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

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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[1-3]. Although PCI associated with SSc appears to be a late complication and is considered a poor prognostic sign, it

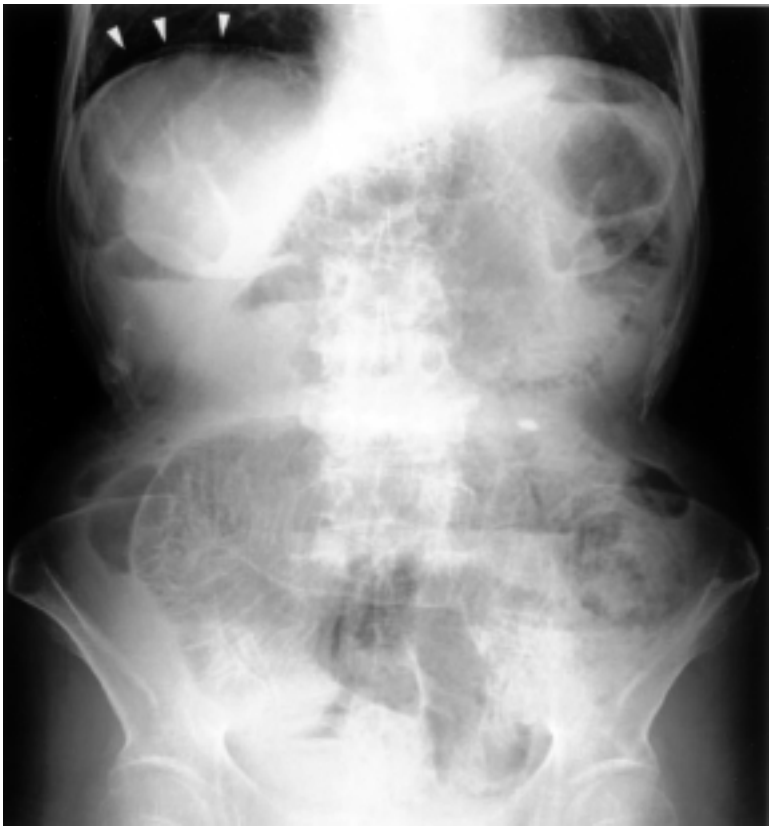

Figure 1. Abdominal radiograph in the standing position shows small free air under the right hemidiaphragm (arrowheads), and distended small intestine with cystic and curvilinear lucencies.

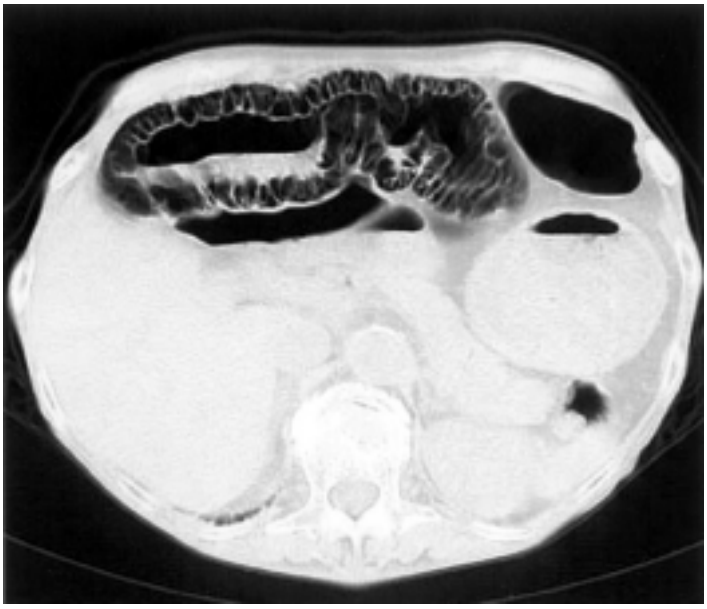

Figure 2. Cross-sectional CT scan shows typical "ladder-like appearance." Intramural cysts have no air-fluid levels, which could be distinguished from intraluminal air. Lung window settings were quite helpful in detecting the intramural cystic gas.

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

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