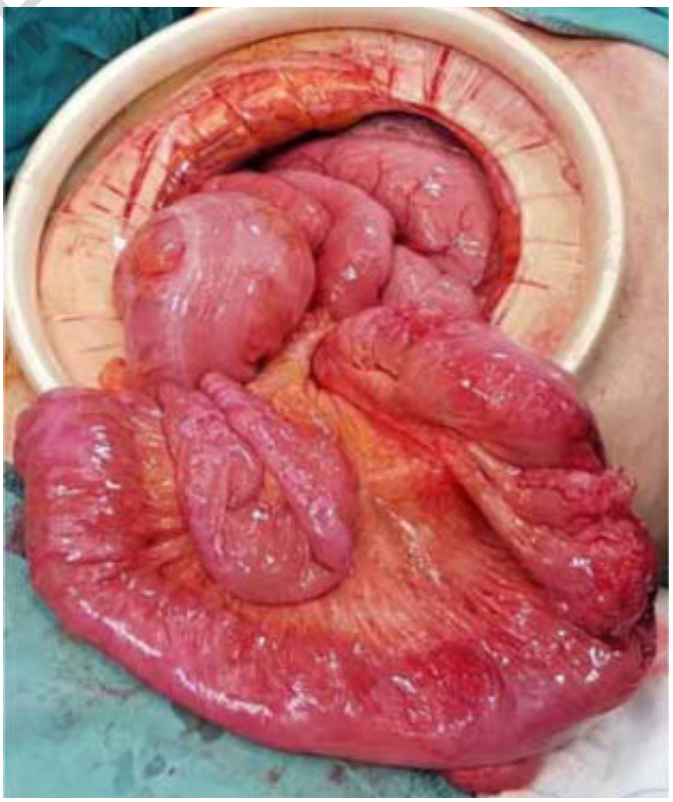

Fig. 2: Intraoperative aspect of the ileum.

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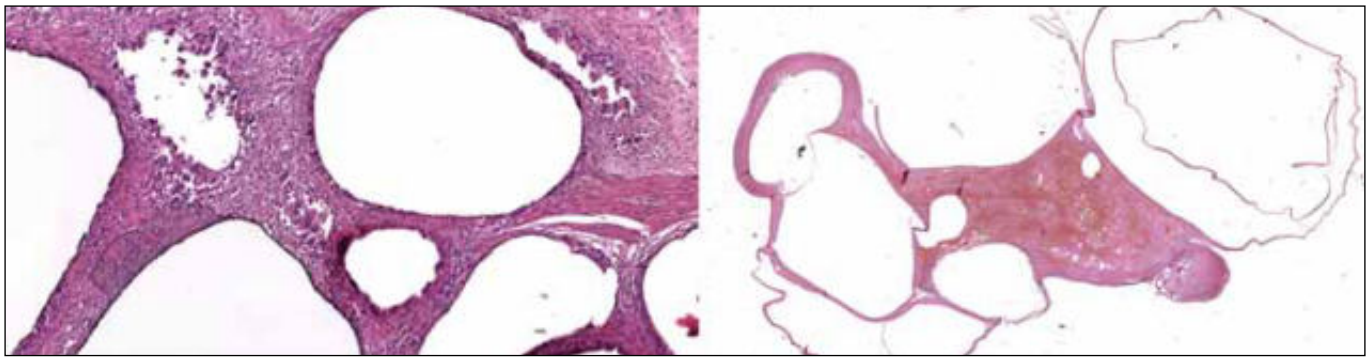

Fig. 3: Histological aspect of the ileal wall.

automatically disappear [16]. Most of the patients are completely asymptomatic. Complications occur in 3% of the cases, due to the occlusion of intestinal lumen, bleeding or necrosis with perforation [17]. PCI is only visible on plain abdominal radiographs in about two-thirds of the patients. On US examination, reverberation due to the air content of the bubbles may be visible [18]. The most characteristic findings are on CT scan, where some grape-like clusters or honeycomb-shaped shadows may be visible along the intestinal wall. These clusters can be associated with free air or fluids, as in the reported case [19,20]. Currently, conservative management with administration of oxygen, to help reabsorption of the bubbles, is recommended in most of the patients. Some Authors propose the application of Bifidobacterium to regulate disbacteriosis and improve the function of gastrointestinal tract [9]. Surgery represents the second choice in case of life-threatening complications such as perforation and intestinal necrosis [21-25]. A laparoscopic approach may be indicated in cases where the conditions of the patient allow the pneumoperitoneum [13]. In the presented case a conservative approach or laparoscopy were contraindicated due to the very poor clinical conditions of the patient, with particular regard to respiratory function.

Conclusions

Despite of the long history of the disease, with the first description in 1783, little is known nowadays about PCI, due to the rarity of symptomatic disease. Further studies are needed to better evaluate the aetiology of the condition, and the prognostic criteria, which may be very important for clinical decisions about conservative or surgical treatment.

Riassunto

Nel presente articolo, viene illustrato il caso di un paziente di 83 anni, presentatosi in Pronto Soccorso con i segni di una perforazione intestinale e conseguente sepsi. La diagnosi si riferiva ad una pneumatosi cistoide intestinale, una rara affezione spesso associata a BPCO, da cui lo stesso risultava affetto. Il pz. è stato sottoposto presso il Nostro Istituto ad intervento di resezione intestinale in urgenza, con decorso post-operatorio privo di complicanze. La pneumatosi cistoide intestinale può essere classificata come primaria (15% dei casi), o idiopatica (85% dei casi). I sintomi clinici possono essere estremamente variabili da paziente a paziente, dato che la patologia può risultare completamente asintomatica o piuttosto esordire con i segni di una peritonite da perforazione intestinale con stato settico generalizzato. Esistono differenti teorie a proposito della formazione delle bolle aeree nel contesto della parete intestinale. La teoria meccanica postula che il gas, erompendo attraverso la parete intestinale, si insinui attraverso la stessa. La teoria batterica, invece, parte dal presupposto che il trattamento antibiotico, per esempio con metronidazolo, potrebbe permettere la creazione di gas da parte di agenti patogeni come il Clostridium Perfringens o il Clostridium Difficile.
Di contro la teoria polmonare considera che la fuoriuscita di aria in seguito alla rottura degli alveoli, insinuandosi nel mediastino e retroperitoneo, potrebbe raggiungere il tratto intestinale. Il trattamento è conservativo nella maggior parte dei casi, escludendo quelli che evolvono in perforazione intestinale. Nonostante la lunga storia della patologia, la cui prima descrizione risale al 1783, ancora oggi esistono molti punti oscuri sulla conoscenza dei meccanismi fisiopatologici e clinici per via della rarità della malattia sintomatica. Sono sicuramente necessari ulteriori studi per definire eziologia e criteri prognostici, anche al fine di guidare il clinico nella diagnosi e tra le opzioni di trattamento conservativo o operatorio.

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