

A Case of Systemic Amyloidosis that Presented as Secondary Achalasia

서울아산병원 소화기내과

서준영, 정기욱

Background: Achalasia is a rare disorder that makes it difficult for food and liquid to pass from the esophagus to the stomach. Primary achalasia is a benign disorder of esophageal motility. Secondary achalasia can be provoked by various etiologies, most frequently malignancies. Herein, we describe a female patient with secondary achalasia caused by amyloidosis.

Case: A 62-year-old female complaining of dysphagia was referred to hospital. She complained of solid-and-liquid type dysphagia, weight loss (22kg/6months), and recurrent vomiting 5 months prior to her visit. The patient was admitted to hospital for evaluation. High resolution manometry revealed relaxation failure of the lower esophageal sphincter, with panesophageal pressurization, suggesting a diagnosis of type2 achalasia(Fig1). Esophagogastroduodenoscopy showed a subepithelial lesion on the high body(Fig2-A,B). Pathologic exam revealed an amorphous afibrillary eosinophilic deposit on lamina propria, and apple-green colored amyloid proteins were observed by polarizing microscopy using Congo-red stain(Fig3-A,B). Various further studies were conducted to elucidate secondary symptoms associated with amyloidosis. Laboratory findings revealed an inverted albumin/globulin ratio (alb: 2.0g/dL, total protein: 5.2g/dL) and proteinuria(1354mg/dL/day). Therefore, kidney biopsy and protein electrophoresis (PEP) were performed. Cast nephropathy with monoclonal gammopathy was noted at kidney biopsy, and PEP revealed Bence-Jones proteinuria. Bone marrow biopsy was performed, supporting a diagnosis of monoclonal gammopathy of undetermined significance(MGUS). Taken together, achalasia, nephrotic syndrome, and MGUS were thought to be related to amyloidosis. She is currently under follow up care with the hematology department.

Conclusion: Secondary achalasia is most frequently caused by a malignant tumor at the esophagogastric junction. Intestinal amyloidosis has been reported to cause rare cases of achalasia, but no previous cases have been reported in South Korea. Therefore, when secondary achalasia is suspected, accurate diagnosis of etiology should be carefully made because amyloidosis, can produce symptoms of achalasia

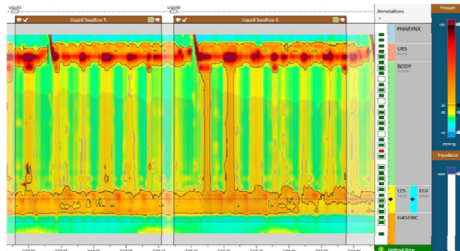


Figure 1. Elevated median integrated relaxation pressure (34 mmHg), and failure of relaxation were noted by high resolution manometry, suggesting a diagnosis of type II achalasia.

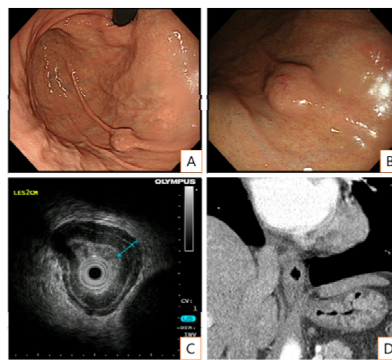


Figure 2. (A), (B) An approximately 1.5-cm sized sub-epithelial lesion with focal nodularity was noted on the great curvature side of the high body. (C) Endoscopic ultrasound showed a thickened esophageal proper muscle layer at the esophagogastric junction, suggesting a diagnosis of achalasia. (D) Computed tomography also showed esophageal wall thickening.

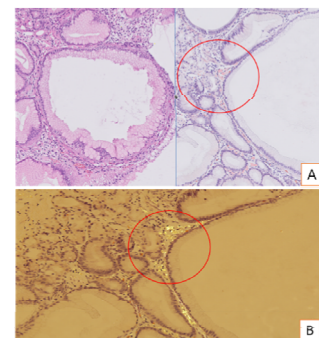


Figure 3. (A) Multiple amorphous afibrillary eosinophilic deposits were noted at the gastric lamina propria by pathologic exam. (B) Congo-red staining revealed scarlet-colored amorphous materials with apple-green birefringence under polarized light, supporting a diagnosis of amyloidosis.