

# Pneumatosis Cystoides Intestinalis in Patients with Antinuclear Antibody Negative Systemic Lupus Erythematosus and Dermatomyositis: Report of Two Cases<sup>1</sup>

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Pneumatosis cystoides intestinalis (PCI) occurring in association with collagen vascular disease is an unusual combination that presents with intramural gas in the gastrointestinal tract. We report two cases of PCI, one with antinuclear antibody (ANA) negative SLE and the other with dermatomyositis, with a review of the relevant literature.

**Index words :** Colon, CT  
Colon, perforation  
Computed tomography (CT)  
Pneumoperitoneum

Pneumatosis cystoides intestinalis (PCI) was first described in 1730 as an uncommon condition that is characterized by multiple air-containing cysts in the submucosa or subserosa of the gastrointestinal wall (1). This condition is classified into primary (idiopathic) and secondary types. The latter arises in the presence of underlying causes, most commonly gastrointestinal disorders (2). In rare cases, collagen vascular disease such as progressive systemic sclerosis, systemic lupus erythematosus (SLE) and dermatomyositis can cause PCI (3 - 6). In Korea, there has been only one report of PCI associated with collagen vascular disease, in which the patient was diagnosed with progressive systemic sclerosis (6). Unlike PCI associated with gastrointestinal disorders or neoplasms, PCI occurring in association with collagen vascular disease is treated conservatively, even in the presence of a pneumoperitoneum (4 - 6). Computed to-

mography (CT) is useful for diagnosing PCI and can exclude predisposing intraabdominal conditions, which can assist in the treatment plan (7).

We report two cases of PCI involving the colon in two patients, one with antinuclear antibody (ANA) negative SLE and the other with dermatomyositis.

## Case Report

### Case 1

An 18-year-old girl was admitted complaining of an abdominal pain, fever, nausea and vomiting. She had a 6-year history of progressive arthritis. SLE had been diagnosed 5 years ago and she was being treated with steroid medication. The serologic test was negative for the lupus preparation and antinuclear antibodies. The plain abdominal radiographs showed a honeycomb appearance along the right colon due to extensive gases within the colonic wall (Fig. 1A). A CT scan with a lung window setting (window width: 2,000 HU, window level: -500 HU) revealed the mottled appearance of gas collections in the walls of the ascending and transverse colons (Fig. 1B). After 7 days without treatment, the intramural gases had disappeared spontaneously. There

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was no recurrence during the follow-up period.

### Case 2

A 42-year-old man with dermatomyositis visited the rheumatology clinic for regular check-up. Dermatomyositis was diagnosed 18 months earlier. At that time, the chest CT revealed interstitial lung disease (Fig. 2A) and the patient was prescribed low-dose steroid. The patient had no significant symptoms. The plain abdominal radiographs revealed honeycomb appearance due to extensive gases within the walls of the ascending, transverse and proximal descending colons (Fig. 2B). A CT scan with a lung window setting (window width: 2,000 HU, window level: -500 HU) showed the multiple curvilinear appearance of intramural gas in the colon with a pneumoperitoneum (Fig. 2C). The patient was managed conservatively with high dose corticosteroid pulse therapy. After 5 days, the plain radiographs showed an improvement in the PCI.

After three months, the patient was again admitted to our hospital because of abdominal pain. The abdomen CT again revealed intramural gases in the transverse colon with a pneumoperitoneum. The patient was treated with antibiotics and high-flow oxygen therapy. After one month, he was completely asymptomatic and was discharged with the radiographs showing the resolution of the PCI and pneumoperitoneum.

### Discussion

Pneumatosis cystoides intestinalis (PCI) is a rare condition that manifests as the cystic accumulation of gas in the bowel wall. Two subtypes, primary (idiopathic) and secondary, of PCI have been described (1). A review by Heng *et al.* (2) showed that secondary PCI occurs with several distinctive clinical settings: 1) in premature infants with necrotizing enterocolitis; 2) in adults with obstructive pulmonary disease; 3) in adults and infants with a mechanical bowel obstruction, infection, drug therapy (particularly steroids), and collagen vascular disease; and 4) as an incidental finding on endoscopy.

In most cases, the pathogenesis of PCI is unknown. Two hypotheses, mechanical and bacterial, have been suggested (1). According to the mechanical hypotheses, intestinal intramural air might develop from a retroperitoneal dissection extending down from the mediastinum. It occurs as a result of an alveolar rupture with air dissecting interstitially along the bronchopulmonary bundles to the mediastinum and then retroperitoneally along the vascular supply of the viscera. This dissection of gas can account for the retroperitoneal gas and the pneumoperitoneum when the cysts have ruptured. PCI may also result from the escape of intraluminal gas secondary to the loss of mucosal integrity (8). According to the bac-

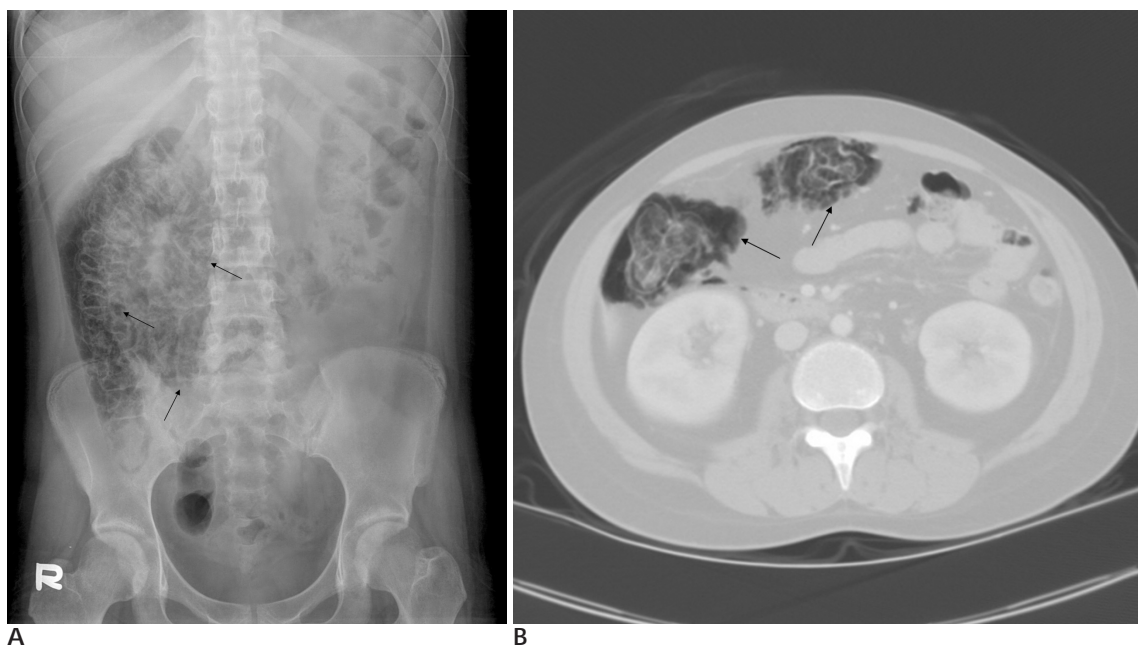
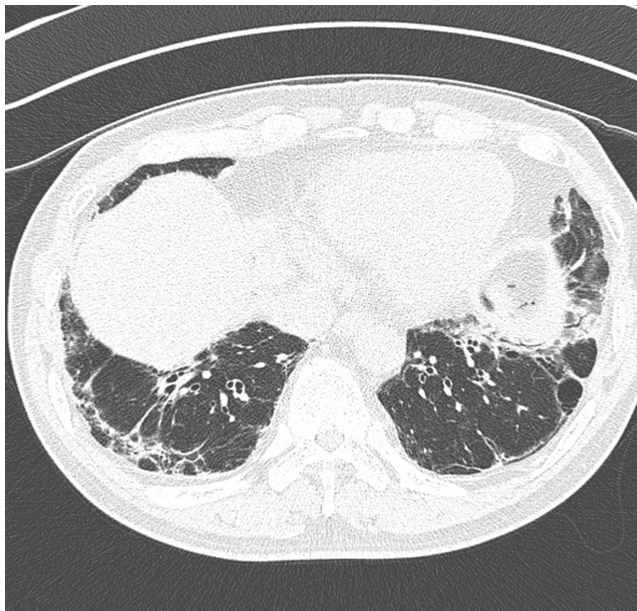


Fig. 1. Case 1: A 18-year old girl with SLE.

A. Plain abdominal radiograph shows the curvilinear collection of intramural gases (arrows) in the right upper abdomen.

B. CT scan with lung window setting (window width: 2,000 HU, window level: -500 HU) shows multiple gaseous cysts (arrows) within the ascending and transverse colons.



A



B



C

**Fig. 2.** Case 2: A 42-year old man with dermatomyositis.

**A.** Chest CT with a lung window setting (window width: 2,000 HU, window level: - 500 HU) shows coarse reticular densities in the subpleural area consistent with interstitial lung disease.

**B.** The plain abdominal radiograph shows the curvilinear collection of intramural gases (arrows) in the ascending, transverse and proximal descending colons.

**C.** Abdominal CT with lung window setting (window width: 2,000 HU, window level: - 500 HU) shows multiple gaseous cysts (arrows) within the transverse colon and pneumoperitoneum (large arrows).

terial hypothesis, increased fermentation by gas-forming bacteria and the subsequent production of a large volume of gas predispose an individual to developing PCI (9). In rare occasions, collagen vascular disease can cause PCI. In 1966, Hughes et al. (10) reported the first case in a patient with collagen vascular disease. Since then, other cases have been described, which have mostly been associated with progressive systemic sclerosis (3, 6). Other collagen vascular diseases that predispose an individual to PCI such as rheumatoid arthritis (3), dermatomyositis (5), and SLE (3, 4) have been reported. In Korea, there has been only one report of PCI associated with collagen vascular disease, in which the patient was diag-

nosed with progressive systemic sclerosis (6).

Several hypotheses for the pathogenesis of PCI associated with progressive systemic sclerosis has been suggested. These include mucosal stretching and fissuring due to a chronic distention and an elevated intraluminal pressure, or from small focal areas of ischemia and ulceration due to vascular compression in the submucosa by fibrous connective tissue deposition (3). In contrast to the extensive fibrosis observed in progressive systemic sclerosis, vasculitis is the mechanism of PCI in cases of SLE. PCI can occur due to focal areas of mucosal ischemia or ulceration as a result of a small vessel occlusion. Lupus vasculitis results in ischemia that affects the mucosa more than

the submucosa (4). There have been only a few reports on PCI occurring in conjunction with dermatomyositis, particularly in adults. The mechanism of PCI in cases of dermatomyositis may involve an intramuscular thrombosis because there is inflammation of the intramuscular arterioles with a thrombus and infarction (5).

The radiological features of PCI vary. The aim of imaging techniques is to differentiate between the primary type, which generally has a favorable prognosis, and the secondary type, the course of which is determined by the underlying disease. In the primary type, PCI is usually located in the submucosal layer of the left hemicolon or its mesentery, and the intramural gas collections are a spherical, well-defined cystic appearance. In the secondary type, PCI is usually limited to the subserosal layer of the stomach, small intestine and right colon, and intramural gas may appear as curvilinear lucencies that circumscribe a loop of the bowel when observed en face and parallel to a long segment when seen lengthwise (4, 7). It is widely accepted that abdominal radiographs and CT can demonstrate the intraluminal gas. However, a recent report suggested that CT shows the cysts quite clearly and also allowed the localization of the affected bowel segments, thereby making CT a more sensitive and discriminating diagnostic tool. Diverticulosis, gas-containing abscess, and gas inclusions or gas layer resulting from coprostasis are potential sources of error related to the CT diagnosis of PCI (7).

After excluding the treatable cause or an occult malignancy, the treatment for PCI in association with collagen vascular disease is largely conservative. Oxygen therapy leads to a relative decrease in the partial pressure of the non-oxygen gases in the blood, which in turn

diffuse out of the cysts leading to resorption. Antibiotics and elemental diets can provide temporary relief. PCI may be complicated by pneumoperitoneum. In contrast to the pneumoperitoneum of other origins, a PCI-related pneumoperitoneum does not usually cause signs of diffuse peritonitis. A pneumoperitoneum can be managed non-operatively if it occurs without peritonitis. The CT demonstration of cysts and the patient's history can help to avoid the need for an unnecessary exploratory laparotomy in patients with PCI (4 - 7).

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