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CASE REPORT

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A case of pneumatosis cystoides intestinalis in a patient with polymyositis and interstitial pneumonia

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Abstract We report the case of a 49-year-old man who presented with pneumatosis cystoides intestinalis associated with polymyositis and interstitial pneumonia. Three months after the administration of prednisolone and cyclosporine, he noticed mild abdominal distension, and a radiographic examination showed intraperitoneal free gas and intramural gas, suggestive of pneumatosis cystoides intestinalis (PCI). Additional treatment with a combination of doxycycline and high-dose oxygen therapy was effective in relieving the clinical symptoms and reducing the intramural gas.

Key words Intersitial pneumonia · Pneumatosis cystoides intestinalis (PCI) · Polymyositis

Introduction

Pneumatosis cystoides intestinalis (PCI) is a condition characterized by the presence of intramural gas cysts in the gastrointestinal wall.[1–3] This rare complication is known to be associated with various medical conditions, including autoimmune disorders such as systemic sclerosis, systemic lupus erythematosus, mixed connective tissue disease, overlap syndrome, polyarteritis nodosa, and dermatomyositis.[4–15] In pure polymyositis (PM), however, gastrointestinal involvement is rarely observed except for pharyngeal dysphagia, and only a few case reports of PCI associated with PM have been documented.[16–19] In this report, we describe an interesting case of PCI associated with PM and interstitial pneumonia.

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Case report

A 49-year-old man complaining of muscle weakness and dry cough was admitted to our hospital on March 28, 2001. The patient had no family history of muscle disease or autoimmune disease. He had been well until December 2000, when symptoms of general fatigue and muscle weakness appeared. On physical examination, he had a temperature of 36.8°C, blood pressure of 120/68 mmHg, and a regular pulse of 64/min. He had no history of Raynaud's phenomenon or gastrointestinal symptoms. There was no malar rash or heliotrope rash on his face. A slight fine crackle was audible at the bilateral lower back, but no abnormalities were found in the abdomen. Gottron's sign, sclerodactyly, and telangiectasia were not recognized on his skin. Neurological examination revealed moderate bilateral muscle weakness of the proximal limbs. Reflexes and sensations appeared to be normal.

Laboratory tests showed a normal blood cell count, and serum electrolytes and renal function were within the normal ranges. However, transaminase levels were slightly elevated and the level of creatine phosphokinase (CPK) was markedly high at 1780 IU/l. Serological testing showed the patient to be negative for antibodies against nuclear factor, dsDNA, RNP, Scl-70, and Jo-1. His serum level of KL-6 was elevated at 1080 U/ml (normal <500 U/ml). Blood gas analysis gave the following results: pH 7.431; $PaCO_2$ 46.7 mmHg; PaO_2 79.5 mmHg; HCO_3- 30.5 mmol/l; BE 5.1 mmol/l; SaO_2 96.6%. A chest computed tomography (CT) scan showed reticulo-nodular shadows at the posterior side of the bilateral lower lobes. An abdominal CT scan, gastroendoscopy, and colonoscopy showed normal findings. Cardiac ultrasonography showed normal cardiac function

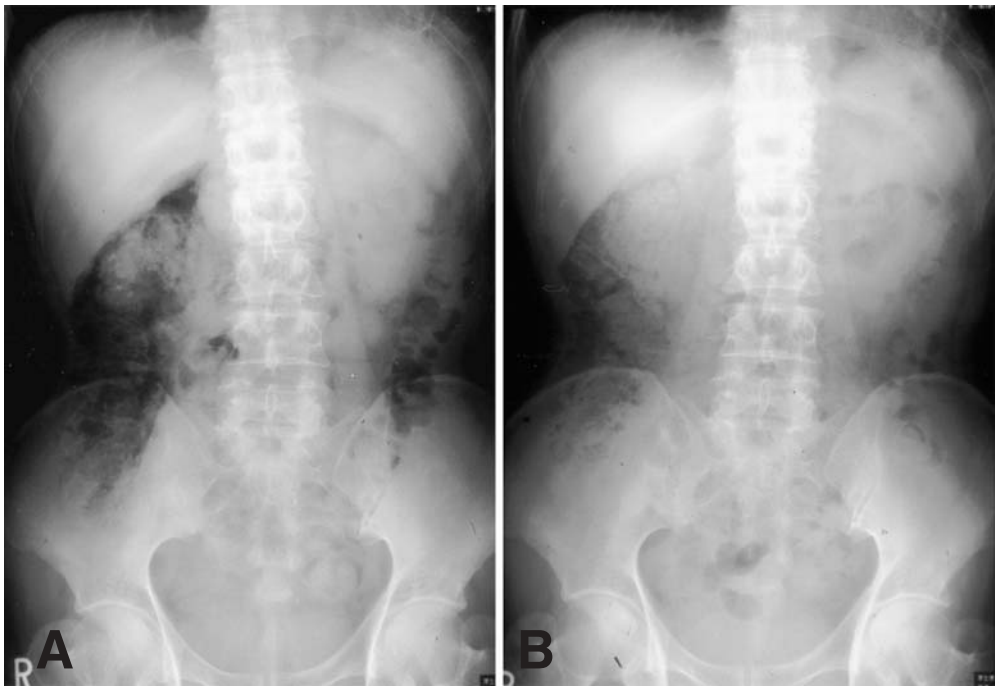

Fig. 1. **A** Abdominal radiography on July 5, 2001, showing intraperitoneal free gas and intramural gas around the liver. **B** Abdominal radiography 2 weeks after treatment showing the slight reduction of intramural gas compared with **A**

without pulmonary hypertension. Electromyography showed low-amplitude and short-duration patterns, and a muscle biopsy revealed thin and necrotic fibers with infiltration of mononuclear cells around the small vessels, which are the characteristic features of PM. A lung biopsy was also performed using video-associated thoracoscopy (VATS), and the specimen revealed nonspecific interstitial pneumonia (type 2). Based on these findings, the patient was diagnosed as having pure PM associated with interstitial pneumonia. Corticosteroid (prednisolone 1 mg/kg) and cyclosporine (150 mg/day) were started on April 17, and the dosage of prednisolone was tapered by 5 mg every 4 weeks. The clinical symptoms, including the muscle weakness and dry cough, gradually improved, together with normalization of the serum CPK level.

In late June, the patient began to notice mild abdominal distension. An abdominal radiographic examination was performed on July 5, and this showed intraperitoneal free gas and intramural gas around the liver (Fig. 1A). Physical and laboratory examinations showed no evidence of bowel perforation. An abdominal CT scan was performed immediately, and intramural gas was found in the small bowel (Fig. 2). Colonoscopy revealed a submucosal cyst in the terminal ileum, which had not been detected at the time of admission. A diagnosis of PCI was made, and doxycycline (100 mg per day) together with high-dose oxygen therapy (10 l/min via a face mask for 4 h per day) was started. Arterial blood gas analysis showed pH 7.443, $PaCO_2$ 45.1 mmHg, and PaO_2 210.8 mmHg while using the facemask. The patient's abdominal distension improved, and follow-up abdominal radiography showed a slight reduction of intramural gas (Fig. 1B). During a 24-month follow-up period, he has been treated with 10 mg prednisolone and 100 mg cyclosporine daily, and no gastrointestinal symptoms have been observed.

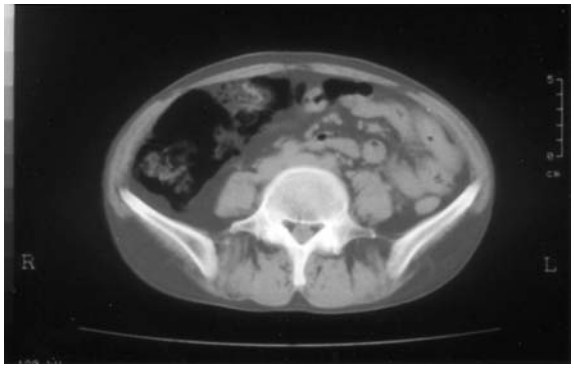

Fig. 2. Abdominal CT scan on July 5, 2001, showing intramural gas in the small bowel

Discussion

PCI is characterized by the presence of intramural gas cysts in the gastrointestinal wall. This rare complication is known to occur in patients with various medical conditions such as gastrointestinal disease, chronic obstructive lung disease, sepsis, organ transplantation, graft versus host disease, HIV infection, and autoimmune disorders.[1–3]

Recent reviews of PCI have proposed several pathological mechanisms.[1–3] Bowel necrosis can cause gaseous cysts in the bowel, which arise from bowel ischemia or infarction,

Table 1. Case reports of pneumatosis cystoides intestinalis associated with polymyositis, dermatomyositis, and overlap syndrome

Case	Age/sex	Disease	Duration	Activity	Clinical symptoms	IP Therapy	Prognosis
1	46/F	PM	3 months	Remission	Vomiting, diarrhea	(+) (−)	Good
2	76/F	PM	10 months	Remission	Abdominal distension, vomiting	(−) High-dose oxygen	Good
3	29/F	PM	10 months	Remission	Abdominal distension	(−) High-dose oxygen, doxycycline	Good
4	38/M	PM	4 years	Remission	Colicky abdominal pain, body weight loss	(−) High dose IVIG, metronidazole, total parenteral nutrition, cisapride octreotide	Good
5	49/M	PM	3 months	Remission	Abdominal distension	(+) High-dose oxygen, doxycycline	Good
6	4/F	DM	1 year	Remission	Abdominal pain, diarrhea, constipation	(−) (−)	Good
7	12/F	DM	3 years	High	Abdominal pain	(−) PSL20 mg/day, MTX	Good
8	8/M	DM	13 months	High	Abdominal pain, diarrhea, vomiting	(−) m-PSL, MTX	Death
9	12/F	DM	3 years	High	Dysphagia	(−) Steroid	Good
10	7/F	DM	3 months	High	Abdominal pain, appetite loss	(−) Total parenteral nutrition	Good
11	80/F	DM	16 years	Remission	Abdominal pain	(+) Metronidazole	Good
12	61/F	DM	18 months	Remission	Abdominal pain, vomiting body weight loss	(−) Total parenteral nutrition, laparotomy	Good
13	60/F	OS	21 months	Remission	Abdominal distension, nausea, vomiting	(−) Total parenteral nutrition High-dose oxygen	Death
14	58/F	OS	3 years	Remission	Abdominal distension	(−) Laparotomy, oxygen Total parenteral nutrition	Death

PM, polymyositis; DM, dermatomyositis; OS, overlap syndrome; duration, duration after onset of disease; activity, disease activity; IP, interstitial pneumonia; IVIG, intravenous immunoglobulin administration; PSL, prednisolone; m-PSL, methylprednisolone; MTX, methotrexate

necrotizing enterocolitis, and sepsis. Bacterial infections, which produce lesions, may damage the bowel wall and produce gas, which is thought to penetrate into the gastrointestinal wall. PCI is also found in patients with mucosal disruption due to ulceration, erosion, and trauma. Another mechanism of mucosal disruption can occur in patients with pulmonary diseases due to gas leakage from the lung interstitium. In addition, patients receiving immunosuppressive therapy have a higher risk of developing PCI due to increased bowel wall permeability.

PCI is known to be one of the rare complications associated with rheumatic disorders, especially systemic sclerosis (SSc), and has also been reported in patients with systemic lupus erythematosus (SLE), mixed connective tissue disease (MCTD), overlap syndrome, polyarteritis nodosa, and dermatomyositis (DM).[4–15] Although esophageal impairment has been reported to occur in as many as 25%–60% of patients with PM/DM, other gastrointestinal complications, including PCI, intestinal pseudo-obstruction, colonic perforation, or hemorrhage due to vasculitis can occur, but are less common.[20]

There have been five previous case reports of PCI associated with PM (Table 1).[16–19] In these cases, the disease activity of PM was mostly controlled by the administration of corticosteroid at the time of PCI onset. Interstitial pneumonia was found in two cases, including the present one, and the disease activity was also controlled. Although cases 1, 2, and 4 were associated with gastrointestinal dysmotility or pseudo-obstruction, these clinical symptoms improved without any need for surgical treatment and the prognosis was reportedly favorable in all patients.

PCI is reported to be a rare gastrointestinal manifestation in patients with DM, especially children (Table 1).[4–10] Juvenile DM (JDMS) is characterized by systemic vasculopathy involving mainly muscle and skin.[21] Among five reported cases of PCI with JDMS, four patients had high disease activity at PCI onset, and one patient died of bowel necrosis and perforation, suggesting the occurrence of digestive vasculitis.[4–8] However, the clinical courses of two PCI patients with adult DM were almost the same as those of patients with PM.[9,10]

The clinical manifestations of PCI with PM/DM are somewhat different from those of SSc and overlap syndrome (Table 1), which are usually observed in patients with end-stage disease with a poor prognosis.[13,14,22,23] It has been suggested that tissue ischemia and hypoperfusion based on inappropriate vasoconstriction and abnormal autonomic responses lead not only to progressive gastrointestinal wall damage, but also to PCI onset in patients with SSc and overlap syndrome. Similarly, the association with subclinical vasculopathy in the bowel mucosa of JDMS patients may be related to the occurrence of PCI. Although the etiology of PCI is still unclear in PM and adult DM, the infrequent and mild gastrointestinal wall damage is likely to have a more favorable prognosis than that of SSc and JDMS.

Treatment of PCI is only recommended for symptomatic patients.[1–3] Conservative therapy consists of nasogastric suction with a balanced diet, administration of antibiotics to control abnormal bacterial overgrowth in the gastrointestinal tract, and high-flow oxygen therapy to help the absorption of cyst gas by capillary blood.[24] Surgery is required only in severe cases where there is bowel necrosis and perforation. In our patient, the administration of doxycyclin and high-flow oxygen therapy were selected to control his abdominal distension. Although the patient had interstitial pneumonia associated with PM, no deleterious effect of high-flow oxygen therapy was observed in his lungs. This

combined therapy improved his clinical symptoms without any side-effects during the follow-up period.

In conclusion, we have reported a case of PCI in a patient with PM and interstitial pneumonia. The patient responded well to conservative therapy and the prognosis seems to be favorable. Further similar cases will be needed in order to clarify the etiology and characteristics of PCI associated with PM.

References

1. Heng Y, Schuffler MD, Haggitt RC, Rohrmann CA. Pneumatosis intestinalis: a review. Am Coll Gastroenterol 1995;90:1747–58.
2. Pear BL. Pneumatosis intestinalis: a review. Radiology 1998;207: 13–9.
3. Voboril R. Pneumatosis cystoides intestinalis: a review. Acta Medica (Hradec Kralove) 2001;44:89–92.
4. Mueller CF, Morehead R, Alter AJ, Michener W. Pneumatosis intestinalis in collagen disorders. Am J Roentgenol 1972;115:300–15.
5. Oliveros MA, Herbst JJ, Lester PD, Ziter FA. Pneumatosis intestinalis in childhood dermatomyositis. Pediatrics 1973;52:711–2.
6. Fischer TJ, Cipel L, Stiehm ER. Pneumatosis intestinalis associated with fatal childhood dermatomyositis. Pediatrics 1978;61:127–30.
7. Braunstein EM, White SJ. Pneumatosis intestinalis in dermatomyositis. Br J Radiol 1980;53:1011–2.
8. Stefanski JC, Shetty AK. Abdominal pain in a girl with juvenile dermatomyositis. Clin Pediatr 1988;37:561–3.
9. Pasquier E, Wattiaux MJ, Peigney N. First case of pneumatosis cystoides intestinalis in adult dermatomyositis. J Rheumatol 1993;20:499–503.
10. Morris-Stiff GJ, Williams RJLL. Pneumatosis cystoides intestinalis in a patient with dermatomyositis. J R Soc Med 1999;92:366–7.
11. Carbera GE, Sopelitis E, Cuellar ML, Silveira LH, Mena H, Espinoza LR. Pneumatosis cystoides intestinalis in systemic lupus erytematosus with intestinal vasculitis: treatment with high-dose prednisolone. Clin Rheumatol 1994;13:312–6.
12. Samach M, Brandt LJ, Bernstein LH. Spontaneous pneumoperitoneum with pneumatosis cystoides intestinalis in a patient with mixed connective tissue disease. Am J Gastroenterol 1978;69:494–500.
13. Shirako J, Yoshikawa T, Ohinishi T, Nakai M, Asano H, Kawakami T, et al. Paralytic ileus, pneumatosis cystoides intestinalis and pneumoperitoneum complicated with overlap syndrome (progressive systemic sclerosis and polymyositis): report of an autopsy case. Nippon Shokakibyo Gakkai Zasshi 2000;97:1031–7.
14. Karube M, Kaneda F, Nakabayashi K, Yamada A, Nagasawa T. Pneumatosis cystoides intestinalis in overlap syndrome manifested by dermatomyositis and scleroderma. Nippon Naika Gakkai Zasshi 2002;91:3278–81.
15. Buffo G, Deitch J. Pneumatosis intestinalis in a patient with polyarteritis nodosa. Gastrointest Radiol 1986;11:286–8.
16. Hanawa M, Toda H, Kobayashi H, Fujisawa H, Matsuoka M. Interstitial pneumonia, intestinal pseudo-obstruction and pneumatosis cystoides intestinalis associated with polymyositis: report of a case. Stomach Intestine 1982;17:1021–7.
17. Park YH, Knoh T, Nishida O, Yoshida Y, Ohkuma M, Uchino H, et al. A case of polymyositis with ileus-like symptoms and pneumatosis cystoides intestinalis, improved by the treatment of high-flow oxygen therapy. Nippon Naika Gakkai Zassi 1985;74:138–42.
18. Kuroda T, Ohfuchi Y, Hirose S, Nakano M, Gejyo F, Arakawa M. Pneumatosis cystoides intestinalis in a patient with polymyositis. Clin Rheumatol 2001;20:49–52.
19. Elkayam O, Gaspi D, Flusser G. Pneumatosis intestinalis in a patient with polymyositis. Clin Exp Rheumatol 2001;19:483.
20. Marie I, Hachulla E, Lecesque H, Reumont G, Ducrotte P, Cailleux N, et al. Intravenous immunoglobulins as treatment of life-threatening esophageal involvement in polymyositis and dermatomyositis. J Rheumatol 1999;26:2706–9.
21. Pachman LM. An update on juvenile dermatomyositis. Curr Opin Rheumatol 1995;7:437–41.
22. Sjogren RW. Gastrointestinal features of scleroderma. Curr Opin Rheumatol 1996;8:569–75.
23. Lock G, Holstege A, Lang B, Scholmerich J. Gastrointestinal manifestations of progressive systemic sclerosis. Am J Gastroenterol 1997;92:763–71.
24. Holt S, Gilmour HM, Buist TAS, Marwick K, Heading RC. High-flow oxygen therapy for pneumatosis coli. Gut 1979;20:493–8.