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 affecting 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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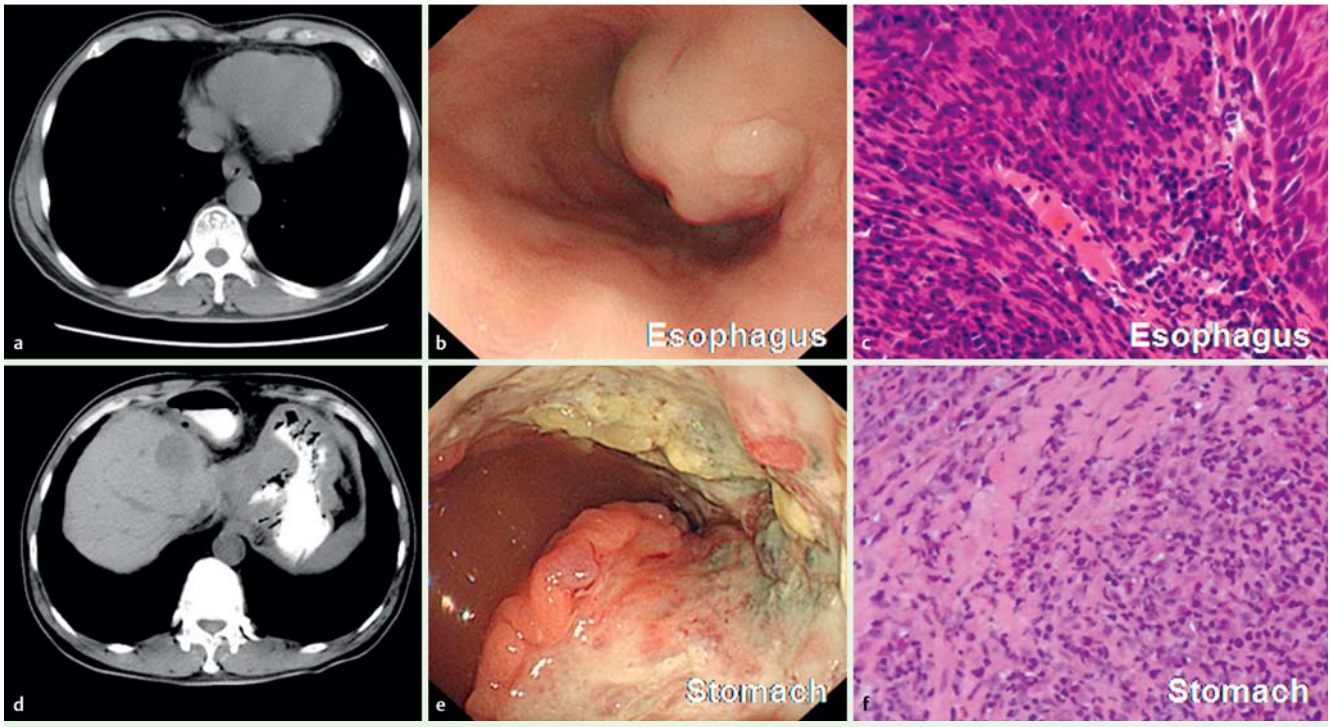

Fig. 1 Immunoglogbulin G4-related disease (IgG4-RD) in a 60-year-old man. **a** Esophageal mass seen at computed tomography (CT); **b** endoscopic view in the lower esophagus of hard fixed mass covered with normal appearing mucosa; **c** histological examination showed abundant lymphocytes, plasma cell 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.

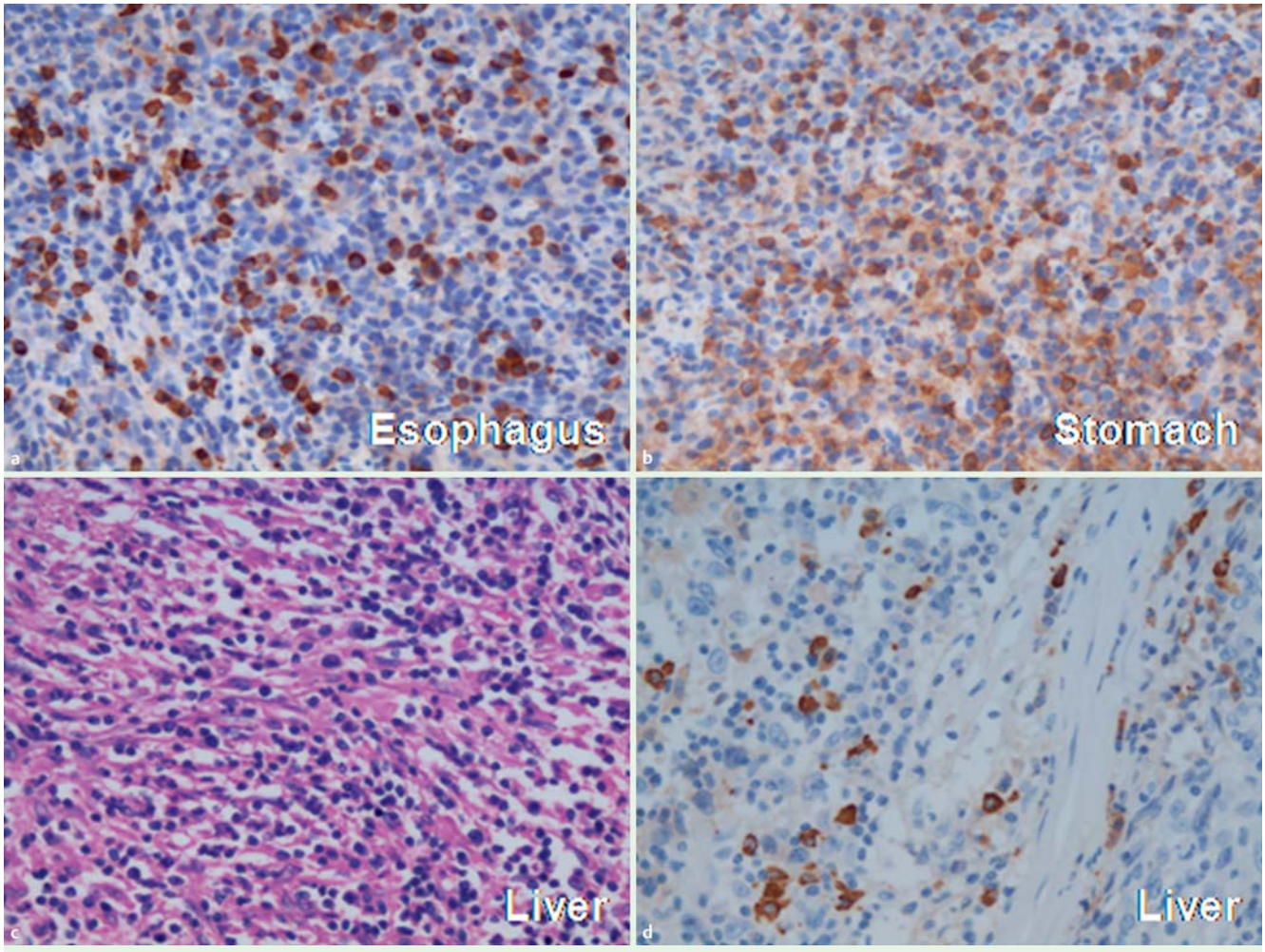

Fig. 2 Immunoglogbulin G4-related disease (IgG4-RD). **a, b** IgG4-positive cells seen in esophageal and stomach biopsies in the 60-year-old patient. **c, d** Similar histological findings from the inflammatory liver pseudotumor excised from the same patient 9 years previously.

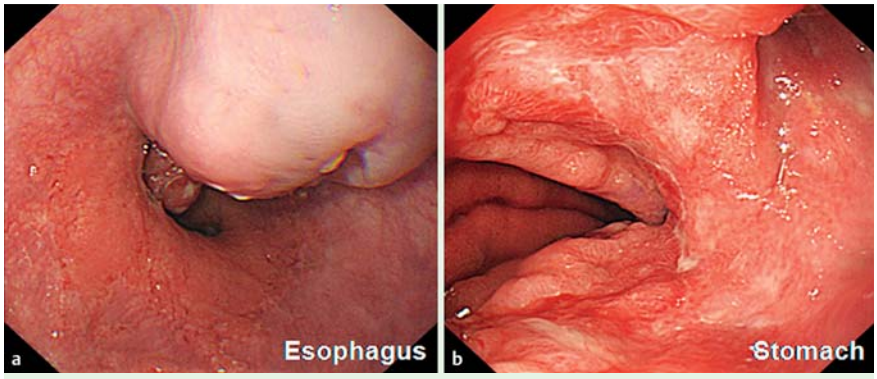

Fig. 3 Endoscopic view of the lesions after the 3-month period of therapy; **a** esophagus showing neither regression nor progression of the mass; **b** stomach showing fresh epithelial tissue regenerated in the margin of the stomach ulcer, with a softer stomach wall.

References

1 *Smyrk TC*. Pathological features of IgG4-related sclerosing disease. Curr Opin Rheumatol 2011; 23: 74 – 79
2 *Morse B*, *Centeno B*, *Vignesh S*. Autoimmune pancreatitis: updated concepts of a challenging diagnosis. Am J Med 2014; 127: 1010.e1 – 1010.e9
3 *Kim JH*, *Byun JH*, *Lee SS* et al. Atypical manifestations of IgG4-related sclerosing disease in the abdomen: imaging findings and pathologic correlations. AJR Am J Roentgenol 2013; 200: 102 – 112

Bibliography
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