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Intestinal Cystic Pneumatosis (ICP), a Rare Cause of Acute Abdomen Case Report and Literature Review

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ABSTRACT

Intestinal cystic pneumatosis is a rare, benign and often an asymptomatic disease. It is considered to be one of the main causes of non-surgical pneumoperitoneum. We report the case of a 53 year-old woman with diffuse pneumatosis cystoides intestinalis revealed by a pseudo-occlusive syndrome and whose diagnosis was suggested by imaging. The evolution under medical treatment was simple and without having to resort to surgery.

Introduction

Intestinal cystic pneumatosis (ICP) is a rare cause of acute abdomen, defined by the presence of a gas cyst in the intestinal wall that can affect the entire digestive tract with a predilection for the small intestine and the colon. It can affect all ages, especially adults after the age of 50. It preferentially affects men with a sex ratio of 3.5 / 1. (1-2)

In this observation we will discuss the case of a patient treated at the begining as a peritonitis but in fact it was just a cystic intestinal pneumatosis which regressed without any surgery.

Observation

We report a case of a 53-year-old patient with a history of peritonitis due to perforation of a peptic ulcer in the past, no other known comorbidity were detected.

The patient was admitted to the emergency for dyspnea with diffuse abdominal pain and a 40 degree temperature.

Clinical examination found polypnea at 28 cycles per minute, tachycardia at 110 beats per minute with normal blood pressure at 118/58 mm Hg, with an increase in abdominal volume.

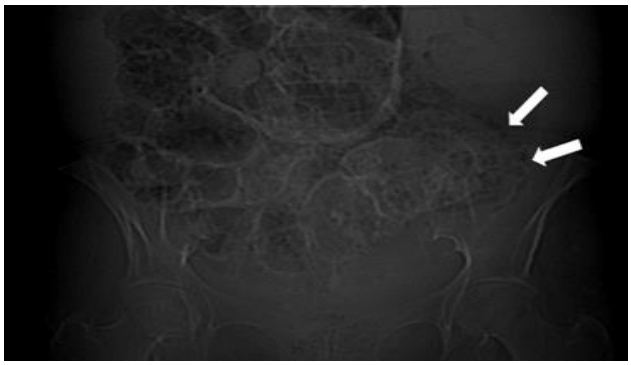

Figure 1. X-ray of the abdomen showing rounded hyperclar images adjacent to the digestive lumen grouped together in "bunches of grapes" or "chains", achieving an aspect of double gas contour (arrows).

Laboratory assessment did not find any notable abnormality other than 5.9 mmol / L kalaemia without electrical cardiac signs and elevated markers of inflammation. The abdomen X-Ray showed distension with hydro-aeric levels (Figure 1).

Abdominal CT was performed, confirming a diffuse digestive distension containing multiple parietal cystic formations as a “grape cluster”, explaining the occlusive syndrome (Figure 2 (a, b)) with an important hydro-pneumoperitoneum.

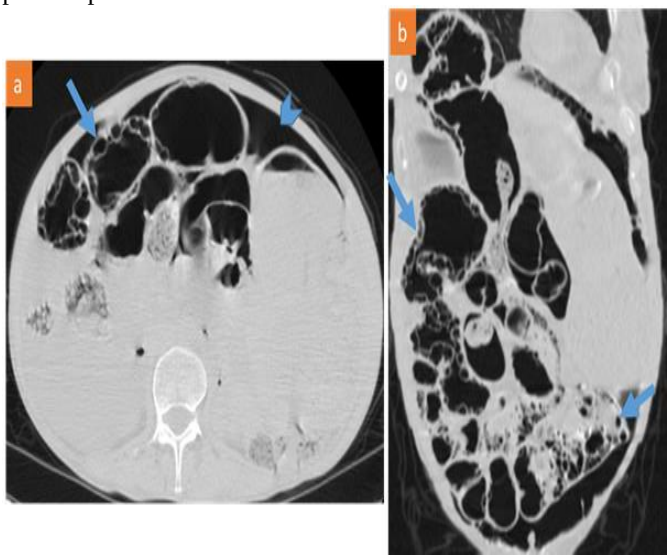

Figure 2. Abdominal CT in axial (a) and coronal (b) section in pulmonary window showing typical pattern of gas bubbles in the wall of the intestine (arrows) associated with a hydro-pneumoperitoneum (arrowhead).

The surgical team opted for a symptomatic treatment with oxygen, pain killer and antibiotic based on protected amoxicillin.

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The outcome was favorable with resolution of the symptoms with regression in the control abdomen CT scan.**Discussion**

Intestinal Cystic pneumatosis (ICP) is a rare cause of acute abdomen but its frequency is probably underestimated. It is defined by the presence of a gas cyst in the intestinal wall which can reach the entire digestive tract with a predilection for the small intestine and the colon.

It can reach all ages, frequently described in adults after 50 years. intestinal Cystic pneumatosis preferentially affects men with a sex ratio of 3.5 / 1, although some series have found more female predominance (1-2).

It is classic to distinguish between the primitive or idiopathic forms located mainly in the left colon and the secondary forms which are essentially thin (2,3).

From an anatomo-pathological point of view, cystic formations are especially developed in the submucosa, in particular in colonic damage, and / or in the subserosa, in particular in the small intestine.

The extension of these cystic formations can be done at the level of the mesentery, in the subperitoneal cellulo-fatty spaces of the retro-peritoneum and of the abdominal walls and can lead to a benign pneumoperitoneum or retro-pneumoperitoneum testifying to the rupture of a sub-serous cyst. It is found in about 15% of cases of the slime forms (4).

The size of the cysts varies from a few millimeters to several centimeters in diameter (1.5). Their gaseous content is very rich in hydrogen and nitrogen; there is also a small amount of oxygen, carbon dioxide and methane (1).

ICT can be idiopathic or more often secondary to various gastrointestinal pathological associations (pyloric stenosis, peptic ulcer, intestinal obstruction, abdominal trauma, inflammatory bowel disease, intestinal anastomoses, celiac disease, after digestive endoscopies) or extra gastrointestinal (COPD, heart disease, cystic fibrosis, lupus, scleroderma), (5). In our case, given the prior visceral surgery, we concluded that a secondary cystic pneumatosis of digestive origin is based on mechanical theory (1); suggesting the penetration of air through the digestive mucosa by trauma thereof and / or by increased endoluminal pressure. The presence of Helicobacter pylori on gastric biopsies led us to suggest an association of two mechanisms; mechanical and bacterial theories, the latter suggests the invasion of the intestinal wall by gas-producing bacteria, in particular hydrogen.

Clinically, ICT is usually asymptomatic or paucisymptomatic. When it exists, the symptoms are related to luminal occlusion, extrinsic compression, or mucosal pain favored by cysts (1).

Very often, the diagnosis of intestinal cystic pneumatosis is accidental, on the occasion of a symptomatology borrowed by radiological or endoscopic examination (1,5). More rarely, the symptomatology is related to a complication such as volvulus, intussusception, perforation or hemorrhage in an acute abdomen picture.

The diagnosis of ICT can be evoked from the X-Ray in front of rounded air images joined in bunches of grapes or chains, lining the wall of the digestive tract.

Two indirect signs are important to look for (3): the sign of Moreau Chilaïditi which corresponds to the interposition of multiple clusters of bubbles between the liver and the right diaphragmatic dome and a pneumoperitoneum testifying to the rupture of a subserous cyst.

Computed tomography has good diagnostic accuracy. It shows gaseous cystic formations in the digestive wall, better visible in axial slices and in the pulmonary window.

Ultrasound examination is nonspecific, however it may suspect the diagnosis by showing thinning of the intestinal wall and echoes with acoustic shadow.

On endoscopy, colonic submucosal gas cystic lesions can mimic adenomatous polyposis. Typically, the cyst collapses during an attempted biopsy with a ‘’popping sound’’, thereby correcting the diagnosis (3).

Treatment varies depending on the etiology. For primary forms, antibiotic is indicated as a first-line treatment. If this fails, mask or hyperbaric oxygen therapy, which will promote the replacement of hydrogen by oxygen, should be attempted. For the secondary forms, the treatment is that of the causal affection.In most cases, intestinal cystic pneumatosis is asymptomatic and no treatment is necessary. Surgery, for its part, should remain reserved for particularly serious forms. (1)

Usually the progression is benign, however there are severe forms with a reserved prognosis, especially in immunocompromised patients.

Conclusion

Cystic intestinal pneumatosis is one of the main causes of non-surgical pneumoperitoneum, its recognition is important to avoid engaging in abusive therapeutic attitudes.

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