Simultaneous Pneumatosis Cystoides Intestinalis and Pneumomediastinum in a Patient with Systemic Sclerosis

To the Editor:

Interstitial lung disease (ILD) is a common pulmonary manifestation in patients with systemic sclerosis (SSc). Pneumomediastinum occasionally develops in patients with connective tissue diseases (CTD) and ILD, but is rare in SSc. Although gastrointestinal (GI) involvement is common in patients with SSc, pneumatoses cystoides intestinalis (PCI), which is characterized by the presence of gas-filled cysts in the intestinal wall, is a rare complication. We encountered a patient with SSc who showed simultaneous occurrence of pneumomediastinum and PCI.

A 61-year-old Japanese woman had been diagnosed with SSc 6 years earlier based on the findings of proximal scleroderma, positive antinuclear antibody (1:640 with nucleolar and homogeneous patterns), positive anti-Scl-70 antibody (1:16), and ILD. Fingertip ulceration had developed 4 years before. Prednisolone (20 mg/day) had been started 2 years before for her skin sclerosis. She had gradually lost her appetite and her weight had declined by 15 kg over the previous 6 months. She was admitted to our division because of poor appetite and abdominal distension. On admission, she was being treated with 10 mg/day prednisolone. Physical examination revealed that her abdomen was distended and tympanic. Bilateral fine crackles were heard in the lung bases, but she had no respiratory symptoms. Plain abdominal radiographs showed marked dilation of the small bowel with prominent Kerckring’s folds and linear lucencies along the bowel wall. Computed tomography of the thorax and abdomen revealed multiple linear and cystic accumulations of gas in the walls of the small bowel, as well as ascites (Figure 1). There was also a small amount of free air around the dilated stomach and under the diaphragm, which extended into the retroperitoneal space. Further, there was free air around the trachea and in the subcutaneous tissues of the chest wall (Figure 2). These findings confirmed the simultaneous presence of PCI and pneumomediastinum. Her radiological abnormalities improved after a few weeks of conservative therapy, consisting of bowel rest and high-flow oxygen (5 l/min) by a face mask.

Pneumomediastinum is a rare complication of GI endoscopy, and it usually indicates perforation leading into the peritoneal cavity or the retroperitoneal space that results from trauma to the GI mucosa. Air enters the bowel wall by the mucosal wound and ascends through the retroperitoneum, tracking upward to reach the mediastinum. This can occur without any overt evidence of perforation. This ascending route from the intestine is more reasonable in our patient, since her chief complaint was abdominal distension, and abdominal radiographs showed marked small bowel dilation (similar to that after endoscopic insufflation).

Both PCI and pneumomediastinum are rare in patients with SSc. It is highly unlikely that 2 rare conditions would occur simultaneously. Rather,
it appears that retroperitoneal and peritoneal free air derived from PCI escaped upward along the para-aortic tissues and through the esophageal hiatus into the mediastinum, finally leading to pneumomediastinum and the subcutaneous emphysema in our patient. Unlike the previous cases in patients with SSc, this is not “spontaneous” pneumomediastinum. A similar case was reported in a patient with DM with simultaneous pneumomediastinum and PCI. Corticosteroid therapy and/or rapid weight loss might be associated with loosening and weakening of the mediastinal connective tissues in such patients.

KYOKO HONNE, MD; AKIHITO MARUYAMA, MD; SACHIKO ONISHI, MD; TAKAO NAGASHIMA, MD, PHD; SEIJI MINOTA, MD, PHD, Professor of Medicine, Division of Rheumatology and Clinical Immunology, Department of Medicine, Jichi Medical University, Yakushiji 3311-1, Shimotsuke, Tochigi, 329-0498, Japan. Address correspondence to Dr. Nagashima; E-mail: naga4ma@jichi.ac.jp

REFERENCES

J Rheumatol 2010;37:10; doi:10.3899/jrheum.100254

Figure 2. Computed tomography scan of the chest. There is free air around the trachea, which extends into the subcutaneous tissues of the anterior chest wall. The esophagus is also dilated.