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Clinical Image

Pneumatosis intestinalis in a patient with anti-synthetase syndrome

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We report a rare case of anti-synthetase syndrome (ASS) complicated by pneumatosis intestinalis (PI).

A 71-year-old Japanese woman presented with anorexia and abdominal fullness. Eleven years prior, she had been diagnosed with ASS due to myositis, interstitial lung disease (ILD), Raynaud phenomenon, arthritis and anti-EJ antibody positivity. She was successfully treated with prednisolone (PSL) and tacrolimus, but subsequently experienced multiple ILD flares. Her condition was successfully controlled with PSL (7 mg/d), mycophenolate mofetil (1000 mg/d), abatacept (125 mg/week s.c.) and nintedanib (300 mg/d) in the 6 months before presentation. Two weeks before presentation, she was anorexic and felt worsening abdominal fullness and diarrhea. On her visit, she was sick, and the abdomen was mildly distended but soft and non-tender. CT revealed intramural air in the colon and small intestine (Fig. 1). Air was also detected around the aorta (blue arrows) and liver (blue arrowhead). She was diagnosed with PI, hospitalized and treated nonsurgically with fasting and intravenous supplemental fluids, followed by a digestible diet, antibiotics (ampicillin) and oxygen therapy. During her 2-week hospitalization, her condition improved, she was able to eat and the intramural air in the intestine disappeared.

PI is characterized by intramural gas in the intestine and is associated with many underlying diseases, including trauma, obstruction, infection, and pulmonary and autoimmune diseases [1, 2]. In autoimmune diseases, scleroderma is common; however, cases of myositis have been reported [3], but rarely of ASS [4]. Intramural gas purportedly originates from the intestinal tract and lung [1, 5]. Alveolar rupture may cause air migration along the mediastinal vessels and through the retroperitoneum to the mesentery of the bowel, resulting in PI [1, 5]. In the present case, air in the mediastinum (blue arrows) and retroperitoneum (blue arrowhead) was more apparent in the former than in the latter, supporting this mechanism [1, 5]. We assumed that the fragility of the intestinal mucosa due to inflammation and glucocorticoids, abnormal bowel movement enhanced by nintedanib and ILD contributed to the development of PI in the present case.

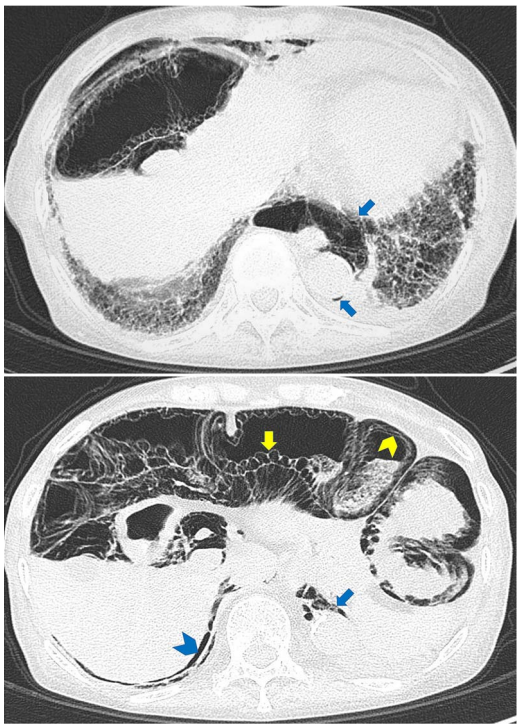

Figure 1. CT reveals reticular opacities in the lung and a low-density bubbly (yellow arrow) or linear pattern (yellow arrowhead) of gas in the bowel wall, indicating ILD and PI, respectively. Gas was detected in the mediastinum and retroperitoneum (blue arrows), as well as in the peritoneum (blue arrowhead).

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PERMISSION FROM THE PATIENT OF THIS MANUSCRIPT

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DATA AVAILABILITY

The data are available from the corresponding author upon reasonable request.

ETHICAL APPROVAL

Not applicable.

CONSENT

The patient provided written permission for the publication of her case.

GUARANTOR

Kazuhiro Kurasawa is the guarantor for this manuscript.

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