REVIEW ARTICLE

PNEUMATOSIS CYSTOIDES INTESTINALIS - A REVIEW

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Summary: Pneumatosis cystoides intestinalis is a rare disease characterized by presence of multilocular cysts in the gastrointestinal wall. Idiopatic and secondary forms of the disease can be distinguished. There are presented several theories explaining pneumatogenesis in this article. The specific and non-specific symptoms are described. Attention is drawn to the pneumoperitoneum without signs of peritoneal irritation, what is a typical complication of this disease. The suspition of pneumatosis cystoides intestinalis may be based on plain abdominal X-ray, and is usually confirmed by computer tomography or magnetic resonance imaging. The therapy can be conservative or surgical. In conclusion, although pneumatosis cystoides intestinalis is a rare disease, it may represent a problem in differential diagnosis of abdominal pain.

Key words: *Pneumatosis cystoides intestinalis; Pneumoperitoneum; Cysts*

Introduction

Pneumatosis cystoides intestinalis (PCI) is not too a frequent disease characterized by multilocular gas cysts localized in the wall of the alimentary tract. The rupture of the cysts leads to pneumoperitoneum in the absence of the signs of peritoneal irritation, which is considered to be pathognomic for this disease. Purpose of this paper is to draw attention to pneumatosis cystoides intestinalis – a rare entity, but an important one for clinical practice, futher to show recent view on this problem and notice the supposed etiology, clinical presentation and treatment.

Etiology and pathogenesis

According to Kreiss et al. (33) PCI can be clasified either the idiopathic with unknown etiology (15%) or the secondary one (85%), in which the mechanism of cysts origin has been explained. Several theories elucidating the pathogenesis of this disorder have been proposed.

Mechanical theory

The mechanical theory explains the pathogenesis of pneumatosis by physical factors. Two pathogenetic mechanisms have been proposed: air leakage from lung interstitium to mediastinum, retroperitoneum, mesenterium and intestinal wall, and leakage of intraluminal gas through gaps in intestinal mucosa. The first mechanism has been proposed in patients with chronic obstructive lung disease or with other illness of the respiratory system (48). A rise in intraalveolar pressure leads to alveolar rupture and leakage of air into lung interstitium. Air from lung interstitium is thought to dissipate via mediastinum, retroperitoneum and mesenterium into the gut wall (37). The second mechanism is represented by increase of intraluminal bowel pressure, which in connection with damage of the mucosa leads to intramural penetration of gas. This can explain the fact that PCI is often present in patients who have gastrointestinal disease – peptic ulcer disease, Crohn's disease (24,48) or necrotizing enterocolitis (11,27,32). Necrotizing enterocolitis with PCI has been induced experimentally by arterial and lymphatic ligation (46). Some others (41) explain PCI as a consequence of reparation after bowel ischemia.

Bacterial theory

Bacterial theory explains the pathogenesis of PCI by bacterial infection. This infection either damages the intestinal wall with subsequent intramural penetration of gas, or produces gas, which then penetrates into the gut wall. Gas can also enter the lymphatic vessels and cause their dilatation. This theory is supported by experiments in rats, where PCI was induced by Clostridium perfringens (52). The microorganisms playing role in origin of PCI are Clostridium difficile, cytomegalovirus (44) or Clostridium perfringens (7).

Immunopathological inflammatory reaction

Based on observation by Holl et al. (22) and their demonstration of histiocytes and foreign-body giant cells present in the afflicted part of the bowel, immunopathological inflammatory reaction has been proposed as a cause of PCI. The presence of monocytes and similar mononuclear cells has been confirmed by Gagliardi et al. (19).

PCI after bone marrow transplantation

PCI has been also described after bone marrow transplantation (6). The effect of long term steroid use, infection, immunosuppression, graft-versus-host disease are thought to cause disorder in these cases (36).

PCI in connective tissue disease

The increase of PCI incidence in patients with a connective tissue disease has been observed (2,10,21,30,34). PCI in these patients is probably caused by damage of the gut wall primary by this illness or secondary owing to the ischemia after failure of vessels supply.

Failure of activity hydrogen metabolizing bacteria

PCI is characterized by high level of breath hydrogen, patients with PCI excrete more hydrogen than others. Clinical features of PCI may be in consequence of abnormal hydrogen metabolism. In normal subjects hydrogen is consumed by methanogenic and sulfatereducing bacteria. The activity of these bacteria is missing in patients with PCI. This leads to the intraluminal gas accumulation, to an increase of intraluminal pressure and thus to intramural gas penetration (12,13). The mechanism just described can explain cysts origin. According to Levitt et al. (35) the hydrogen hyperproduction is only the initial reason for cysts origin. Their futher persistence is caused by nitrogen and oxygen, which diffuse from blood (35).

Clinical presentation

Presence and character of symptoms

The character of symptoms is dependent on the localization of PCI and on presence or non-presence of basic disease. Symptoms, which can appear, are either non-specific or specific ones. Abdominal distension (29,34), diarrhoea (10,19,29,39), abdominal pain (10,39), constipation (19,39), mucus discharge (19,39), hematemesis (34), rectal bleeding (19,39), meteorism (14) and weight loss (10) belong to non-specific symptoms. Among the specific symptoms there belong cysts, which can be source of origin of invagination (1) or volvulus (5) and can cause interception of motility and the mechanical obstruction (26). It is especially necessary to draw attention to cysts ruptures, which lead to pneumatosis specific complication – pneumoperitoneum without alarming signs of peritoneal irritation (16,23,26,28,31). Pneumoretroperitoneum develops by cyst rupture in retroperitoneal part of bowel. The mentions of pneumoretroperitoneum are repeated in the literature (28,31,37). During these complications patients usually complain of non-specific abdominal disorder. Pneumoperitoneum diagnosed on abdomen X-ray examination is followed by laparotomy when perforation of alimentary tract in common location is not found (3). Clinical course of PCI may be benign, however also alarming and requiring surgical intervention (8).

Localization of damage

Occurence of PCI was described in right colon (21), further in transversum (6) or left colon (18,19). Moote et al. (40) conclude that sigma has predisposition to the occurence of the illness. The cysts in sigmoid localization cause sigmoid colon redundancy by affecting sigmoid mesentery. Rectum is usually spared (49). There is also described the incidence of PCI in other parts of alimentary tract - small intestine (35,42), stomach (4,6). Small intestine infliction can be connected with malabsorption (39) or with coeliac disease (45), the gastric infliction is unusual (15).

Age and sex

Beyond infancy the PCI is rare (44). In Bertram's et al. (7) opinion the period with the most frequent incidence of PCI is age between 30 and 50 years, a clear sexual predominance doesn't exist.

Diagnostics

When PCI suspected the first examination is plain radiography of abdomen (26,51). There are seen the cysts in the bowel wall or free air under diaphragm in the case of their rupture and pneumoperitoneum appearance. The next diagnostic method can be represented by barium enema examination (7,40). Further it is possible to complete other investigations, which are more sensitive but also more expensive. Sonography that is able to diagnose the cysts is according to some authors (20,51) suitable method, too. Computer tomography remains the most successful technique for initial diagnosis and subsequent follow up (17,51). Its disadvantage is high radiation stress and financial severity. It is also possible to use magnetic resonance in PCI diagnostics (43). Furthermore, but less frequently, PCI can be diagnosed by other ways, as diagnostic laparoscopy (38), endoscopical methods (15,18,34,51) and H_2 test, which enables to detect higher breath level of hydrogen by patients with PCI (12).

Therapy

In asymptomatic patients with PCI no special therapy is recommended (9,18). If a basic disease is present, then it is necessary to treat it and secondary cysts regression is usually observed. PCI therapy could be conservative or surgical one.

Conservative therapy

Conservative therapy can be causal or symptomatical. The causal therapy includes ways suppressing supposed etiological mechanisms. Inhibition principles of these mechanisms consist either in restriction of intestinal gas producting microflora – administration of antibiotics, es-

pecially metronidazol (7,29,31,47), or in inhibition of process leading to the hydrogen hyperproduction - hyperbaric oxygen inhalation (7,31,42). In another way of treatment there is possible to include a diet low in flatulence-producing carbohydrates (14), parenteral nutrition (29,31), endoscopical puncture and cysts sclerotherapy (25), therapy with long-acting somatostatin analogue (30). Symptomatical therapy suppresses single symptoms (19) – as pain, constipation, diarrhoea.

Surgical therapy

Patients with pneumoperitoneum without signs of peritoneal irritation when diagnosis of PCI is known are not necessary to operate (23,44), it is sufficient enough to observe them (23). Surgery is indicated only in fulminant cases (3). The most frequent surgical solution is gut resection (7) or limited colectomy (50).

Conclusion

To conclude: pneumatosis cystoides intestinalis is a rare entity with uncomplicated recognition by modern diagnostic methods. The practical importance of this paper is to inform about this problem and thus enable to avoid the laparotomy in patients suffering from PCI with pneumoperitoneum without signs of peritoneal irritation.

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