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Case Report

Pneumatosis cystoides intestinalis: A rare cause of intraabdominal free air

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ABSTRACT

Pneumatosis cystoides intestinalis is a rarely seen disease characterized by cysts filled with multiple gases in the wall of the small or large intestine. Many factors have been suggested in the etiology and pathogenesis, including peptic ulcer, pyloric stenosis, and endoscopic trauma. Because various clinical characteristics and symptoms are observed together, diagnosis is generally difficult. It may be a cause of free air observed on direct radiographs. Treatment is directed at the cause and although there is generally a good course, it can sometimes lead to severe complications. In this paper, we present the case of a 33-year old male being prepared for surgery for pyloric stenosis due to a peptic ulcer. In the preoperative tests, because acid was determined within the abdomen and free air below the diaphragm, pneumatosis cystoides intestinalis was considered in the differential diagnosis. Definitive diagnosis was made during the operation and surgical treatment was applied.

Keywords: Ascites, pyloric stenosis, intraabdominal free air, pneumatosis cystoides intestinalis

INTRODUCTION

Pneumatosis cystoides intestinalis (PCI) is a rarely seen disease that is characterized by cysts filled with multiple gases in the subserosa or submucosa of the small or large intestine. It may cause intra-abdominal free air (1). In 85% of the cases, PCI is seen secondary to another pathology. These secondary pathologies are pyloric stenosis, peptic ulcer, appendicitis, ischemic and inflammatory intestinal diseases, diverticular disease, sigmoid volvulus, obstructive pulmonary disease, connective tissue diseases, various parasitic diseases of the intestine, and surgical or endoscopic trauma (2). Because PCI is seen with many different clinical findings and symptoms, most cases are found incidentally. Therefore, when free air is seen in the abdomen, it should be considered in the differential diagnosis. Generally, treatment includes the removal of the concomitant secondary pathology and includes resection of the involved segment of the intestine. Although it can lead to serious complications, the course is generally good (3). In this paper, a case of PCI seen together with pyloric stenosis due to acid and a peptic ulcer is discussed.

CASE PRESENTATION

A 33-year-old male patient presented at our clinic with complaints of nausea, severe vomiting, epigastric pain, and swelling. Questioning revealed a 10-year history of dyspeptic complaints and that he had been diagnosed endoscopically with a duodenal ulcer 5 years previously. Although medical treatment was administered in the subsequent period, endoscopy and biopsy procedures were applied on further two occasions as the complaints continued. Despite a good appetite, the patient had lost 30 kg in weight over 10 years. He was cachectic in appearance and there was succussion splash and sensitivity in the epigastric region on physical examination. No pathology was determined in the laboratory tests. On the postero-anterior pulmonary radiograph, free air was determined below the right diaphragm (Figure 1).

In the esophago-gastro-duodenoscopy that was applied, a large amount of solid and liquid food remnants were seen in the stomach despite 12 h of fasting. Grade A esophagitis was determined, and loosening of the lower esophageal sphincter, antral gastritis, and pyloric stenosis to a degree that would not permit the passage of the endoscope were observed. The biopsy taken on the basis of an ulcer was reported as chronic gastritis. On the abdominal ultrasonography (USG) applied to the patient who did not have findings of acute abdomen, widespread free fluid was observed. On the computed tomography (CT) imaging of the whole abdomen, the stomach was full, there was increased thickness of the pyloric wall, and widespread free fluid was observed in the abdomen. In the parasynthesis applied under USG, acid-like fluid was aspirated. No pathological finding was determined on examination of this fluid. After 2 days of hydration and nasogastric decompression, the patient was admitted for surgery. When the abdomen was surgically entered, approximately 1 L of acid fluid was aspirated. In the subsequent exploration, the stomach was dilated and hypotonic to an advanced degree and scar tissue and stenosis in the pylorus were observed. As additional pathology, multiple cystic structures of varying sizes were noticed that were located subserosally involving a 20 cm colon segment starting from the splenic flexura towards the distal region (Figure 2). Additional to the distal subtotal gastrectomy because of the

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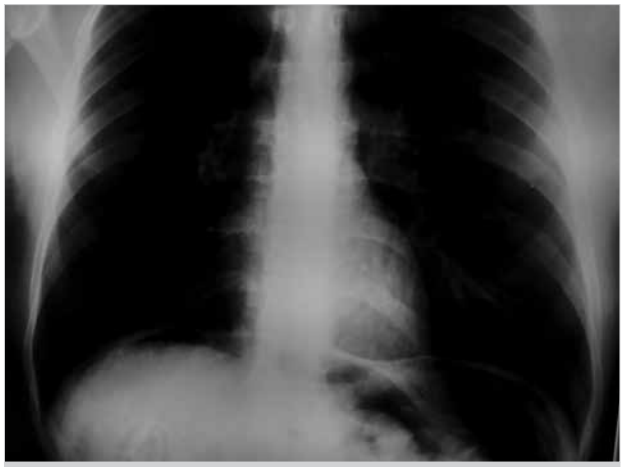

Figure 1. Free air seen under the right diaphragm on the posteroanterior chest radiography

Figure 2. Gas-filled cysts seen in the colon serosa

pyloric stenosis, a resection of the colon segment where PCI was determined and end-to-end anastomosis were applied. In the postoperative follow-up, no complications developed and the patient was discharged on the 7th day.

The pathology examination of the excised material determined ulceration in the pylorus, gastritis in the antrum, intermittent smoothing of the mucosa in the colon segment, and air-filled cysts ranging from a few mm to a few cm on the serosal surface. At the postoperative 3-month follow-up examination, the patient had put on 10 kg in weight and none of the preoperative complaints remained.

DISCUSSION

Pneumatosis cystoides intestinalis is seen in the small intestine in 42% of cases, in the colon in 36%, and in both the colon and small intestine and extra-intestinal tissues such as the mesentery and omentum in 22% (4). In the current case, the air cysts were localized in the colon subserosa and mesentery. Pneumatosis cystoides intestinalis is seen most often in those aged 30–50 years and in males (5). The case presented here was that of a 33-year-old male. The incidence of PCI has been reported as 0.37% (6) and although the etiology is unknown, many hypotheses have been suggested. Pneumatosis cystoides intestinalis etiology may be classified as primary (idiopathic) or secondary. The form seen not together with any other intestinal or systemic disease is named "primary" (15%) and that seen together with another disease, "secondary" (85%) (7). These secondary factors are pyloric stenosis, peptic ulcer, appendicitis, ischemic and inflammatory intestinal diseases, diverticular disease, sigmoid volvulus, obstructive pulmonary diseases, connective tissue diseases, nephrotic syndrome, AIDS, transplantation, the use of some medications (chemotherapeutic agents, steroids, sorbitol, lactulose) various parasitic diseases of the intestine, hematological tumors, and surgical or endoscopic trauma (2). The current case was accepted as the secondary form as there was acid accompanied by pyloric stenosis associated with a peptic ulcer and more than one endoscopy and biopsy procedure had been applied.

There is no evident relationship between the severity of the clinical findings and involvement of the PCI disease (4). Pneumatosis cystoides intestinalis is generally asymptomatic, but depending on the localization, non-specific symptoms such as diarrhea, constipation, abdominal pain, distension, meterorism, tenesmus, and bloody or mucous stools or specific symptoms of gas-filled cysts such as invagination, volvulus, or mechanical obstruction may develop. In addition, pneumoperitoneum may result from ruptured cysts without any peritoneal irritation findings (8). When PCI is suspected, simple methods such as direct radiograph (two-thirds of the cases are diagnosed) and USG and advanced diagnostic techniques such as colonoscopy and CT, may be requested (3). Because PCI is seen with a wide range of clinical findings and symptoms, most cases are found incidentally. In the current case, sub-diaphragmatic free air was seen on the direct pulmonary radiograph taken during the preparation for surgery for pyloric stenosis. Because there were no findings of acute abdomen, PCI was considered and definitive diagnosis was made during the operation. Clinically, the presence of intra-abdominal free air with the rupture of subserosal cysts without peritoneal findings is known as "benign pneumoperitoneum" (4). When free air is seen in the abdomen radiologically, attention must be paid to the indications for surgical intervention and there must be good knowledge of the clinical and physical examination findings and the underlying disease of the patient.

The treatment of PCI depends on the spread of the disease and the underlying reason. The clinical findings are corrected. In patients with symptomatic but uncomplicated cases, good results have been reported with wide spectrum antibiotics, metronidazole, prednisone, normobaric or hyperbaric oxygen, somatostatin, and bismuth (3). Surgical treatment is useful in cases where the underlying reason for PCI is an obstructive gastrointestinal disease or an acute abdomen event such as perforation, which leads to peritonitis. In the current case, during the surgical intervention for pyloric stenosis, segmental colon resection and end-to-end anastomosis was applied to a segment of approximately 20 cm with PCI involvement seen in the colon. Recurrences have been reported after treatment (9). In the current case, where an intestinal segment with lesions was removed and pyloric stenosis was corrected, no recurrence was determined throughout a 14-month postoperative period.

Despite surgery, the mortality rates have been reported in complicated cases as 44% (4). In a case of mortality with PCI,

it seemed to be associated with the underlying reason or pathology rather than the PCI itself (10).

CONCLUSION

Pneumatosis cystoides intestinalis is rarely seen and several factors are held responsible in the etiology. Delayed treatment may be a reason for serious morbidity and mortality. When intra-abdominal free air is seen, intestinal perforation or bacterial infections, which can create gas should be considered in the differential diagnosis.

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