

Immunoglobulin G4-related disease (IgG4-RD) affecting the esophagus, stomach, and liver

A 60-year-old man, with occasional acid reflux, was found on computed tomography (CT) to have multiple masses in his esophagus (● Fig. 1 a), stomach (● Fig. 1 d), and liver. He had a history of partial hepatectomy for inflammatory liver pseudotumor on two occasions, 22 and 9 years earlier. Physical examination revealed no superficial lymph node. Blood test showed that hemoglobin was 105 g/L, and the tumor biomarkers CEA, AFP and CA199 were normal. Gastroscopy revealed a hard, fixed mass in the lower esophagus covered with normal appearing mucosa (● Fig. 1 b), and a giant stomach ulcer with a clean and hard base (● Fig. 1 e). Histological examination showed abundant lymphocytes, plasma cell infiltration, and fibrosis in biopsies from the esophagus (● Fig. 1 c), and stomach (● Fig. 1 f). The number of IgG4 positive cells was greater than 50 per high-power field (hpf) (● Fig. 2 a, b), and the ratio of IgG4-positive to

IgG-positive cells was greater than 30% in both specimens. Meanwhile, similar results were found in the inflammatory liver pseudotumor excised 9 years earlier (● Fig. 2 c, d). Moreover, the concentration of serum IgG4 was 1590 mg/L (normal range: 80–1400 mg/L). Therefore, the patient was diagnosed as having immunoglobulin G4-related disease (IgG4-RD). After a 3-month period of steroid therapy and anti-ulcer treatment, fresh epithelial tissue had regenerated in the margin of the stomach ulcer and the stomach wall had become softer (● Fig. 3 b). Neither regression nor progression of the mass in the esophagus were observed by endoscopy (● Fig. 3 a), or CT scanning. IgG4-RD is an autoimmune disorder characterized by IgG4 positive plasma cell infiltration, fibrosis, phlebitis, and increased serum IgG4 [1]. IgG4-RD always mimics malignancy clinically and responds to steroids [2]. Many cases of IgG4-RD affect-

ing extrapancreatic organs have been reported [3]. To the best of our knowledge, this is the first documented case of IgG4-RD affecting the esophagus, stomach, and liver. It is highlighted that IgG4-RD should be considered as a possible diagnosis for multiple masses in the gastrointestinal tract, which may avoid unnecessary surgery or chemotherapy.

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Competing interests: None

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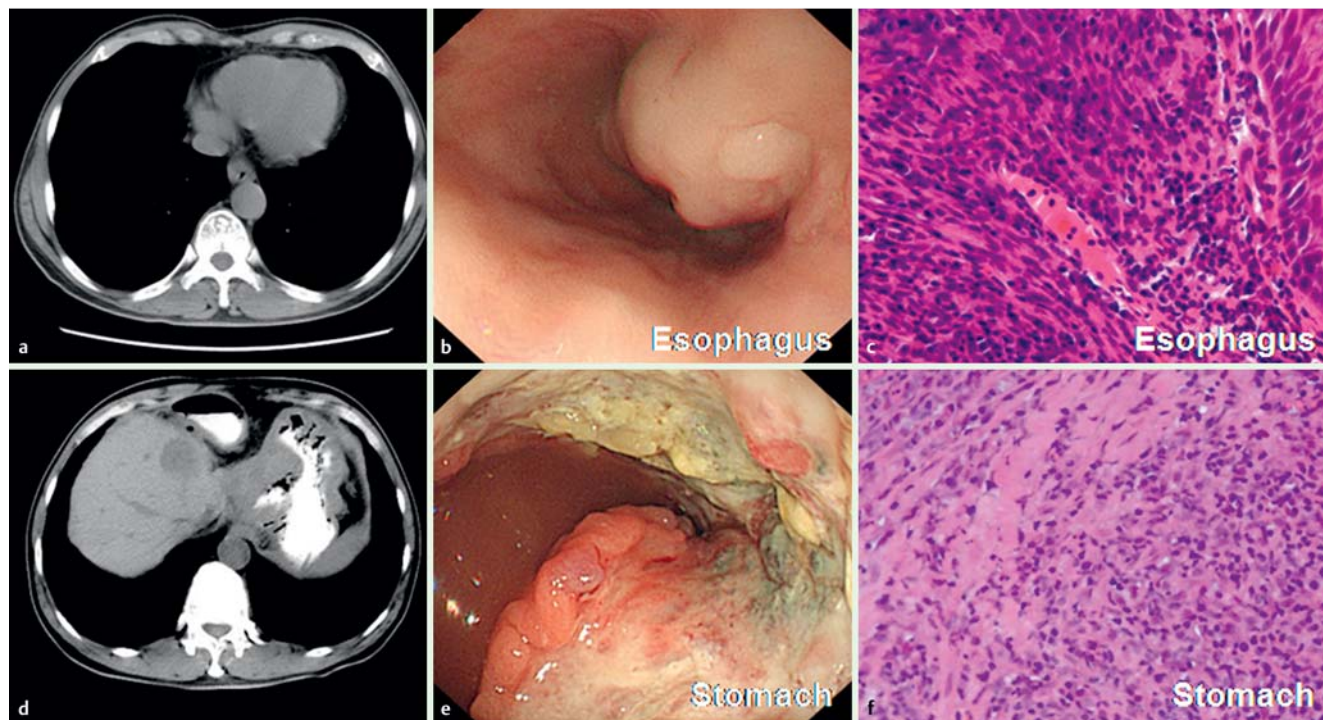


Fig. 1 Immunoglobulin G4-related disease (IgG4-RD) in a 60-year-old man. **a** Esophageal mass seen at computed tomography (CT); **b** endoscopic view in the lower esophagus of hard fixed mass covered with normal appearing mucosa; **c** histological examination showed abundant lymphocytes, plasma cell infiltration, and fibrosis. **d** Stomach mass seen at CT; **e** endoscopic view of giant stomach ulcer with clean hard base; **f** histological findings were similar to those for the esophagus.

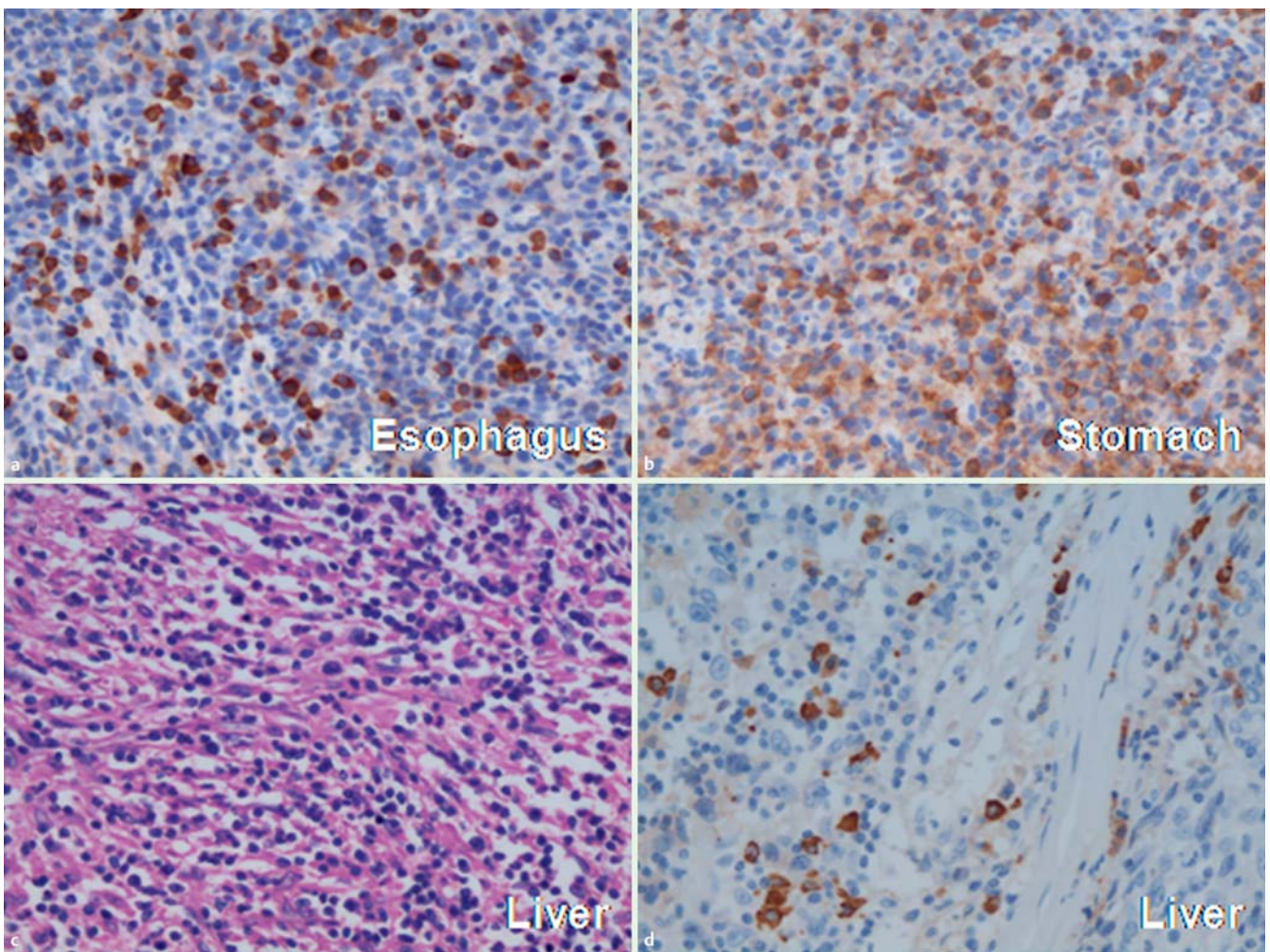


Fig. 2 Immunoglobulin G4-related disease (IgG4-RD). **a, b** IgG4-positive cells seen in esophageal and stomach biopsies in the 60-year-old patient. **c, d** Similar histological findings from the inflammatory liver pseudotumor excised from the same patient 9 years previously.

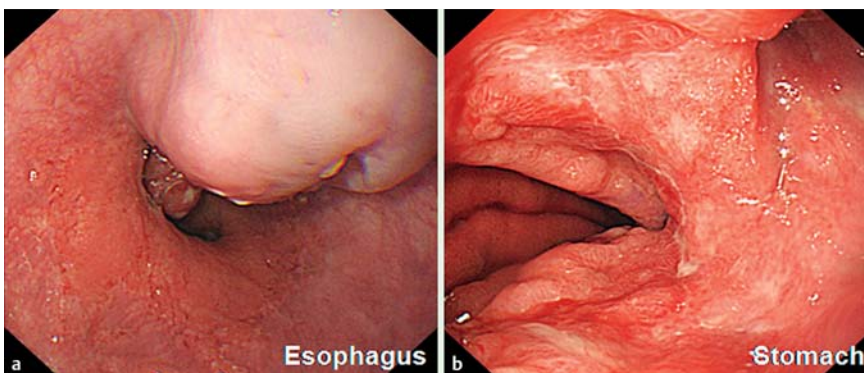


Fig. 3 Endoscopic view of the lesions after the 3-month period of therapy; **a** esophagus showing neither regression nor progression of the mass; **b** stomach showing fresh epithelial tissue regenerated in the margin of the stomach ulcer, with a softer stomach wall.

Bibliography

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