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CASE REPORT

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A case of systemic lupus erythematosus complicated with pneumatosis cystoides intestinalis

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Abstract Pneumatosis cystoides intestinalis (PCI), which is characterized by the presence of multiple gas-filled mucosal, submucosal, or subserosal cysts located throughout the colon and/or small intestine, is an unusual complication of systemic lupus erythematosus (SLE). We report a case of a 33-year-old woman with a 5-year history of SLE with PCI. Her symptoms improved with conservative management. Although PCI is a rare manifestation of SLE, clinicians should be alert to the differential diagnosis of this complication.

Key words Abdominal symptoms · Pneumatosis cystoides intestinalis (PCI) · Systemic lupus erythematosus (SLE)

Introduction

Pneumatosis cystoides intestinalis (PCI) is a relatively uncommon condition characterized by the presence of multiple gas-filled mucosal, submucosal, or subserosal cysts located throughout the colon and/or small intestine.[1,2] The first possible case of PCI was reported by Du Vernoi in 1730, but Meyer in 1925 was the first to use the term PCI.[3,4] There are many conditions associated with PCI.[5–8] They include obstructive pulmonary diseases (bronchial asthma, chronic obstructive lung disease, cystic fibrosis), obstructive intestinal pathology (pyloric or esophageal stenosis, intussusception, volvulus), and trauma by surgery or colonoscopy. In addition, it is well known that PCI is occasionally associated with certain connective tissue diseases, particularly with systemic sclerosis (SSc), but rarely with systemic lupus erythematosus (SLE).[6,9] We report a rare case of PCI in a patient with SLE.

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Case report

A 33-year-old woman with SLE presented to the emergency unit of our university hospital because of abdominal pain. She had no remarkable family history or medical history except for SLE. Her 5-year history of SLE had been characterized by discoid rash, fever, oral ulcer, proteinuria, and leukocytopenia. Treatment with oral prednisolone (PSL) 20 mg/day was started and the dose was gradually tapered to 12.5 mg/day with clinical improvement. Her lupus had been both clinically and serologically inactive for the past several months. She did not have scleroderma or other symptoms suggestive of SSc. On physical examination, her abdomen was soft and slightly distended. She had spontaneous pain and tenderness in the epigastric lesion, but no rebound tenderness. An abdominal radiograph revealed extensive gas within and around the wall of ascending and transverse colon, which was distended by gas and feces (Fig. 1). A diagnosis of PCI was made, and she was admitted to our hospital.

On admission, her body temperature, blood pressure, and pulse rate were 37.0°C, 90/60 mmHg, and 78 beats/min, respectively. Laboratory data revealed white blood cell count of 4750/μl (Neut. 75.4%, Eos. 1.7%, Baso. 0.2%, Mono. 8.8%, Lymph 13.9%), C-reactive protein 0.779 mg/dl, positive antinuclear antibody (1:320) (homogeneous 320, speckled 320), positive anti-DNA antibody (RIA) 15 IU/ml, ferritin 23.0 ng/ml, C3 47 mg/dl, C4 6 mg/dl, CH_{50} 21.8 U/ml. Red blood cell and platelet counts were normal. Anti-RNP antibody, anti-Sm antibody, anti-SS-A antibody, and anti-Scl70 antibody were negative. Computed tomography (CT) of the abdomen showed extensive pneumatosis involving the submucosal and subserosal layers of the ascending and transverse colons (Fig. 2). Barium enema showed an extensive gas layer surrounding the ascending colon, suggestive

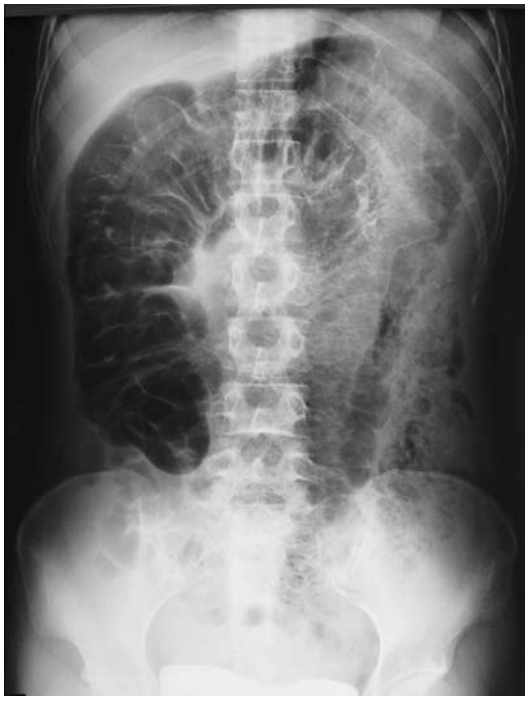

Fig. 1. Abdominal radiograph revealed extensive gas within and around the wall of the ascending and transverse colons, which were distended by gas and feces

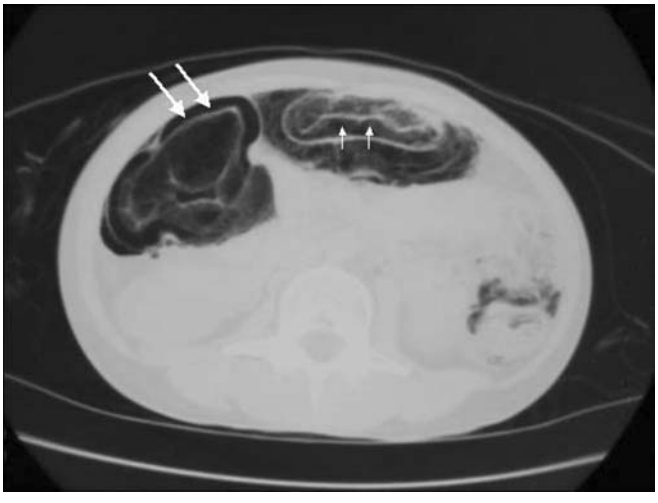

Fig. 2. Computed tomography (CT) of the abdomen showed extensive pneumatosis involving the submucosal (*small arrows*) and subserosal (*large arrows*) layers of the ascending and transverse colons

of a subserosal lesion (Fig. 3). Subsequent colonoscopy suggested mild colitis and scarring but showed no polypoid lesion. Empirical treatment with intravenous cefotiam hydrochloride and adequate hydration were initiated with bowel rest. The patient's symptoms gradually improved with conservative management, and on the 12th day after admission she resumed oral intake. She was discharged 16 days after admission when the resolution of PCI was confirmed on radiographs (Fig. 4A). Six months after discharge, a relapse of PCI was incidentally found on follow-up abdominal radiographs (Fig. 4B). She has been completely asymptomatic with regular diet and did not need any specific treatment during the 3 months of follow-up after relapse of PCI.

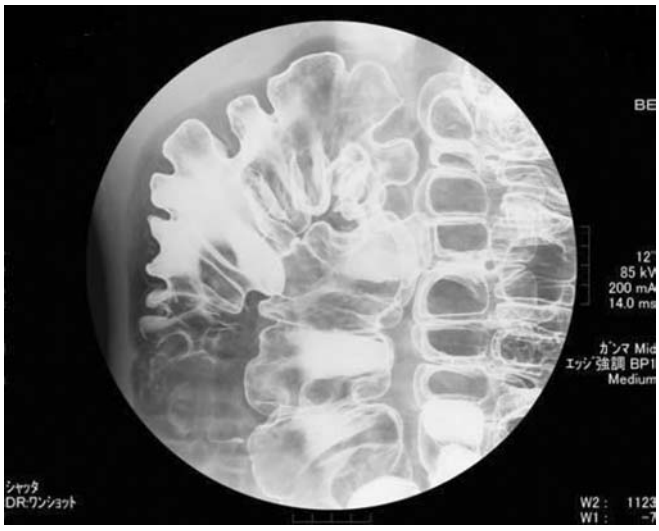

Fig. 3. Barium enema showed an extensive gas layer surrounding the ascending colon, suggestive of subserosal lesion

Discussion

Pneumatosis cystoides intestinalis is a relatively uncommon condition characterized by the presence of multiple gas-filled mucosal, submucosal, or subserosal cysts located throughout the colon and/or small intestine.[1,2] In general, there are several speculative theories regarding the etiology of PCI, including mechanical and bacterial hypotheses.[10] Mechanical factors, such as a break in the mucosal integrity with increased intraluminal pressure, may cause intestinal gas to dissect into the bowel wall. This situation may occur in intestinal obstruction, colonoscopy, and bowel surgery. An intermittent air leak through a peptic or malignant ulcer crater could cause PCI in some patients.[11] In addition, chronic obstructive pulmonary disease could lead air to dissect along bronchi into the mediastinum to the retroperitoneum.[12] Retroperitoneal air may subsequently dissect along a vascular channel into the bowel wall. It is suggested that alveolar rupture with gas dissection might be pathogenic in PCI. A bacterial mechanism[13] may involve the invasion of gas-producing bacteria from the intestinal lumen.

Symptoms of PCI include anorexia, nausea, vomiting, diarrhea, and abdominal pain, but PCI may also be totally asymptomatic. The diagnosis is usually made by abdominal radiographs or contrast studies.[4,14] Abdominal radiographs show bowel dilation and mottled, bubbly, and linear collections of intramural intestinal gas.[15] Contrast studies show defects in the intestinal wall which suggest submucosal

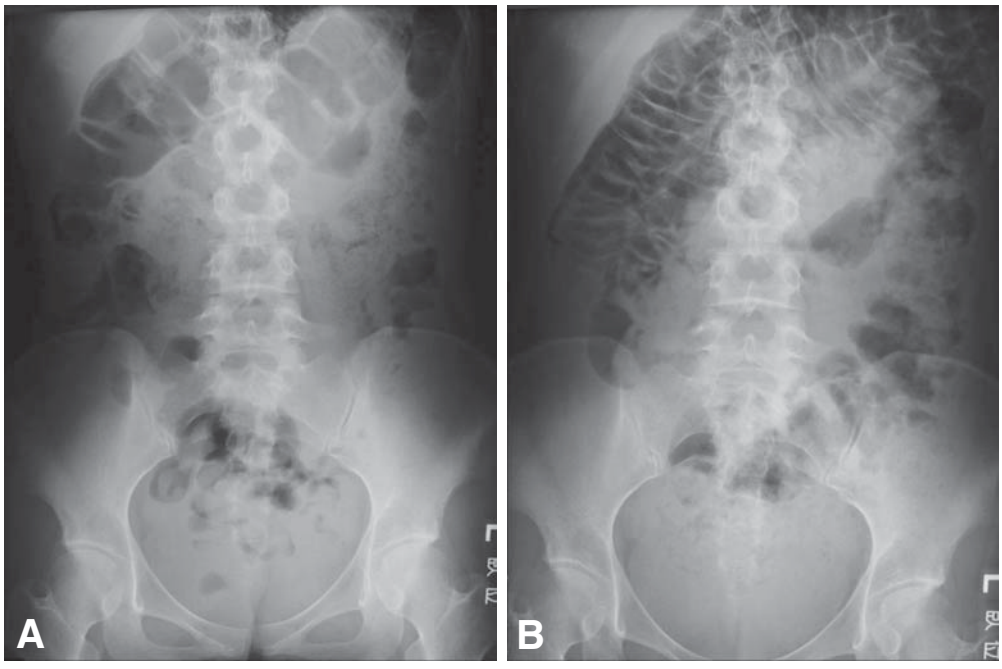

Fig. 4. **A** At discharge. The resolution of pneumatosis cystoides intestinalis (PCI) was confirmed on radiographs. **B** Six months after discharge, a relapse of PCI was incidentally found on follow-up abdominal radiographs

lesions. Endoscopic studies typically show multiple polypoid lesions, which consist of air-filled cysts covered by intestinal mucosa. The mucosal surface covering the cysts may be focally eroded.[14]

In general, PCI follows a benign course and treatment is usually unnecessary.[1,14] In some cases, resolutions of the cysts may be brought about by inhaling highly concentrated oxygen for a few days.[16,17] In rare cases with difficulty in intestinal passage caused by gaseous cyst, or PCI caused by intestinal passage disorder, in addition to cases indicative of intestinal necrosis, surgical management may be required.[14]

PCI comprises one of the gastroenteropathies seen in connective tissue diseases, particularly in SSc.[8,18–21] Atrophy and fibrosis in the inner circular muscle layer, the findings consistent with so-called scleroderma bowel,[20,21] may play a key role in the invasion of air into the bowel wall in SSc patients. Another possible mechanism of PCI in SSc includes the changes in bowel bacterial flora by persistent hypokinesis of the gut.

Pneumatosis cystoides intestinalis is a rare complication in SLE. To the best of our knowledge, a total of 13 cases of PCI complicating SLE have been reported in the literature (Table 1). Although the precise pathogenesis is unknown in most patients, one or more pathogenic mechanisms may be present. Some case reports suggest the exclusive role of vasculitis in the development of PCI in SLE. According to this hypothesis, some patients are reported to have been successfully treated with steroids and/or cyclophosphamide. Cabrera et al.[4] described PCI in a patient with active SLE and intestinal vasculitis, who underwent intestinal resection for acute abdomen and had an excellent response to high-dose PSL. The high-dose PSL improved active SLE as well as abdominal symptoms that were caused by vasculitis in the abdominal area. This speculation of the role of vasculitis is possible, but the fact that PCI seldom complicates other vasculitis syndromes that often accompany gastrointestinal symptoms suggests other mechanisms. Another controversial theory is the role of steroids in the pathogenesis of PCI.[16,26] The hypothesis is based on the speculation that steroids initiate shrinkage of lymphoid tissues in the Peyer patches of the intestine, and in some manner impair the structural integrity of the bowel and permit dissection of intramural gas.[27] Cases of PCI are reported in other diseases treated with steroids; nephrotic syndrome, leukemia, and transplant patients.[27,28]

The clinical characteristics of case reports of PCI in SLE are shown in Table 1. Generally, PCI in SLE, as well as in other conditions, is considered as a benign disorder, especially in adults. In fact, some cases resolved without any specific treatment. Pruitt et al.[1] reported a case of PCI with SLE who was treated with conservative management and had no recurrence. Atsumi et al.[25] reported a patient with PCI who underwent operation that was considered unnecessary. However, there seems to be a distinct disease population of PCI in SLE associated with vasculitis, requiring aggressive therapy including steroids and/or immunosuppressive agents, with a poor prognosis. Decrop et al.[5] reported a patient who underwent urgent laparotomy whose histologic examination showed evidence of vasculitis with necrosis of the mucosal layer. He concluded that the management of the acute abdomen in SLE is a challenging therapeutic and surgical issue; an acute intestinal infarction and perforation can be ruled out by upright abdominal radiograph and paracentesis, and a therapeutic trial of

Table 1. Systemic lupus erythematosus (SLE) with pneumatosis cystoides intestinalis

First author (year)[Ref.]	Age (years)/sex	Abdominal symptoms	Clinical features	Free air	Steroid	Pathogenesis	Treatment	Prognosis
Freiman (1975)[6]	54/F	(–)	Active SLE	(–)	(–)	Unknown	(–)	Death
Morrison (1976)[7]	29/F	(–)	Active SLE	(–)	(–)	Mesenteric arteritis	(–)	Death
Kleinman (1976)[9]	35/F	Abdominal pain, acute abdomen, intestinal vasculitis	Active SLE	(+)	(+)	Intestinal vasculitis	Surgery	Death
Binstadt (1977)[22]	15/F	Abdominal pain, vomiting, diarrhea	Active SLE	(+)	(+)	Unknown	(–)	Death
Derksen (1978)[23]	28/F	Abdominal pain	Active SLE	(–)	(–)	Generalized mesenteric arteritis	O_2 therapy	Remission
Pruitt (1988)[1]	58/F	Abdominal pain, nausea, vomiting, bloody stool	Inactive SLE	(+)	(+)	Unknown	(–)	Remission; completely asymptomatic without recurrence during 24 months
Laing (1988)[24]	21/F	Abdominal pain, nausea, vomiting, rectal bleeding	Active SLE	(–)	(+)	Small vessel arteritis	Cyclophosphamide	Remission
Decrop (1990)[5]	30/M	Abdominal pain, ileus, ischemic bowel disease	Shock, active SLE	(–)	(+)	Small bowel vasculitis	Surgery	Death
Atsumi (1991)[25]	51/F	(–)	Inactive SLE	(+)	(+)	Unknown	Surgery	Remission; renal failure got worse after surgery and dialysis was introduced
Cabrera (1994)[4]	31/F	Abdominal pain, tenderness, muscular defense, diarrhea	Active SLE	(–)	(–)	Vasculitis of the arterioles and venules of the bowel and the mesentery	Laparotomy, steroid pulse	Remission; dramatic improvement:receiving predonisolone 20 mg/day and hydroxychloroquine
Nonaka (1998)[16]	51/F	(–)	Inactive SLE	(+)	(+)	Unknown	O_2 therapy	Remission; no recurrence
Hiraishi (1999)[26]	13/F	(–)	Inactive SLE	(+)	(+)	Unknown	(–)	Remission; no recurrence
Alcocer-G. (2000)[15]	14/F	Abdominal pain, tenderness, muscular defense, vomiting	Inactive SLE	(+)	(+)	Bowel vasculitis	Laparotomy, cyclophosphamide	Remission
Yamaguchi (present case)	33/F	Abdominal pain, tenderness, No rebound tenderness	Inactive SLE	(–)	(+)	Unknown	(–)	Temporal remission, reccurence (+) after 6 months, completely asymptomatic

high-dose steroids and broad-spectrum antibiotics should be started. If the patient's condition deteriorates, surgical intervention should be promptly considered. To improve the prognosis of these severe cases, and to avoid unnecessary laparotomy in general benign cases, patients need careful evaluation and follow-up.

In our patient, clinical and serological findings suggest that PCI was not due to vasculitis. Her PCI improved with conservative management, and the relapse of PCI was shown on follow-up abdominal radiographs, but she has been completely asymptomatic with no specific treatment. Long-term use of steroids might have a role in the pathogenesis of PCI in our case.

Although PCI is a rare manifestation of SLE, clinicians should be alert to the differential diagnosis of this complication. As the disease course of PCI in SLE is diverse, all cases should be followed by a multidisciplinary team in close communication, including primary care physicians, rheumatologists, gastroenterologists, and surgeons.

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