Zbigniew Antosz[1], Maciej Zaniewski[2]

Pneumatosis Cystoides Intestinalis – a Report of Two Cases

[1]Division of Pathology, Regional Specialistic Hospital, Tychy,
[2]Department of Surgery, Silesian Medical University, Regional Specialistic Hospital, Tychy

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 Vernoi 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 jejunomicrostomy 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.

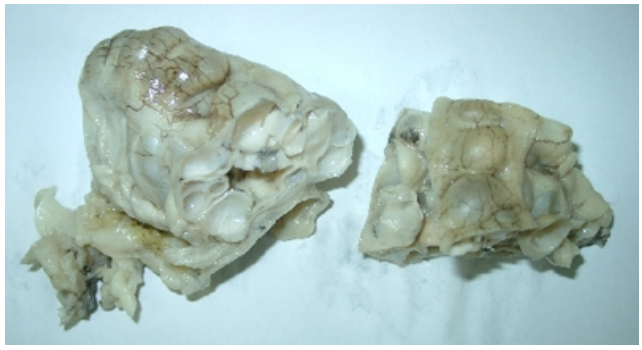

Fig. 1. Cystic cavities in all layers of the large bowel wall.

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).

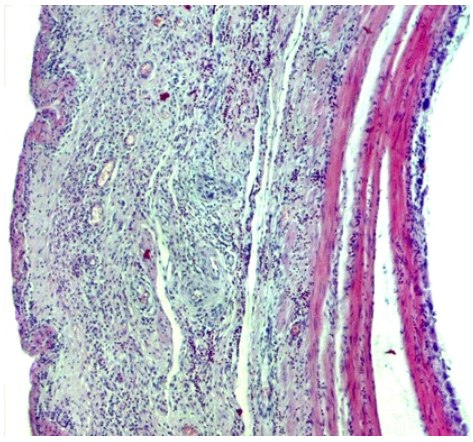

Fig. 2. Subserosal inflammation and fibrosis. HE. Magn. 40x.

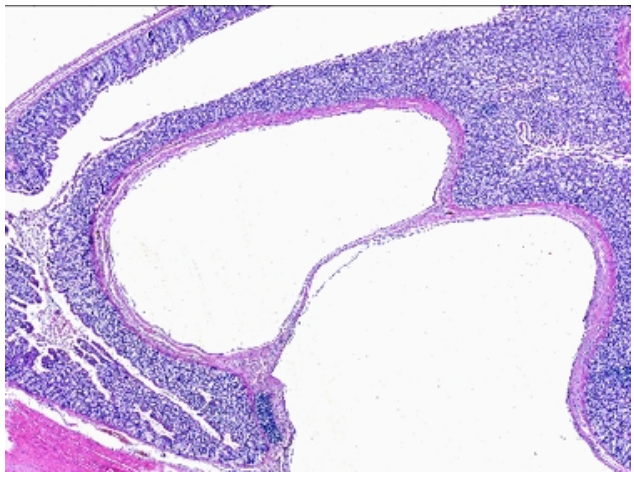

Fig. 3. Cystic cavities in mucosa. HE. Magn. 20x.

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 "pseudonecrotic" 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

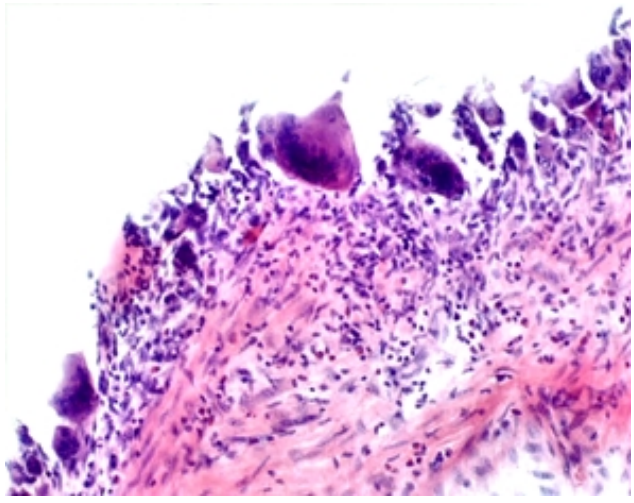

Fig. 4. Multinucleated foreign body giant cell reaction. HE. Magn. 200x.

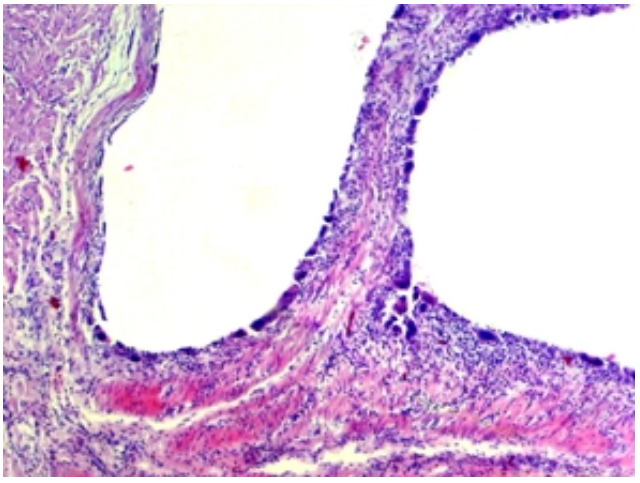

Fig. 5. Lympho-plasmacytic inflammatory infiltrate with fibrosis. HE. Magn. 100x.

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 Case1 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

1. *Andorsky RI:* Pneumatosis cystoides intestinalis after organ transplantation. Am J Gastroenterol 1990, 85, 189-194.
2. *Borns PF, Johnston TA:* Indolent pneumatosis of the bowel wall associated with immature suppressive therapy. Am Radiol 1973, 16, 163-166.
3. *Cabrera GE, Scopelitis E, Cuellar ML, Silveira LH, Mena H, Espinoza LR:* Pneumatosis cystoides intestinalis in systemic lupus erythematosus with intestinal vasculitis: treatment with high-dose prednisone. Clin Rheumatol 1994, 13, 313-316.
4. *Collins CD, Blanshard C, Cramp M, Gazzard B, Gleeson JA:* Case report: pneumatosis intestinalis occurring in association with cryptosporidiosis and HIV infection. Clin Radiol 1992, 46, 410-411.

5. *Day DL, Ramsay NK, Letourneau JG:* Pneumatosis intestinalis after bone marrow transplantation. AJR Am J Roentgenol 1988, 151, 85-87.
6. *Earnest DL, Hixson LJ:* Other diseases of the colon and rectum. In: Gastrointestinal Disease: Pathophysiology, Diagnosis, Management. Sleinsenger MH, Fordtran JS, eds. Philadelphia Pa WB Saunders 1993.
7. *Galandiuk S, Fazio VW:* Pneumatosis cystoides intestinalis: a review of the literature. Dis Colon Rectum 1986, 29, 358-363.
8. *Galandiuk S, Fazio VW, Petras RE:* Pneumatosis cystoides intestinalis in Cronh's disease: report of two cases. Dis Colon Rectum 1985, 28, 951-956.
9. *John A, Dickey K, Fenwick J, Sussman B, Beeken W:* Pneumatosis intestinalis in patient with Cronh's disease. Dig Dis Sci 1992, 37, 813-817.
10. *Khouri MR, Levine MS, Dabezies M, Saul SH:* Benign pneumoperitoneum in a patient with celiac sprue. J Clin Gastroenterol 1989, 11, 70-72.
11. *Knechtle SJ, Davidoff AM, Rice RP:* Pneumatosis intestinalis: surgical management and clinical outcome. Ann Surg 1990, 2, 160-165.
12. *Kobayashi T, Kobayashi M, Naka M, Nakajima K, Momose A, Toi M:* Response to ocreotide of intestinal pseudobstruction and pneumatosis cystoides intestinalis associated with progressive system sclerosis. Intern Med 1993, 32, 607-609.
13. *Koss LG:* Abdominal gas cysts (pneumatosis cystoides intestinorum hominis). Arch Pathol 1952, 53, 523-549.
14. *Meihoff WE, Hirschfield JS, Kern F:* Small intestinal scleroderma with malabsorption and pneumatosis cystoides intestinalis. JAMA 1968, 10, 854-858.
15. *Mirables M, Hinojosa J, Alonso J, Berenguer J:* Oxygen therapy in pneumatosis coli: What is the maximum oxygen requirement? Dis Colon Rectum 1983, 26, 458-460.
16. *Pietrese AS, Leong AS, Rowland R:* The mucosal changes and pathogenesis of pneumatosis cystoides intestinalis. Hum Pathol 1985, 16, 686-688.
17. *Polinsky MS, Wolfson BJ, Gruskin AB et al:* Development of pneumatosis cystoides intestinalis following transperitoneal renal transplantation in a child. Am J Kidney Dis 1984, 4, 419.
18. *Ramos AJ, Powers WE:* Pneumatosis cystoides intestinalis. Report of a case. Am J Roentgenol Radium Thor Nucl Med 1957, 77, 678-683.
19. *Reyna R, Soper RT, Condon RE:* Pneumatosis intestinalis: report of twelve cases. Am J Surg 1973, 125, 667-671.
20. *Reynolds HL, Gauderer MWL, Hrabovsky EE, Shurin SB:* Pneumatosis cystoides intestinalis in children beyond the first year of live: manifestations and management. J Pediatr Surg 1991, 26, 1376-1380.
21. *Sachse RE, Burke GW 3rd, Jonas M, Milgrom M, Miller J:* Benign pneumatosis intestinalis with subcutaneous emphysema in a liver transplant recipient. Am J Gastroenterol 1990, 85, 876-879.
22. *Sackier JM, Smith EJ, Wood CB:* Cystic pneumatosis in coeliac disease. Gut 1988, 29, 852-855.
23. *Silliman CC, Haase GM, Strain JD et al:* Indication for surgical intervention for gastrointestinal emergencies in children receiving chemotherapy. Cancer 1994, 74, 203-206.
24. *Smith BH, Welter LH:* Pneumatosis intestinalis. Am J Clin Pathol 1967, 48, 455-465.
25. *Suarez V, Chesner IM, Price AB, Newman J:* Pneumatosis cystoides intestinalis. Histological mucosal changes mimicking inflammatory bowel disease. Arch Pathol Lab Med 1989, 113, 898-901.
26. *van Leeuwen JCJ, Nossent JC:* Pneumatosis intestinalis in mixed connective tissue disease. Neth J Med 1992, 40, 299-304.
27. *West KW, Rescorla FJ, Grosfeld JL, Vane DW:* Pneumatosis intestinalis in children beyond the neonatal period. J Pediatr Surg 1989, 24, 818-822.

Address for correspondence and reprint requests to:
Zbigniew Antosz M.D.
Division of Pathology
Regional Specialistic Hospital
Edukacji 102, 43-100 Tychy