Review

Pneumatosis cystoides intestinalis (PCI)

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Abstract. Pneumatosis cystoides intestinalis (PCI) is a rare disease usually occurring in association with a large variety of gastrointestinal (GI) and non GI conditions in the majority of cases, although idiopathic PCI is also known to occur. There are two theories regarding the development of these intramural gas cysts – the mechanical and bacterial theories. PCI usually runs a benign course, although fulminant PCI can be present both in infants and adults. The importance of this condition for the surgeon lies in its early recognition, in order to prevent unnecessary surgical intervention, especially when pneumoperitoneum without clinical evidence of peritonitis is encountered. Oxygen therapy has been shown to lead to regression of PCI, although recurrences have been reported. Elemental diets and antimicrobial agents have provided symptomatic relief in a few reported cases. The association of PCI with a wide variety of conditions leads us to conclude that PCI may not be a disease in itself, but a sequel to these varied conditions.

Introduction

Pneumatosis cystoides intestinalis (PCI) is a rare condition characterised by the presence of multiple gas-filled cysts within the wall of some part of the gastrointestinal tract, and has been variously described as peritoneal pneumatosis, intestinal gas cysts, pneumatosis cystoides intestinorum, peritoneal pneumatosis or cystic lympho-pneumatosis. Mayer first coined the term “pneumatosis cystoides intestinorum” in 1825, although the first descriptions of the disease were by du Vernois in France and John Hunter in England in the 18th century [1].

Amongst the most extensive reviews of this condition was that published by Koss in 1952; he described the majority of lesions being small bowel cysts, 58% being associated with lesions of the “pyloric area”, the disease affecting males more frequently than females (ratio 3:5:1), peak age of occurrence between 30 and 60 years [1]. Forgacs reported an equal incidence in both sexes [2].

Pathogenesis

Various theories have tried to explain the mechanism of formation of the cysts.

The mechanical theory postulates that gas is forced into the bowel wall by one of several mechanisms [1, 3, 4]; (a) breach in bowel mucosa as occurs in ulceration – supported by Koss’ observation of pyloric stenosis in 55% of cases; (b) direct trauma to bowel wall following endoscopic procedures, enteric tube placement, or blunt abdominal trauma [5–7]; (c) anastomoses – following jejunoileal bypass [8, 9]; (d) obstruction – increased intraluminal pressure and increased intraluminal gas, along with increased peristalsis proximal to the site of obstruction may lead to penetration of gas through minute mucosal defects [1, 10, 11]; (e) increased pulmonary pressure with alveolar rupture and dissection of gas through the mediastinum, along the great vessels to the retroperitoneum, perivascular space in the mesentery to the bowel wall – in chronic obstructive pulmonary diseases and cystic fibrosis [12–14].

The bacterial theory is supported by the high hydrogen content of these cysts, a product of bacterial metabolism and not a product of mammalian cells [4, 15].

Yale et al. [16–18] experimentally reproduced PCI after intraluminal, intramural and intraperitoneal injection of pure cultures of Clostridial organisms. However, the absence of an inflammatory reaction around these cysts and the rarity of peritonitis associated with pneumoperitoneum in patients with PCI are sufficient arguments against a bacterial cause [1, 19, 20].

Other theories suggested include the biochemical and dietary mechanisms, which postulate that increased lactic acid levels due to disturbed carbohydrate metabolism results in decreased carbon dioxide and oxygen resorption, with cyst formation [1, 4]. The deficiency of the enzyme disaccharidase in infants has been suggested, resulting in incomplete carbohydrate digestion, which, along with bacterial fermentation, produces large amounts of gas, predisposing to PCI [21].
The results of several analyses of cyst gas reveal high levels of hydrogen and nitrogen, small amounts of nitrous oxide, carbon dioxide and traces of butane, isobutane, propane, methane, ethane and argon [2, 15, 22]. The increased hydrogen content of cyst gas favours the bacterial theory, hydrogen being a result of bacterial metabolism [15], and is against the mechanical theory, hydrogen being present in only minute quantities in alveolar air [4].

Normally, gas-containing cavities within the body but not in communication with the atmosphere gradually deflate and eventually disappear. Why, then, do these cysts persist? A direct communication with the lumen of the bowel has never been demonstrated [1, 2, 4]. Their persistence suggests that they are replenished at a rate that equals or exceeds the rate of absorption. Oxygen therapy is based on the principle of alteration of the balance between diffusion of gases into and out of the cysts favouring their absorption [2, 20, 22].

**Pathology**

Macroscopically, multiple sessile or pedunculated gas cysts are present on the serosal and mucosal surface of the involved bowel segment, ranging in size from a few millimetres to several centimetres [1, 4, 20]. The submucosal cysts give the bowel a spongy, crepitant consistency. The trapped gas is under pressure, puncture resulting in its release with an audible hiss. Serosal cysts are predominantly on the mesenteric border [4].

Microscopically, the cysts are found in the serosa, subserosa, mucosa and submucosa, the muscular layer being least affected [1, 20], (Fig. 1). The cysts are lined by endothelial cells with large multinucleated foreign body giant cells and macrophages in close proximity [1, 2, 4, 19, 20]. In clusters, the cysts are separated from one another by thin bands of hyaline connective tissue sparsely infiltrated by mononuclear cells, predominantly lymphocytes [1].

Earlier reports of this condition described these cysts to be lymphatic vessels distended by gas. Koss [1] suggested that progressive fibrosis eventually leads to decrease in size and disappearance of cysts.

**Classification and incidence**

PCI has adult and infantile forms; the infantile form runs a fulminant course as acute necrotising enterocolitis, with a poor prognosis. A much better classification divides PCI into benign and fulminant forms, since the fulminant type has also been observed in adults, associated with pseudomembranous colitis and Crohn’s disease [3, 21].

PCI can also be classified as idiopathic (primary) or secondary, the latter occurring in association with a wide variety of gastrointestinal (GI) and non GI conditions constituting 85% of PCI [4].

The conditions associated with PCI are listed in Table 1.

PCI seen following transplantation may be due to prolonged steroid therapy leading to depletion of submucosal lymphoid tissue – the denuded Peyer’s patches may produce mucosal defects, permitting entry of gas into the bowel wall [27, 30]. In cystic fibrosis, PCI is seen in the later stages of the disease, indicating severity of the existing pulmonary disease [14].

PCI has been reported as occurring more frequently in the small bowel, but large bowel involvement is being increasingly reported [4, 21, 39, 40].

PCI involving the left colon is usually idiopathic, secondary disease usually affecting the small bowel and ascending colon [4, 41].