CASE REPORT

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Idiopathic Pneumatosis Cystoides Intestinalis in A 35-Year-Old Man Who was Diagnosed Incidentally

Rastlantısal Olarak İdiyopatik Pnömatozis Sistoides İntestinalis Tanısı Konulan 35 Yaşındaki Hasta

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ABSTRACT

Signs and symptoms of pneumatosis cystoides intestinalis (PCI) vary depending on the localization of cystic lesions in the gastrointestinal tract. Since PCI is not a frequent disease, diagnosis is usually made incidentally. PCI mostly occurs as a result of various clinical conditions. Principally, treatment is managed according to the cause. It is usually benign and rarely causes serious complications. In this case report, we introduce a 35-year-old man who was admitted to the hospital with non-specific symptoms.

Keywords: Idiopathic, pneumatosis cystoides intestinalis, incidentally

ÖZ

Pnömatozis cystoides intestinalis'in (PCI) belirti ve semptomları, gastrointestinal sistemdeki kistik lezyonların lokalizasyonuna göre değişir. PCI sık görülen bir hastalık olmadığından tanı genellikle tesadüfen konur. PCI çoğunlukla çeşitli klinik durumların bir sonucu olarak ortaya çıkar. Prensip olarak tedavi, nedene göre yönetilir. Genellikle iyi bir seyri vardır, PCI nadiren ciddi komplikasyonlara yol açar. Bu olgu sunumunda, hastaneye non-spesifik semptomlarla başvuran 35 yaşında bir erkek hastayı sunuyoruz.

Anahtar Kelimeler: İdiyopatik, pnömatozis sistoides intestinal, rastlantısal

Introduction

Pneumatosis cystoides intestinalis (PCI) is a very uncommon entity. This clinical picture may be diagnosed incidentally in radiologic or endoscopic examinations performed for different clinical indications. Usually, submucosal, intramucosal or subserosal layers of gut are occupied with single or multiple cysts full of air bubbles. There are very few reports of PCI occurring in the duodenum and rectum. Although the etiopathogenesis of PCI is not known precisely, it is thought to arise due to inflammation, physical damage of the intestinal mucosa, malnutrition, dysbacteriosis in which the balance of beneficial-harmful bacteria is impaired, gastrointestinal dysmotility and immune dysfunction (1,2,3). It may cause intra-abdominal free air. In 85% of the cases, PCI is associated with another pathology. Clinical findings are generally not clear but rarely PCI presents with serious clinical conditions like bleeding or perforation. In this study, we present a 35-year-old man incidentally diagnosed as PCI.

Case Report

A 35-year-old patient complaining moderate to severe abdominal pain with flatulence and epigastric pain was admitted to gastroenterology department of our institution. He did not have any chronic disease, any medication, any illicit or over-the-counter drug usage. He was suffering from abdominal pain and intermittent constipation for six years. The patient especially reported that abdominal discomfort was proportional to the amount of food intake. Physical examination was normal except epigastric sensitivity and abdominal distension with palpation. Biochemical laboratory tests were all normal.

Upper gastrointestinal endoscopy revealed hyperemic antral gastropathy. Five bubble-like and grape shaped submucosal cystic polypoid lesions in the distal part of descending colon were reported in colonoscopy. They were 10-30 mm in diameter and resembling like lipomatous polyps or gastric fundal varices (Figure 1). There were also

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two distinct polyps with a diameter of 10 mm below and above this area. Smooth and bluish colored polypoid mucosa differing from adjacent non-polypoid tissue was noteworthy (Figure 1). The patient's history, laboratory results and physical findings were not consistent with portal hypertension and there was no pillow sign indicating a lipomatous polyp.

The bubble-like appearance made us think to use a sclerotherapy needle to test whether these subepithelial lesions contained gaseous material. Polyps suddenly became deflated when punctured and the mucosa turned to normal color like adjacent area. White epithelial ring surrounded the deflated polypoid region (Figure 2). This sign also strengthened our diagnosis of submucosal PCI. Later, we deflated the other ones with sclerotherapy needle. In the upper parts of the descending colon, the other whitish epithelial rings indicating the spontaneous explosion of these submucosal cystic polyps also were noted. In addition, written consent was obtained from the patient.

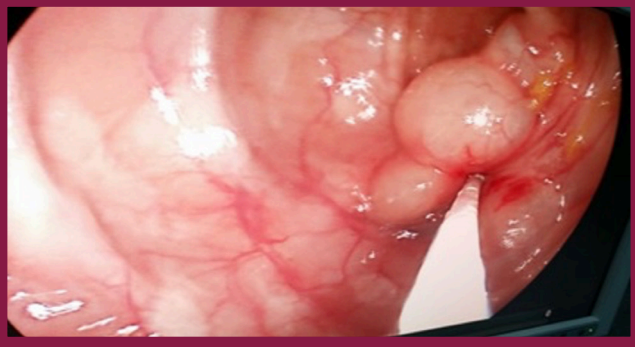

Figure 1. Deflating of the gas filled cystisc lesions in colonoscopy

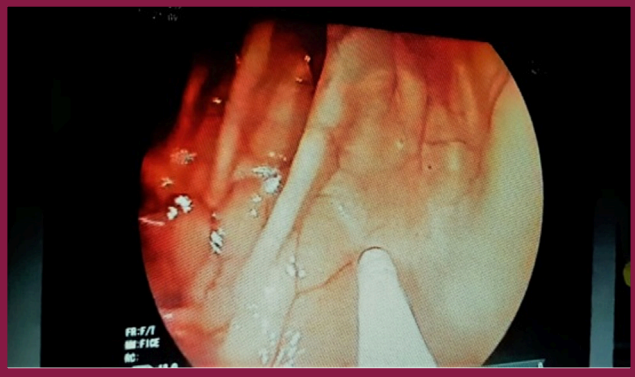

Figure 2. Appearance of the cystic lesion after deflation. Whitish and rim like epithelial ring surrounded the deflated cyst

Discussion

PCI is typically explained as being gas-filled cysts in the intestinal submucosa and/or subserosa. PCI is a rarely seen condition and formerly reported with the incidence of 0.03%. PCI may be encountered in all age groups (1,2,3). Since it coincides with other clinical scenarios and some cases have vague symptoms, determining the exact incidence is very difficult. First description in literature was made by Olmsted and Madewell (4) in the 18th century and then subcategorized by Koss (5). It is predominant in males between the ages of 30 and 50 years.

PCI may present with different clinical conditions. It usually associates with severe clinical entities and may lead to complications including acute abdominal pain, gastrointestinal bleeding, perforation, pneumoperitoneum, and volvulus (6,7). Nevertheless, it may present with benign situations as in this case. Prognosis usually correlates with the underlying disease.

Because of the wide spectrum of clinical settings and sometimes having silent clinical picture, PCI can be underdiagnosed. The exact pathophysiology of PCI is not entirely explained. This gray zone is probably multifactorial. PCI may be idiopathic (15%) or secondary (85%) to a lot of gastrointestinal and non-gastrointestinal problems (8). Secondary gastrointestinal conditions are intestinal ischemia, infarction and perforation, inflammatory bowel diseases, AIDS enteropathy, peptic ulcer disease, Coronavirus disease-2019 tuberculosis, and C. difficile infections. Secondary non-gastrointestinal disease may include chronic obstructive pulmonary disease, cystic fibrosis, diabetes mellitus, anti-diabetic medications like acarbose or voglibose, scleroderma, lymphoproliferative disorders and bone marrow or solid organ transplantation (8). During infancy, most causes are secondary to necrotizing enterocolitis. Our patient did not have any comorbidity.

Mechanical, bacterial, and biochemical explanations have been put forward to explain the exact pathogenesis of PCI. In line with the mechanical theory, gas enters into the intestinal wall from either the luminal surface through breaks in the mucosa or through the serosal surface by tracking along mesenteric blood vessels (9). In bacterial theory, PCI has been reproduced experimentally by injecting the gas-forming bacillus *Clostridium perfringens* into the bowel wall of rats. Further supporting the bacterial theory is that PCI may resolve following treatment with antibiotics (10). Finally, elemental diets have been reported to improve PCI presumably by removing substrate for the production of gas by bacteria (11). Our patient also noted that his abdominal discomfort was proportional to the amount of food intake and this situation may be supportive of bacterial theory of

PCI. Food content way also induce gas forming bacteria in the intestinal tract. Biochemical theory depends on the idea that luminal bacteria produce large amounts of hydrogen gas through the fermentation of mainly carbohydrates and other kind of foods, and hydrogen gas invades mucosal layers. All pathogenetic factors may contribute to the formation of PCI.

PCI can locate in any part of the gastrointestinal tract except the stomach. The small bowel is the most affected region and the colon is the second one. In some cases, both regions may become affected. The cysts may involve all three layers (mucosa, submucosa, subserosa). Subserosal cysts are more commonly seen in small intestinal pneumatosis while submucosal cysts are more commonly seen in colonic pneumatosis as in this case (12). There was not any evidence of small intestinal pneumatosis in plain abdominal X-ray (Figure 3). Endoscopic procedure may report bubble-like pattern, grape or beaded circular forms, linear or cobblestone gas formation and irregular forms. The bubble-like lesions need to be differentiate from intestinal polyps (1,2). Our case had bubble-like and grape patterns. Symptoms may vary depending on the region affected. In a series of 919 patients, small intestinal disease associated symptoms were vomiting, abdominal pain, abdominal distention, weight loss, and diarrhea. In patients with colonic pneumatosis, most frequent symptoms were diarrhea, hematochezia, abdominal pain, distention and prolonged constipation. Other symptoms were flatulence, loss of appetite, and tenesmus, which were reported in our case (12).

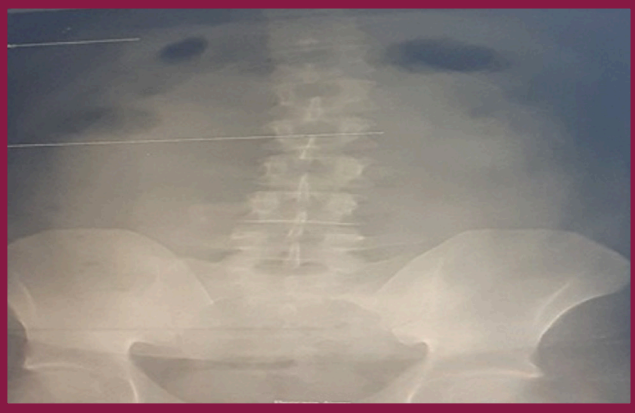

Figure 3. X-ray graphy of the abdomen after colonoscopy

Conclusion

Besides non-specific gastrointestinal symptoms, life-threatening conditions may accompany PCI at presentation. This clinicopathological entity should be taken into consideration in clinical practice.

Ethics

Informed Consent: Written consent was obtained from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: İ.K., Y.Y., M.K., A.T., Concept: İ.K., Y.Y., M.K., A.T., Design: İ.K., Y.Y., M.K., A.T., Data Collection or Processing: İ.K., Y.Y., M.K., A.T., Analysis or Interpretation: İ.K., Y.Y., M.K., A.T., Literature Search: İ.K., Y.Y., M.K., A.T., Writing: İ.K., Y.Y., M.K., A.T.

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