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Case Report

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An asymptomatic case of pneumatosis cystoids intestinalis: A case report

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Abstract

Introduction: pneumatosis cystoides intestinalis is an uncommon disease characterized by the presence of gas within the submucosa or subserosa of the intestine. Pneumatosis cystoides intestinalis can be often localized in colon and it may be asymptomatic.

Case presentation: A 53's man, with an autistic spectrum disorder and no other comorbidities nor symptoms, underwent a colonoscopy because of finding of a wall thickening of the rectum on the last abdominal computerized tomography. The colonoscopy revealed the presence of multiple cysts, resembling a bunch of grapes at the left-colonic flexure with normal overlying mucosa.

Conclusion: pneumatosis cystoides intestinalis may be asymptomatic or sometimes diagnosed incidentally during colonoscopy. The treatment approach depends on underlying cause and it can be conservative or surgical in restricted cases.

Introduction

Pneumatosis cystoides intestinalis (PCI) is an uncommon disease with an unknown etiology, characterized by the presence of gas within the submucosa or subserosa of the intestine [1]. PCI can be localized anywhere within the gastrointestinal tract [2]. PCI may be asymptomatic or sometimes diagnosed incidentally during colonoscopy requested for various symptoms (i.e., abdominal pain, diarrhoea) [3] and treatment approach is related on the underlying cause.

Case Report

A 53's-man, outpatient, with an autistic spectrum disorder and no other comorbidities, underwent surgery and radiotherapy on 2012 for a seminoma and was on annual follow up with CT. The last one showed a wall thickening of the rectum. So, he underwent a colonoscopy. In the period of the colonoscopy, he had no abdominal symptoms, laboratory findings were normal, and he was not taking any therapy. The colonoscopy revealed the presence of multiple cysts, resembling a bunch of grapes and compressible with forceps, at the left-colonic flexure. The overlying mucosa was normal, confirmed by use of NBI (Figure 1-3). Furthermore, the rectum and the other colonic segments were normal. A subsequent review of CT images confirmed the presence of gas collections within the left-colonic flexure wall (Figure 4). So, we confirmed the diagnosis of PCI and prescribed rifaximin therapy.

Discussion

Pneumatosis cystoides intestinalis (PCI) is an uncommon disease with an unknown etiology, characterized by the presence of gas within the submucosa or subserosa of the intestine. In a recent study about Chinese population including 239 PCI cases, was found a M:F ratio of 2,4:1 and the mean age of patients was 45.3 ± 15.6 years (range: 2-81 years) [1]. PCI can be localized anywhere within the gastrointestinal tract from the oesophagus to the rectum: it involves the large intestine in 46% of the cases, the small intestine in 27% of the cases, the stomach in 5% of the cases and both large and small intestine in 7% of the cases [2].

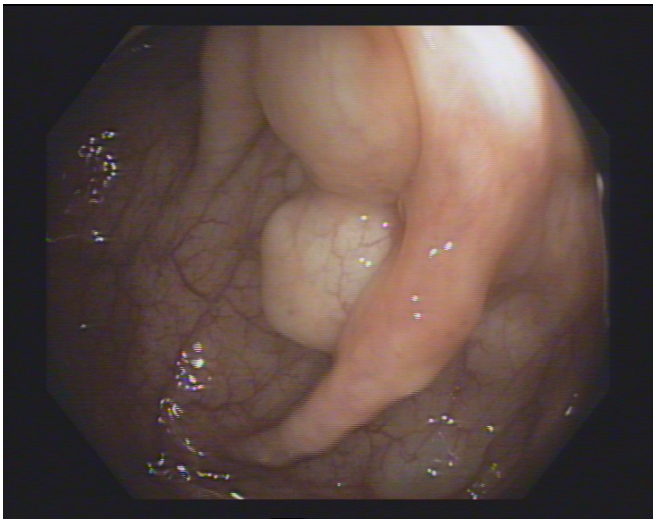

Figure 1. Multiple cysts with overlying normal mucosa, resembling a bunch of grapes at the left-colonic flexure

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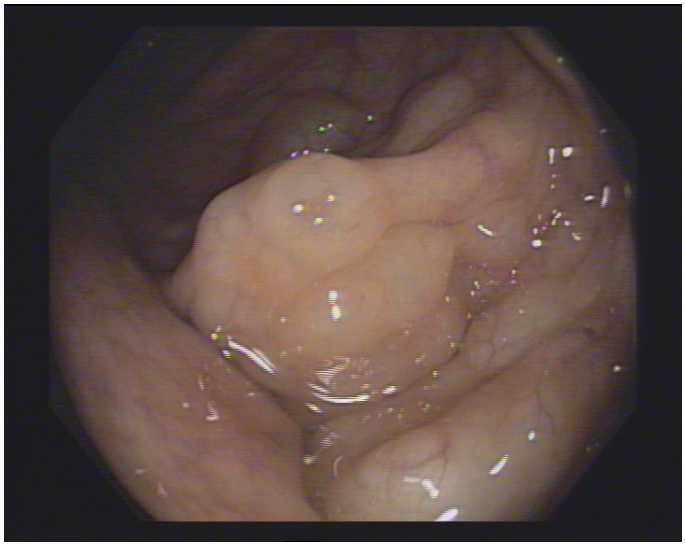

Figure 2. Multiple cysts with overlying normal mucosa, resembling a bunch of grapes at the left-colonic flexure

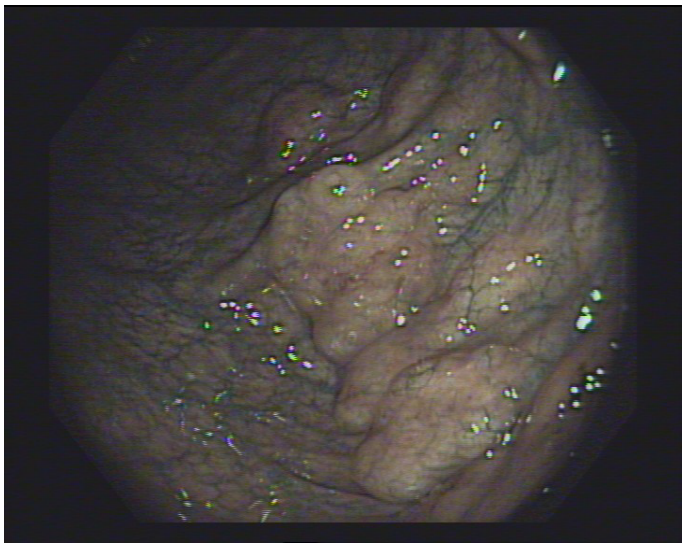

Figure 3. NBI use confirmed normal appearance of mucosa

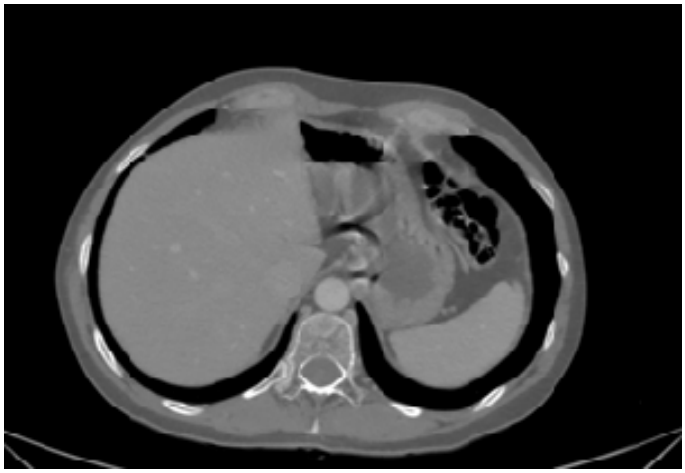

Figure 4. CT-scan image

PCI is mainly asymptomatic and is often diagnosed incidentally, as in our case; some cases present with diarrhoea or constipation, abdominal pain, bloody stool, loss of appetite, weight loss, and even severe events as perforation or pneumoperitoneum [2,4]. PCI can be divided into primary (15%) and secondary (85%) types [5]. Various gastrointestinal and non-gastrointestinal diseases has been considered as causes of secondary types of PCI, such as bowel ischemia [6], inflammatory bowel diseases, bowel infection, gastroduodenal ulcer, necrotizing enterocolitis [5], pyloric stenosis [7], diverticular disease [5], obstructive pulmonary disease [8], drugs (e.g., alpha-glucosidase inhibitors) [9,10], collagen tissue diseases [11,12], acquired immune deficiency syndrome [13] and malnutrition [4,5,14]. Anywhere none of these symptoms and diseases affected our patient, so his type of PCI can be cosidered as primary type.

Pathogenesis

Various theories have been proposed to explain the gas passage or formation in the submucosa or subserosal.

1) the mechanical theory: ischemia, obstruction, traumas, surgery, or even biopsies during a colonoscopy, could cause increase of intraluminal pressure, wall injury and allow the passage of gas within intestinal wall. However, this does not explain why gas remains within intestinal wall [1].

2) the bacterial theory: an increased mucosal permeability and the presence of gas-producing bacteria, in particular hydrogen, could cause the gas passage within the wall. A support to this theory is the disappearance of PCI after antimicrobial therapy [15]. Some authors have demonstrated the appearance of PCI in animals with intestinal infection by Clostridium perfringes [16]. However, is not possible isolate or cultivate bacteria from the cysts.

3) the pulmonary theory: pulmonary diseases, such as chronic obstructive pulmonary disease (COPD), asthma, and interstitial pneumonia, may result in pulmonary alveolar rupture and movement of gas through the mediastinum and retroperitoneum and locates within the bowel mesentery. In some cases, alveolar gas dissects along the aorta, through the diaphragm and into the mesenteric blood vessels and then becoming trapped in the bowel wall. A support to this theory is the presence in literature of various case reports of PCI in patients with pulmonary diseases [17,18,19]. However, this theory alone doesn't explain the fact that hydrogen, a gas that human cells aren't able to produce, may be represent up to 50% of the gas content within the cysts. Nevertheless, many patient affected by PCI are not affected by pulmonary diseases [20]. None of these three theories completely explain the pathogenesis of PCI, perhaps it is more likely a combination of two or more theories.

Diagnosis

Nowadays because of the increase of number of colonoscopies for colorectal cancer screening, it is possible to find PCI incidentally in asymptomatic patients. The endoscopic appearance is the presence of gas-filled blebs resembling a bunch of grapes. The mucosa is often normal or thin or sometimes has erosions or erythema. Sometimes PCI could be mistaken with polyps or submucosal lesions. Typically, blebs collapse when biopsed [21]. In symptomatic patient, for example those with abdominal pain, it's more frequent to make diagnosis with X-ray or CT. Abdominal CT is more sensible than X-ray and shows a thickened bowel wall containing gas within submucosa or subserosa.

Furthermore, CT scan shows complications such as perforation or pneumoperitoneum or signs of the underlying disease [1,22]. In our patient the CT report did not mention PCI, but a subsequent review of the images allowed to highlight the presence of gas in the same sites reported by the colonoscopy.

Treatment

Treatment of PCI is related on the underlying cause. There is not a specific treatment in asymptomatic patients. Conservative approaches (intestinal rest, antibiotic therapy and oxygen) are suitable in patients without complications requiring surgery [23]. A spontaneous remission is also possible. The medical history of the patient and his current drugs should be evaluated carefully to guide further decisions. Khalil et al. proposed a treatment algorithm to support the decision-making process; it is based on evaluation of clinical condition, laboratory and CT findings and medical history. It may help to reduce the rate of patients with benign forms of PCI unnecessarily subjected to exploratory surgery and reduce the delay in surgical therapy for those patients who would profit from early surgical intervention [20]. Our patient was completely asymptomatic, so we decided for rifaximin therapy and follow-up with endoscopic re-evaluation.

Conclusion

Pneumatosis cystoides intestinalis may be asymptomatic or sometimes diagnosed incidentally during colonoscopy. The treatment approach depends on underlying cause and it can be conservative or surgical in restricted cases.

Conflict of interests

The author declares that he has no conflict of interests.

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Declaration of patient consent

Informed consent of patients was obtained.

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