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. 1a), stomach (Fig. 1d), 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. 1b), and a giant stomach ulcer with a clean and hard base (Fig. 1e). Histological examination showed abundant lymphocytes, plasma cell infiltration, and fibrosis in biopsies from the esophagus (Fig. 1c), and stomach (Fig. 1f). The number of IgG4 positive cells was greater than 50 per high-power field (hpf) (Fig. 2a, 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. 2c, 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. 3b). Neither regression nor progression of the mass in the esophagus were observed by endoscopy (Fig. 3a), 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

Lang Yang, Peng Jin, Jian-qiu Sheng
Department of Gastroenterology, General Hospital of Beijing Military Command, Beijing, China

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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.

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Corresponding author
jian-qi Sheng
Department of Gastroenterology
General Hospital of Beijing Military Command
Nanmencang S#
Dongcheng District
100700 Beijing
China
Fax: +86-10-66721299
jianqi@263.net