## Marked Pneumatosis Cystoides Intestinalis in a Patient with Mixed Connective Tissue Disease

YOKO AOKI, MD; TAKAO NAGASHIMA, MD; TAKESHI KAMIMURA, MD; MASAHIRO IWAMOTO, MD; SEIJI MINOTA, MD, Division of Rheumatology and Clinical Immunology, Department of Medicine, Jichi Medical University, Tochigi, Japan. Address reprint requests to Dr. T. Nagashima, Division of Rheumatology and Clinical Immunology, Department of Medicine, Jichi Medical University, Minamikawachi 3311-1, Tochigi, Japan. E-mail: naga4ma@jichi.ac.jp

A 60-year-old woman with mixed connective tissue disease (MCTD) was referred to our rheumatology and clinical immunology unit for the treatment of pneumatosis cystoides intestinalis (PCI). She was diagnosed with MCTD 15 years ago, based upon her Raynaud's phenomenon, digital swelling, biopsy-proven myositis, positive anti-U1-RNP antibody, and interstitial pneumonia. She was prescribed glucocorticoid for the treatment of her interstitial pneumonia since then. Although she had a 6-year history of repeated colonic obstruction, the diagnosis of PCI was made for the first time 6 months ago and she was treated conservatively by oxygen therapy. She lost 10 kg of weight within a year. Physical examination revealed mild abdominal distention without tenderness. Her bowel sounds increased, and she had intermittent diarrhea

with steatorrhea. Her abdominal radiograph showed small free air under the right hemidiaphragm and distended small intestine with marked cystic lucencies in the wall (Figure 1). Her abdominal cross-sectional computerized tomography (CT) scan revealed extensive multiple cystic air densities in parallel with the bowel wall (Figure 2). Treatment with high flow oxygen (8 l/min via face mask with reservoir pouch, 4 h a day for 2 weeks) with intravenous hyperalimentation led to complete symptomatic and radiological resolution.

As reviewed elsewhere, among collagen vascular diseases, systemic sclerosis (SSc) is most often complicated with PCI, followed by systemic lupus erythematosus and, rarely, by MCTD<sup>1-3</sup>. Although PCI associated with SSc appears to be a late complication and is considered a poor prognostic sign, it



Figure 1. Abdominal radiograph in the standing position shows small free air under the right hemidiaphragm (arrowheads), and distended small intestine with cystic and curvilinear lucencies.

Personal non-commercial use only. The Journal of Rheumatology Copyright © 2006. All rights reserved.

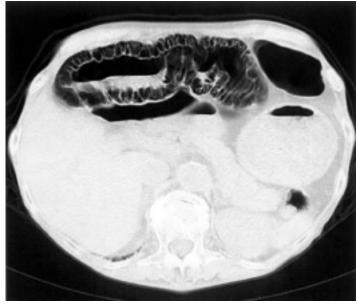


Figure 2. Cross-sectional CT scan shows typical "ladder-like appearance." Intramural cysts have no air-fluid levels, which could be distinguished from intraluminal air. Lung window settings were quite helpful in detecting the intramural cystic gas.

occurs relatively early in the disease course in MCTD<sup>2,4,5</sup>. Typical PCI images with "ladder-like appearance" by CT scan shown in this report have not been previously published for collagen vascular diseases.

## REFERENCES

- Lynn JT, Gossen G, Miller A, Russell IJ. Pneumatosis intestinalis in mixed connective tissue disease: two case reports and literature review. Arthritis Rheum 1984;27:1186-9.
- Sequeira W. Pneumatosis cystoides intestinalis in systemic sclerosis and other diseases. Semin Arthritis Rheum 1990;19:269-77.
- Wakamatsu M, Inada K, Tsutsumi Y. Mixed connective tissue disease complicated by pneumatosis cystoides intestinalis and malabsorption syndrome: case report and literature review. Pathol Int 1995;45:875-8.
- Goulet JR, Hurtubise M, Senecal JL. Retropneumoperitoneum and pneumatosis intestinalis in 2 patients with mixed connective tissue disease and the overlap syndrome. Clin Exp Rheumatol 1988;6:81-5.
- Pun YL, Russell DM, Taggart GJ, Barraclough DR. Pneumatosis intestinalis and pneumoperitoneum complicating mixed connective tissue disease. Br J Rheumatol 1991;30:146-9.