Asymptomatic Localized Gastric Amyloidosis with Two Separate Lesions

Joo Hyun Lim¹, Jung Kim², Ji Yeon Seo¹, Jung Ho Bae¹

Department of Internal Medicine, Healthcare Research Institute, Seoul National University Hospital Healthcare System Gangnam Center¹,
Department of Internal Medicine and Liver Research Institute, Seoul National University College of Medicine², Seoul, Korea

Amyloidosis is a disease in which amyloid is abnormally accumulated in the tissue. The kidney and heart are the most commonly involved organs. Gastric involvement is relatively common in systemic disease; however, localized gastric amyloidosis is rare. Here, we report a case of localized gastric amyloidosis with two separate lesions. A 56-year-old woman underwent a health surveillance checkup without any noticeable symptoms. She was under medication for diabetes and dyslipidemia, and was otherwise healthy. On surveillance upper endoscopy, an irregularly shaped hyperemic elevated erosion at the gastric fundus and a 1.5-cm, yellowish subepithelial tumor-like lesion with intact overlying mucosa at the lesser curvature of the lower body of the stomach were detected. Endoscopic biopsy revealed submucosal eosinophilic material deposition in both lesions. Congo-red staining showed amyloid deposit appearing as a yellow-green birefringence under polarizing microscopy. Echocardiography, abdominal sonography, and colonoscopy revealed no abnormality. The patient was diagnosed as having localized gastric amyloidosis and is now undergoing regular follow-up without any treatment. Localized gastric amyloidosis is a rare disease that may mimic nonspecific gastritis or subepithelial tumor. However, endoscopic biopsy with appropriate staining may be diagnostic and thorough evaluation for systemic involvement is important. (*Korean J Helicobacter Up Gastrointest Res* 2018;18:131-134)

Key Words: Amyloidosis; Endoscopy; Stomach

INTRODUCTION

Amyloidosis is a rare disease in which amyloids, the insoluble fibrillar proteins are abnormally accumulated in the intercellular matrix.¹ It can be categorized into systemic and localized type according to the distribution of accumulated amyloids. In most cases, amyloidosis manifests as a systemic disease and localized amyloidosis is extremely rare. In systemic amyloidosis, the most commonly affected organs are kidney, heart and liver. Gastrointestinal tract is also commonly involved in systemic amyloidosis; however, localized gastric involvement is considerably rare and its manifestations are hardly characterized.²⁻⁴ Here, we report a case of localized gastric amyloidosis with two separate lesions diagnosed during surveillance endoscopy in a healthy adult.

CASE REPORT

A 56-year-old female underwent health surveillance checkup without any noticeable symptoms. She was under medication for diabetes and dyslipidemia and otherwise healthy. On surveillance upper endoscopy, an irregularly-shaped hyperemic elevated erosion was found at the gastric fundus (Fig. 1A) and a 1.5-cm sized yellowish subepithelial tumor (SET)-like lesion with intact overlying mucosa was detected at the lesser curvature side of the lower body of the stomach (Fig. 1B). Endoscopic biopsy revealed submucosal eosinophilic material deposition in both lesions (Fig. 2A, B). Congo-red stain revealed amyloid deposit appearing as yellow-green birefringence under polarizing microscopy (Fig. 2C, D). Stain for smooth muscle actin and desmin were all negative. Laboratory findings demonstrated no specific abnormality. Immunoglobulin G antibody for *Helicobacter pylori* was negative. Urinalysis showed no evidence of Bence Jones proteinuria. Serum free light chain levels were all within normal ranges, including kappa-free light chain 16.37 mg/dL (normal range, 3.3~19.4 mg/dL), lambda-free light 20.47

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Corresponding author: Joo Hyun Lim

Department of Internal Medicine, Healthcare Research Institute, Seoul National University Hospital Healthcare System Gangnam Center, 38~40FL, Gangnam Finance Center, 152 Teheran-ro, Gangnam-gu, Seoul 06236, Korea

Tel: +82-2-2112-5807, Fax: +82-2-2112-5635, E-mail: joohyunlim@snuh.org

ORCID: <https://orcid.org/0000-0002-8437-096X>

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Fig. 1. Endoscopic findings. (A) An irregularly shaped hyperemic elevated erosion at the gastric fundus. (B) A 1.5-cm, yellowish subepithelial tumor-like lesion with intact overlying mucosa at the lesser curvature side of the lower body of the stomach.

Fig. 2. Histopathological findings. (A, B) Endoscopic biopsy from both lesions showing submucosal eosinophilic material deposition (H&E; ×40). (C, D) Congo-red staining showing amyloid deposits appearing as yellow-green birefringence under polarizing microscopy (Congo-red staining; C: ×100, D: ×400).

mg/dL (normal range, 5.71~26.3 mg/dL), and kappa/lambda ratio 0.8 (normal range, 0.26~1.65). Echocardiogram, low dose screening chest computed tomography, abdominal sonogram, and colonoscopy did not show any significant findings. Because this patient had no symptom and no evidence of systemic involvement, she was diagnosed as localized gastric amyloidosis and she did not undergo any treatment. She is now on regular follow-up without any discomfort for 10 months.

DISCUSSION

Amyloidosis is a disease where abnormal protein, amyloid, accumulates in the tissues. Usually it occurs systemically involving multiple organs. However, sole gastric involvement is extremely rare. Systemic amyloidosis involving the gastric tissue usually shows unfavorable prognosis. Although primary localized gastric amyloidosis was previously thought to present poor prognosis,⁵ these days, it is known that localized amyloidosis confined to stomach

has relatively good prognosis.^{6,7} Therefore, accurately confirming the range of involvement is very important. Until 1990s surgical resection was considered as proper treatment for localized gastric amyloidosis,^{2,8} while after 2,000 most cases with localized gastric amyloidosis, especially symptom-free ones, maintained good condition without any treatment.^{3,6,9-14}

So far, there have been no diagnostic criteria for localized gastric amyloidosis. It is only diagnosed by excluding other organ involvement. Once amyloid deposit is detected pathologically, any possible underlying disorder such as multiple myeloma or chronic inflammatory disorders should be ruled out first with physical examination and laboratory findings. And then, to exclude systemic involvement, examinations for other organs which are usually affected in systemic amyloidosis should be performed. To rule out renal involvement, urinalysis for Bence Jones proteinuria is needed, and to exclude cardiac involvement echocardiography is warranted. Also, colonoscopy, chest and abdominal imaging should be performed to exclude other gastrointestinal tract or solid organ involvement.

Gastric involvement of systemic amyloidosis presents as various symptoms and endoscopic findings. Its symptoms vary from nonspecific dyspepsia to hematemesis or epigastric pain and the endoscopic findings are also nonspecific, including erosions, fine granularity, mucosal friability, thickened folds, hematoma, polyps, or ulcer.^{15,16} Similarly, localized gastric amyloidosis does not present any characteristic symptoms or endoscopic findings. Therefore only endoscopic biopsy can confirm the diagnosis. Previously reported cases showed various type of gross morphology, from early gastric cancer-like depressed lesion to scirrhous cancer-like fold thickening or SET. The current case had two separate lesions with different shape; one was erosion-like and the other was SET-like. Although they were initially assumed to have different pathology, both turned out to be amyloidosis. So far there has been only one case of localized gastric amyloidosis presenting two separate lesions with different morphology.⁵ However, that case was more aggressive one with epigastric pain and was treated endoscopically. On the contrary, our case was asymptomatic and continued good condition without any treatment for 10

months. As far as we know this is the first case of asymptomatic localized gastric amyloidosis with two separate lesions showing different endoscopic findings.

In conclusion, since localized gastric amyloidosis can present as various type and number of lesions, gastric amyloidosis should be one of the differential diagnoses for nonspecific abnormal endoscopic findings and accurate diagnosis with biopsy is important. Once pathologic diagnosis is confirmed, thorough examination to exclude systemic involvement should be performed.

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