

Pneumatosis Cystoides Intestinalis – An Incidental Post-Operative Rare Case Report in Port Sudan Teaching Hospital, Sudan

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ABSTRACT

Background: *Pneumatosis intestinalis* also called *Pneumatosis cystoides intestinalis (PCI)* is a rare disease and difficult to diagnosis that confuses many doctors. A vast number of factors are suspected to contribute to its pathogenesis, such as Crohn's disease, intestinal stenosis, ulcerative colitis, drug use, extra-gastrointestinal diseases, and chronic obstructive pulmonary disease.

Case Report: A 41-year-old man presented to port Sudan teaching hospital on 20 July 2021 with symptoms of abdominal pain, abdominal distension. A physical examination revealed tenderness all over the abdomen. Then patient was admitted to the hospital, and resuscitation was done. Laboratory investigations revealed no abnormalities. Radiological investigation: abdominal ultrasonography and plain abdomen x-ray showed signs of intestinal obstruction. Finally, patient was undergone exploratory laparotomy on next day. Final diagnosis reached after result of histopathology was presented as: *Pneumatosis cystoides intestinalis*.

Conclusions: *Pneumatosis intestinalis* difficult to diagnosis, so an efficient recognition of the clinical scenario, encompassing the current clinical context, comorbid conditions, physical examination findings, laboratory data, and radiographic details, assists the clinician in reaching the correct diagnosis and offering appropriate treatment.

Keywords

Pneumatosis cystoides intestinalis, Exploratory laparotomy, Port Sudan, Sudan.

Introduction

Pneumatosis cystoides intestinalis (PCI) was first reported by Du Vernoi in 1730 [1]. In a systematic review and analysis of 239 patients with PCI, Wu et al. reported that the peak age at onset was 45.3±15.6 years (range, 2–81 years), the male to female

ratio was 2.4:1, and the mean disease course was 6 months [2]. After examining 123 patients with PCI, Boerner reported an equal incidence in male and female patients and that remission was achieved in 70% of patients using nonsurgical treatment [3]. PCI is distributed throughout the digestive tract, particularly the subserous or submucosa of the small intestine and colon, in which multiple pneumocysts develop. The distal stump of the transverse splenic flexure colon, particularly the descending and sigmoid colon, is most commonly affected [2].

Case Report

A 41-year-old man was admitted to emergency room in port Sudan teaching hospital, port Sudan, Sudan on 20 July 2021, with symptoms of abdominal pain, abdominal distension, no diarrhea, hematochezia or melena. Pain was mild, cramping, diffuse, and worse with food. There was no tenesmus, and the patient denied nausea or vomiting. No fever, chills, myalgias, and weight loss or systemic signs of illness were present. Past surgical history was and review of systems was otherwise unremarkable. Physical examination revealed a temperature of 36.8°C, blood pressure of 90/60 mm Hg, pulse of 72 beats per minute, 18 respirations per minute, and oxygen saturation of 99% on ambient air. The patient was a nontoxic. Cardiovascular exam showed an irregularly irregular rhythm with regular rate. Pulmonary examination demonstrated lungs which were clear to auscultation; there was no wheezing, rales, or rhonchi. Her abdominal exam revealed a soft abdomen with no significant tenderness to light palpation; however, there was diffuse tenderness to deep palpation. There was no rebound or guarding, and bowel sounds were normal. Initial laboratory investigations were unremarkable. Radiological investigation: abdominal ultrasonography and plain abdomen x-ray showed signs of intestinal obstruction. Finally, patient was undergone exploratory laparotomy on next day: on laparotomy was found multiple sacculations, cysts with bowel perforation in small intestine, so resection and anastomosis were done and specimen sent for histopathology. final diagnosis after result of histopathology was: Pneumatosis cystoides intestinalis.

Discussion

PCI can be divided into types: idiopathic called primary (15%) and secondary to other causes (85%) types [2,4]. The secondary type occurs secondary to diseases such as digestive tract stenosis, obstructive pulmonary disease, abdominal external injury or surgery, and malnutrition [2,3]. There are three hypotheses of PCI pathogenesis: (1) mechanical theory: involving an increase in intraluminal pressure that causes mechanical damage and mucosal rupture of the intestinal wall, leading to the migration of gas from the gastrointestinal cavity to the intestinal wall [1]; (2) pulmonary theory: chronic lung diseases such as chronic obstructive pulmonary disease, asthma, and interstitial pneumonia lead to alveolar rupture, causing mediastinal emphysema and release of gas along the aorta and mesenteric blood vessels into the intestinal wall [5]; and (3) bacterial theory: aerogenic bacteria penetrate the intestinal mucosal barrier, ferment in the intestinal wall, and produce gas [6].

PCI lesions are mainly located in the colon (46%) and small intestine (27%), followed by the large and small intestine (7%) and stomach (5%) [8]. The clinical manifestations of primary PCI are nonspecific, such as abdominal pain (59%), diarrhea (53%), nausea and vomiting (14%), mucus in stool (12%), and hematochezia (12%) [2]. Secondary PCI also has primary disease manifestations. About 3% of the patients with PCI complained of complications, including pneumoperitoneum, volvulus, intestinal obstruction, and intestinal ischemia [1,2,9-11]. Serious complications may alter the decision-making process for the therapeutic schedule.

Prior authors have devised multiple methods to classify the expansive differential diagnosis which encompasses PI. A more inclusive list of conditions associated with PI is discussed elsewhere [14]. From a clinician's view, however, an initial clarification between life-threatening and no urgent pathologies is of utmost importance. Most concerning, PI may be indicative of necrotic tissue allowing gas to penetrate the submucosa. Mesenteric ischemia from low-flow states or infarction from acute arterial occlusion may be the most concerning of the high risk etiologies and can give rise to this clinical picture [15].

A high index of suspicion warrants an urgent intervention and consideration of surgical consultation for suspected ischemia or infarction. Other urgent pathologies which can be associated with ischemia include intestinal obstruction as well as volvulus and malrotation. When considering ischemia or infarction, rapid recognition of the overall clinical picture is imperative. There are several key features of the current clinical scenario, patient past medical history, physical exam, and laboratory data, which can heighten suspicion for life-threatening PI. An immediate appreciation of low-flow vascular states, such as sepsis, CHF, use of IV pressors, and other causes of hypotension, should be made by the physician. Arrhythmias, which can cause both low-flow states as well as precipitate embolic phenomena, should also be taken into account. The patient's past medical history should be examined for peripheral vascular disease and coronary artery disease as well as risk factors for vascular disease such as hypertension, hyperlipidemia, diabetes, and smoking. Physical examination should focus upon the abdomen, with caution being given to an exam revealing the classic ischemic finding of "pain out of proportion to exam." Signs of peritonitis, although uncommonly present, may also suggest ischemia [13]. Laboratory data should include a lactate level, with an elevated lactate raising serious concern for ischemia [12]. Furthermore, details of the radiographic findings can help define the nature of PI. The finding of additional gas in the vasculature, particularly portal venous gas, can be an ominous sign and correlates to transmural bowel necrosis [12]. Other authors have suggested that so-called crescentic or linear gas collections may indicate bowel infarction and are more often associated with more sinister pathology [16]. This should be contrasted with so-called cystic PI, which represents discrete bubbles of gas attached to one another along the digestive tract wall and is usually considered benign. Small bowel PI is more frequently associated with ischemia.

Conclusion

The incidence of PCI is low. Due to a lack of specificity in clinical manifestations and endoscopic findings, it is often misdiagnosed as intestinal polyps, tumors, inflammatory bowel disease, or other conditions. Colonoscopy, computed tomography, and ultrasonography have demonstrated benefit in patients with multiple nodular projections in colon. Compared to the treatment of the above diseases, PCI treatment is effective and convenient, and the prognosis is optimistic. Therefore, clinicians should increase their awareness of PCI to avoid unnecessary misdiagnosis.

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