Primary gastroduodenal amyloidosis

Amyloidosis is characterized by tissue deposition of insoluble fibrillar proteins in various organs [1]. In humans, more than 23 different and unrelated proteins are known to form amyloid fibril [2]. Amyloidosis is divided into primary (i.e. idiopathic) and secondary amyloidosis (i.e. associated with chronic inflammatory conditions, and infectious and neoplastic disorders) [1]. Primary amyloidosis is extremely rare in the gastrointestinal tract. Fewer than 1% of patients with primary amyloidosis in the gastrointestinal tract have any symptoms [3]. We report a case of primary gastroduodenal amyloidosis in which endoscopic ultrasound (EUS) was instrumental in the work-up.

A 76-year-old man presented with a history of fatigue, dyspepsia, and anemia. An upper-gastrointestinal endoscopy revealed prominent gastric folds and gastropathy (Fig. 1a). Gastric biopsies showed only signs of unspecific inflammation. The patient developed diarrhea and weight loss. Abdominal computed tomography (CT) showed unspecific gastric wall thickening. The initial suspicion was a malignant disease such as scirrhous carcinoma, which led to an EUS referral. EUS revealed gastric wall thickening (Fig. 1b) and a complete lack of normal sonographic layers in the stomach wall, suggestive of an infiltrative disease (Fig. 1b), but no sign of malignancy. Another upper-gastrointestinal endoscopy was undertaken with new biopsies from the stomach and bulbus duodeni. Congo red staining revealed amorphous eosinophilic infiltrates (Fig. 2a) and green birefringence under polarized light (Fig. 2b), which is diagnostic for amyloidosis [1]. It may be suggested that tissue deposition of insoluble proteins causes the sonographic disappearance of normal gastric wall layers in amyloidosis. This patient had no systemic disease such as a chronic immune disease or neoplastic disorder, which are the most common causes of secondary amyloidosis. Thus this patient had a primary amyloidosis in the gastroduodenum. In conclusion, this case highlights the use of EUS in the work-up of patients with suspected amyloidosis in the stomach and duodenum.

Fig. 1 a Endoscopic image of the stomach. The endoscopic view shows prominent and irregular gastric folds in the corpus, with food remnants. b Endoscopic ultrasound (EUS) image of the stomach, revealing increased gastric wall thickening (9.6 mm) and loss of the typical 1–3 sonographic wall layers, although layer 4 (muscularis propria) is normal (arrow).

Fig. 2 Photomicrographs from gastric biopsies, stained with Congo red. a Sample viewed with normal light revealed amorphous eosinophilic infiltrates in the interstitium (arrow). b Sample viewed with polarized light showed green birefringence (arrow) of areas stained with Congo red, which is diagnostic for amyloidosis.

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Competing interests: None

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