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Pneumatosis Cystoides Intestinalis – a Report of Two Cases

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The authors present two cases of an extremely rare pneumatosis cystoides intestinalis of large and small intestine in a 48-year old male and in a 77-year old female surgical patients.

Introduction

Pneumatosis cystoides intestinalis (PCI) has not been differentiated as a separate medical entity, but considered rather as a morphological condition of an unknown origin. The condition was for first reported as a post-mortem observation by Du Vernei in the eighteenth century. Although this condition is usually asymptomatic and incidentally found during laparotomy or on radiological investigation of unrelated symptoms, it can also cause abdominal pain, subacute intestinal obstruction, intussusception, or rectal bleeding. It is characterized mainly by the presence of mid-membrane cystoid air-filled cavities found usually in submucosa or subserosa of the bowel [8, 25]. However, it must be highlighted that all parts of gastrointestinal tract may be involved [7, 19].

Typically, PCI has been observed to accompany the following clinical entities: 1) infant necrotizing enterocolitis [20, 27], 2) chronic obstructive pulmonary disease [13], 3) connective tissue diseases (collagenoses) [3, 12, 14, 24, 26], 4) chronic gastric ulcer with pyloric stenosis [6, 13]; a significant rate of PCI was noted as a complication of liver, heart or bone marrow transplantation (in these cases it is thought to be linked to the immunosuppressive cytostatic or long-term steroid treatments) [1, 5, 17, 21, 23]. Other reported cases were found to co-exist with Lesniowski-Crohn disease, ulcerative colitis, celiac disease and various cancers [8, 9, 22].

Literature data report few cases of PCI related to various therapeutic and diagnostic procedures e.g. intestinal anastomoses, endoscopic interventions within bowel (polypectomy), double-contrast X-ray bowel examination and jejunostomy nutrition. PCI has been also described to develop in the course of HIV infections and ischemic bowel disease or even in cocaine addiction [4].

Description of Cases

Case 1

A 48-year old male was admitted on November 2001 to the Department of Medicine in Regional Specialistic Hospital in Tychy for a planned diagnostic procedure, with the initial diagnosis of sigmoid polyposis. The patient suffered from constipation, flatulence and diarrhea with a reoccurring bleeding. A weight loss was reported caused by a dietary regime and there was no history of increased body temperature. The first symptoms had been noticed about eight months earlier. At admission no significant abnormalities were found on the physical examination and the biochemical tests were also within normal range (with the exception of a slightly elevated blood pressure). The patient after having been monitored and prepared for the surgical procedure was transferred to the Department of Surgery and underwent sigmoid resection with subsequent end-to-end anastomosis. The postoperative course was uneventful. The patient was discharged from hospital in a good general condition.

The surgery specimen No. 201761–763 submitted for pathological analysis comprised a fragment of a large bowel with adjacent mesosigmoid up to 2cm-wide. Almost entire serosa of the bowel fragment constituted a “cystically” creased area (pseudopolyps) covered with creases reaching 2cm in diameter and distributed mainly around mucosa band (Fig. 1). The material was fixed in formalin and embedded in paraffin blocks and then routinely stained with HE and additionally with Mayer’s mucicarmine and paS-alcian blue.
Histologically, in all bowel wall layers cystic cavities were revealed (probably air-filled), surrounded by multinucleated foreign-body cells and very scarce inflammatory infiltration composed of lymphocytes and plasma cells. Focally in these inflammation regions fibrosis was visible (Fig. 2). Apart from mechanical compression no other traces of microscopic abnormalities were found in mucosa and submucosa (Fig. 3).

Case 2

A 77-year old female with acute ileus was transferred to the Department of Surgery on August 2002 from one of the local hospitals. When interviewed, the patient reported increasing abdominal pain lasting for at least 5 days with gas and defecation blockage. These symptoms were preceded by some diarrhea and mild abdominal discomfort. Some weight loss was also noticed – about 5kg during three months. The patient had been undergoing a treatment for gastric ulcer and had been operated on twice; one of these surgical procedures was appendectomy and the other – gynecological operation due to an unspecified uterine disease (lack of data). During the initial examination a mild-grave patient status was established and significant flatulence and lack of peristalsis were pointed out. Abdominal X-ray scan revealed typical signs of ileus and biochemical tests showed no abnormalities with exception of an elevated WBC level of 15,000 and a high creatinine level (130µmol/l), hyperglycemia (8.65mmol/l) and elevated alkaline phosphatase level (270U/l). Under these circumstances an emergency operation was performed. In the course of surgical proceedings multiple adhesions were found in the peritoneal cavity; a 50cm-long “pseudo-necrotic” fragment of the small intestine was found and a cyst of left ovary was also discovered. A partial resection of small intestine with an end-to-side anastomosis of remaining bowel was performed. The left ovary cyst was also removed. During postoperative period no early or late postoperative complications were observed, and the patient was discharged from the Clinical Department of Surgery on the eighth postoperative day in a good general condition.

The material submitted for pathological analysis consisted of the 74cm-long small intestine fragment with adjacent mesentery up to 7cm wide; most of the small bowel

Fig. 1. Cystic cavities in all layers of the large bowel wall.

Fig. 2. Subserosal inflammation and fibrosis. HE. Magn. 40x.

Fig. 3. Cystic cavities in mucosa. HE. Magn. 20x.

Fig. 4. Multinucleated foreign body giant cell reaction. HE. Magn. 200x.
displaying “minor cyst cavities” up to 0.5cm in diameter located mainly under the mucosa (specimen No. 209467–701). The formalin-fixed material was embedded in paraffin and the sections were subjected to routine staining, the same as in the Case 1.

Histologically, submucosa and mucosa showed diffusely distributed cyst-like cavities (probably air-filled) surrounded by multinucleated foreign body giant cell reaction (Fig. 4) and lympho-plasmacytic inflammatory infiltrate with accompanying fibrosis (Fig. 5).

Discussion

In pneumatosis cystoides intestinalis (PCI) air-filled cysts are present in the bowel wall and mesentery, and may occur anywhere in the gastrointestinal tract [9]. The cysts (0.5–10cm in size) are found most frequently in the terminal ileum and rarely in the proximal small bowel, stomach [20] and colon [7]. When the air-filled cysts rupture, they cause a pneumoperitoneum, with often is benign in nature [10].

Pneumatosis cystoides intestinalis has a number of different etiologies. Primary PCI (15% of cases) affects primarily the colon, secondary (85% of cases) affects the small intestine and is associated with mucosal breakdown or a bacterial or mechanical etiology. After clinical observation and histopathological examination Case 1 may be regarded as primary or idiopathic, in which there is no other known pathology. The three etiologies in secondary PCI have been proposed as follows. Mucosal breakdown theory – steroids and other immunosuppressive agents cause Peyer’s patches in the bowel wall to shrink, leading to an alteration of mucosal integrity and hence, the potential for air dissection [2]. Those agents also impair tissue-repair mechanisms, further exacerbating ulceration and bowel necrosis. Additionally, ischemia can facilitate decrease in mucosal integrity, allowing access of intraluminal gas to submucosal tissue planes. In bacterial theory, gas-producing organisms invade the bowel wall. This theory was supported by experimental animal model involving the induction of PCI through the intramural, intraluminal and intraperitoneal injection of Clostridium perfringens, a well-known gas-forming organism [14]. In cases of PCI associated with infectious colitis, multiple organisms were cultured from the stool, blood or both: these included fungal, viral, and bacterial agents [4], many of which are not known to be gas-forming. This tends to implicate some additional etiology for observed PCI [16]. A variant of the bacterial theory suggests that bacterial fermentation of carbohydrates within the gastrointestinal tract leads to excessive gas formation [19] inducing absorption of this gas into the bowel wall. The bacterial theory is also supported by the observation that benign PCI often responds to dietary changes, antibiotic therapy, and oxygen, which is toxic to anaerobic intestinal flora and may create a diffusion gradient across cyst wall, accelerating their deflation [15]. Case 2 may be related to a mechanical theory. In this case, air dissects the bowel wall because of increased intraluminal pressure, which can occur as a result of obstruction, increased gas production with absorption and trapping of the air in the bowel wall [16].

Treatment of pneumatosis cystoides intestinalis ranges from supportive care to laparotomy. PCI is often benign and only follow-up is warranted. Surgery is generally indicated in patients with severe pain – see Case 2, rectal bleeding, fever – see Case 1 or an evidence of ischemic bowel. The decision to proceed with explorative laparotomy must be based on the thorough analysis of a detailed history, physical examination, laboratory tests, and radiological studies [9].

References


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