ORIGINAL ARTICLE

Peroral endoscopic myotomy for a pediatric case of suspected congenital esophageal stenosis

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INTRODUCTION

Peroral endoscopic myotomy (POEM) is becoming one of the main treatment options for pediatric achalasia.[1] Congenital esophageal stenosis (CES) is a rare disease that can have similar symptoms, endoscopic features, and radiographic findings to achalasia, which often leads to difficulty in distinguishing between the 2 conditions. We present an extremely rare case that was highly suspicious for CES interprocedurally and was successfully treated with modified POEM (Video 1, available online at www.videogie.org).

CASE

A 5-year-old girl with trisomy 21 was referred to our center for the treatment of suspected achalasia. She experienced dysphagia with every meal, occasional regurgitation, and poor weight gain after she started solid foods as an 18-month-old. Although she received 2 sessions of endoscopic balloon dilation, her symptoms did not improve.

Endoscopic findings revealed a tight esophagogastric junction (EGJ) (Fig. 1A), and an endoscopic miniprobe US demonstrated a thickened muscle layer without any signs of tracheobronchial remnants (Fig. 1B). Dilatation and stasis of the esophagus, with narrowing at the EGJ, was observed during the esophagram (Fig. 1C). High-resolution manometry was not performed because of the patient's age.

We agreed with the suspicion of achalasia because of the endoscopic and radiographic findings, although CES could not completely be excluded. The patient was scheduled for POEM.

Informed consent was obtained from the patient's parent as substitute decision maker for the publication of her information and imaging.

Abbreviations: CES, congenital esophageal stenosis; EGJ, esophagogastric junction; POEM, peroral endoscopic myotomy.

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PROCEDURE

The mucosal entry was made at the posterior wall of the esophagus, and the submucosal tunnel was created. However, the submucosal layer became gradually narrower as we approached the EGJ. At this point, the network of muscle fibers became dense and intricate (Fig. 2A), and separating the muscle layer from the mucosa became difficult. These features are very atypical for achalasia; thus, we suspected fibromuscular thickening–type CES.

Creation of the submucosal tunnel was halted before passage through the stenotic area. Full-thickness myotomy was started from the oral side of the stenosis while separating the muscle layer from the mucosa as much as possible (Fig. 2B). The myotomy continued through the stenotic area and just into the stomach. Excessive air insufflation should be avoided when performing full-thickness myotomy without a submucosal tunnel to not induce gas-related adverse events. Using water jet injection was a helpful technique for ensuring a safe distance from the mucosa to the endoknife and avoiding inadvertent mucosotomy.

Once the myotomy reached the gastric side, EGJ relaxation was observed. The entry was closed after 2 to 3 cm of additional myotomy was added proximal to the stenotic area.

OUTCOME

The Eckardt score was decreased from 4 to 1, and the endoscopic and radiographic findings improved 3 months after the POEM (Fig. 3).

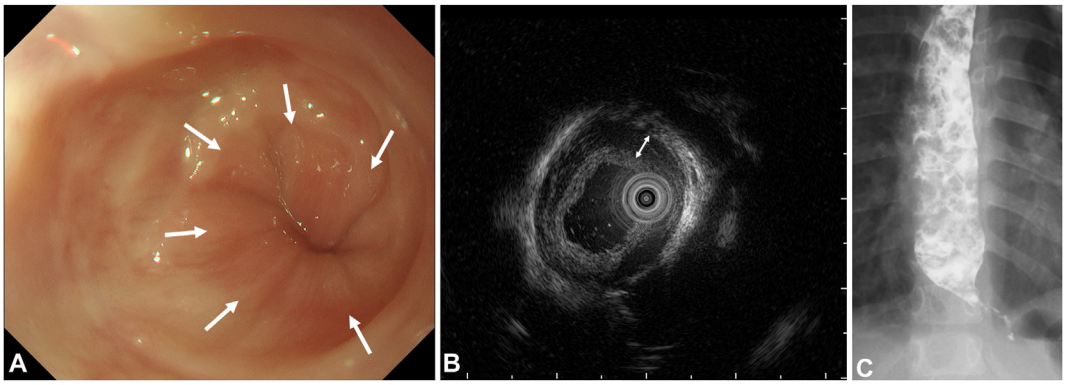

Figure 1. A, Endoscopic findings before peroral endoscopic myotomy. **B,** Miniprobe US (20 MHz). **C,** Esophagram before peroral endoscopic myotomy.

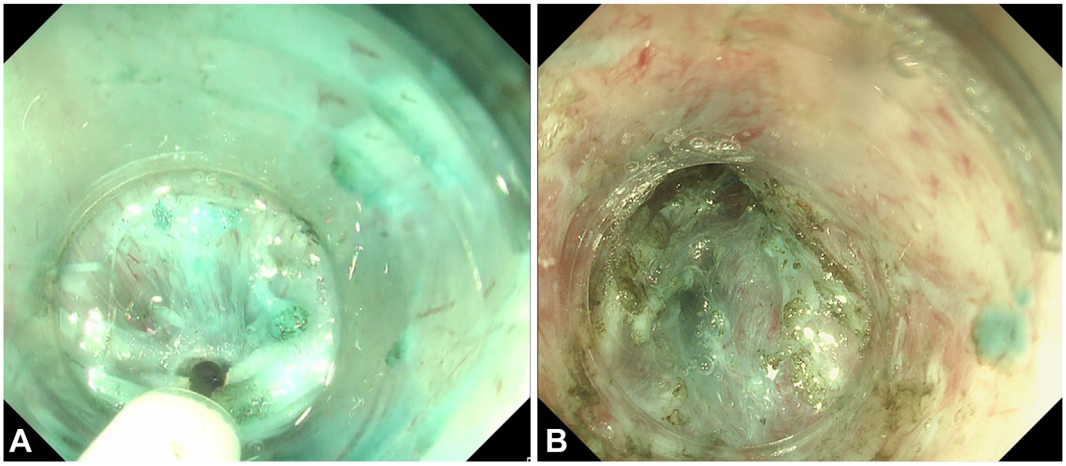

Figure 2. A, Narrowed submucosal layer and intricate muscle fibers at the stenotic area. **B,** Myotomy at the stenotic area.

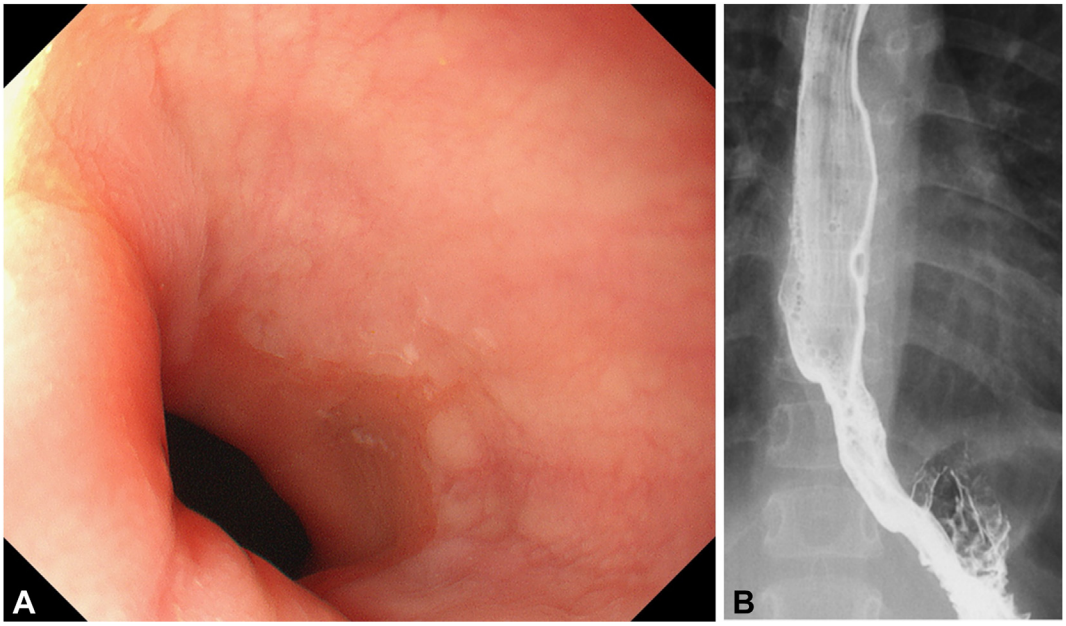

Figure 3. A, Endoscopic findings after peroral endoscopic myotomy. **B,** Esophagram after peroral endoscopic myotomy.

DISCUSSION

To our knowledge, this is the first report of pediatric CES with successful treatment via a modified POEM procedure. CES is divided into 3 types based on its histological findings: tracheobronchial remnants, which involves ciliated pseudo-stratified columnar epithelium, seromucous glands, or cartilage; fibromuscular thickening, which is characterized by hypertrophic muscle fiber with fibrosis; and membranous web.[2,3] The endoscopic findings of the fibromuscular thickening type are particularly similar to achalasia. Cartilaginous components are not present and thus not observed by the EUS, which makes it difficult to distinguish this subtype from achalasia.

Although the definitive diagnosis of CES is made by the pathologic examination of resected esophagus, we concluded the patient has CES based on the suspicious clinical and endoscopic findings. The immediate onset of symptoms after starting solid food and the associated anomaly of trisomy 21^2 point away from achalasia. Endoscopically, findings more typical of CES were present, that is, (1) narrowed submucosal layer, (2) intricate muscle fibers at the stenotic area, and (3) difficulty in separating the muscle layer from the mucosa. Ikeda et al[4] reported the effectiveness of modified POEM for cases of adult congenital stenosis. We applied this technique, which led to a favorable outcome for this pediatric patient.

Other preoperative findings, such as ring-shape stenosis on endoscopy, lopsided hourglass sign on barium esophagogram, and a compartmentalized intrabolus pressure pattern with distinction between the stenotic area and the lower esophageal sphincter on manometry, can be supportive for the diagnosis of CES.[4] Nevertheless, distinguishing CES from achalasia before POEM is sometimes difficult, as shown in the present case. Thus, it is important to recognize the challenge in preoperative diagnosis of CES and the possible need for conversion to a modified approach when the aforementioned intraprocedural findings are observed during pediatric POEM.

DISCLOSURE

Dr Toyonaga has ownership interest in Fujifilm. The other authors did not disclose any financial relationships.

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