Usefulness of Endoscopic Ultrasonography in the Diagnosis of Congenital Esophageal Stenosis

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Background/Purpose: Endoscopic ultrasonography (EUS) is considered a potentially useful tool to investigate structural abnormalities of the esophagus in pediatric patients, as in adults. The aim of this study was to evaluate the usefulness of EUS for the diagnosis of congenital esophageal stenosis.

Methods: High-frequency catheter probe EUS was performed under general anesthesia in 2 patients who had congenital esophageal stenosis.

Results: A 4-year-old boy with anorectal anomaly showed tapered narrowing in the distal esophagus, which was not ameliorated with balloon dilatation. High-frequency catheter probe EUS showed hypertrophy of the muscular layer in the esophageal wall at the narrowed portion, but no images suggested the presence of tracheobronchial remnants. The histologic diagnosis of fibromuscular hypertrophy was confirmed at esophagoplasty. A 5-month-old boy with Gross C-type esophageal atresia and symptomatic gastroesophageal reflux showed tapered narrowing in the middle esophagus on esophagography. The symptoms of stenosis were not ameliorated by balloon dilatation performed 4 times. High-frequency catheter probe EUS showed hyperechoic lesions suggesting cartilage at the esophageal narrowing. The diagnosis of tracheobronchial remnants was confirmed by the finding of 2 pieces of cartilage in the specimen obtained at the time of esophageal resection.

Conclusion: EUS can be applied to show structural abnormalities of the esophageal wall even in pediatric patients with congenital esophageal stenosis and is useful for planning the therapeutic strategy.

INDEX WORDS: Endoscopic ultrasonography, congenital esophageal stenosis, fibromuscular hypertrophy, tracheobronchial remnants, high-frequency catheter probe.

CONGENITAL esophageal stenosis (CES) is a rare anomaly, which usually presents in infancy or childhood. The causes of CES are divided into 3 histopathologic types: tracheobronchial remnants (TBR), membranous diaphragm (MD), and fibromuscular hypertrophy (FMH). Resection of the esophagus has been considered essential in cases of TBR, whereas dilatation is effective in some cases of FMH. It would be worthwhile to make a differential diagnosis of FMH and TBR in planning the therapeutic strategy.[1] We report our experience of application of high-frequency catheter probe endoscopic ultrasonography (EUS) to make the diagnosis of FMH and TBR in pediatric patients with CES.

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CASE REPORTS

Case 1

A 3-year-old boy with trisomy 21 experienced vomiting after colostomy closure for anorectal anomaly, coinciding with an increase of solid food intake. Esophagography showed a tapered narrowing in the distal esophagus with dilatation of the proximal esophagus (Fig 1A). Esophagoscopy showed a narrowing 2 cm proximal to the esophagogastric junction. Esophageal pH monitoring showed absence of gastroesophageal reflux (GER). The diagnosis of CES was made, and balloon dilatation of the stenotic esophagus was performed with a Rigiflex balloon dilator under general anesthesia. Because of persistent dysphagia, the patient underwent esophagoscopy followed by EUS at 4 years of age. EUS using a high-frequency catheter probe (UM-BS20-26R, 20MHz; Olympus Optical, Tokyo, Japan) was performed with a balloon sheath attached onto the esophageal mucosa, and the image was observed on an endoscopic ultrasonographic processor (EU-M30S; Olympus Optical). EUS showed that the thickness of the esophageal muscular layer was 2.5 mm at the narrow portion, which was 1 mm thicker than that in the other normal portion (Fig 2A). No images suggested the presence of TBR in the esophageal wall. Balloon dilatation was performed again with a Rigiflex balloon dilator. Heinecke-Mikulicz–type esophagoplasty was performed at 4 years, 6 months of age because of the recurrence of symptoms. The specimen obtained from the esophageal wall showed hypertrophy of the muscular and submucosal layers with fibrosis, consistent with the findings of EUS.

Case 2

A boy weighing 1,522 g was born at 33 weeks' gestation. Gross C-type esophageal atresia was diagnosed, and he underwent primary repair with gastrostomy. Postoperative esophagography showed a mild

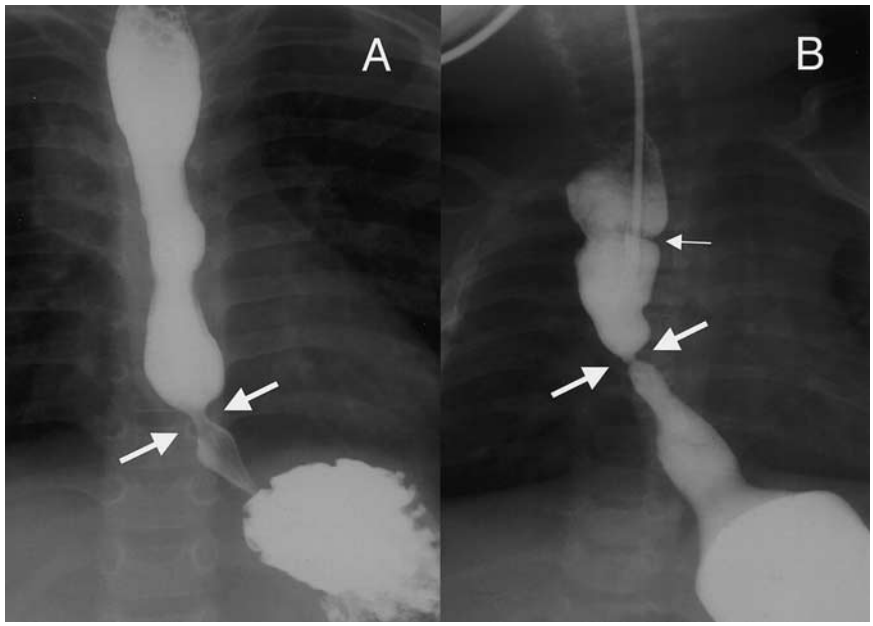

Fig 1. (A) Esophagogram in patient 1 at 3 years of age. A tapered narrowing (arrows) in the distal esophagus with dilation of the proximal esophagus is seen. (B) Esophagogram in case 2 at 5 months of age. A severe narrowing in the middle esophagus (large arrows) with GER is seen. Small arrow indicates the anastomotic site of primary repair for esophageal atresia.

tapered narrowing in the middle esophagus 2 cm anal to the anastomotic site. GER also was seen. Although associated CES was suspected and balloon dilatation was performed 4 times, the stenosis worsened gradually because of reflux esophagitis (Fig 1B). EUS using a high-frequency catheter probe (UM-S30-20R, 30MHz; Olympus Optical) was performed. After filling the lumen with deaerated water, the probe was positioned in the narrowing via endoscopy. The image was observed on an endoscopic ultrasonographic processor EU-M30S. EUS showed 2 hyperechoic lesions with an acoustic shadow in the stenotic portion, suggesting the presence of cartilage in the esophageal wall (Fig 2B). Segmental resection of the esophageal narrowing, 2 cm in length, followed by Collis-Nissen fundoplication, was performed at 8 months of age. Ectopic tracheobronchial tissues, including cartilage and ciliated epithelium, were found in the specimen. The clinical course after surgery was uneventful.

DISCUSSION

Nihoul-Fékété[2] classified the causes of congenital malformation of esophageal wall architecture into 3 types; the presence of ectopic tracheobronchial tissue (TBR), the presence of a membranous diaphragm (MD), and segmental hypertrophy of the muscularis and submucosal layers with diffuse fibrosis (FMH). CES caused by TBR or MD tends to require surgical correction, whereas many patients with CES caused by FMH are treated only by bougienage or dilatation.[2,3] To differentiate these 3 types may be helpful in predicting the effectiveness of these conservative therapies.[1,3] To make a therapeutic strategy, a differential diagnosis of these types is desirable before treatment.

There have been few reports in which the presence of FMH was proved by EUS in adult patients with CES.[4,5] Takamizawa et al[1] has reported recently pediatric patients with CES in whom the presence of TBR was shown by EUS. We have shown the presence of TBR

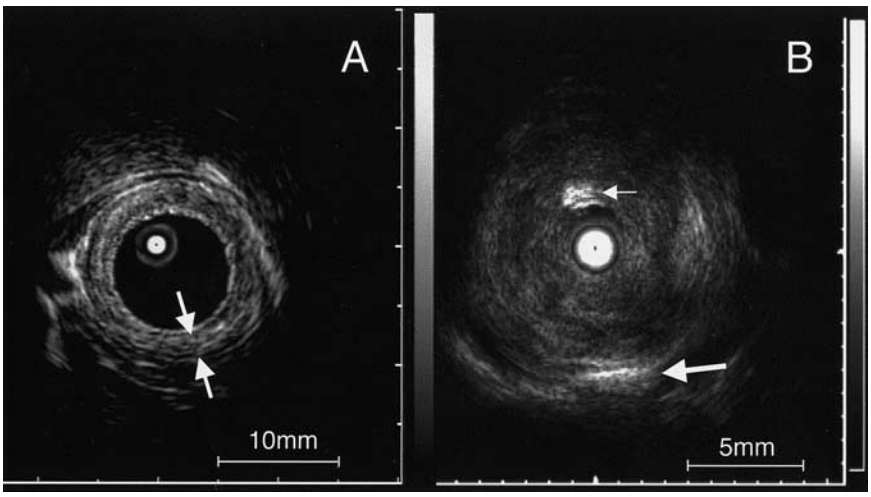

Fig 2. (A) Ultrasonography of the esophageal narrowing in case 1. The muscular layer (white arrows) is thickened to 2.5 mm. (B) Ultrasonography of esophageal narrowing in case 2. Two pieces of cartilage are seen as hyperechoic lesions. Small white arrow indicates cartilage just underneath the esophageal mucosa. Large white arrow indicates cartilage located deep in the muscle layer.

before surgery using high-frequency catheter probe EUS. High-frequency catheter probe EUS has been found to be a useful imaging technique in the evaluation of mucosal and submucosal lesions.[6] We attempted to use 2 kinds of catheter probes. One was 20 MHz in frequency and 2.6 mm in diameter with a balloon sheath, and the other was 30 MHz in frequency and 1.7 mm in diameter without a balloon sheath. Both of them were able to be passed through a 2.8-mm channel of an endoscope without difficulty. The higher the frequency of the probes, the higher resolution becomes, but the visual field is shallower.[7] In case 1, the balloon sheath system was useful to attach the probe onto the esophageal mucosa at a site where it was hard to retain deaerated water. We selected a thinner but higher-resolution probe in case 2, because the stricture was more severe and TBR was suspected. In that case, it was not difficult to fill the lumen with water because of severe narrowing. Although Takamizawa et al[1] reported that the cartilaginous component was visualized as a sonolucent area, cartilage was seen as a hyperechoic structure in our study. The inside of the cartilage layer is homogeneous and is imaged as a hypoechoic structure, whereas the cartilage interface with other tissues is imaged as a hyperechoic structure.[8] The difference in echogenic characteristics between these 2 studies may be caused by differences in the thickness of the ectopic cartilaginous structures. In the future, the accumulation of experience using EUS for examining the causes of CES would contribute to more accurate diagnosis, even in small infants

The causes of CES were shown clearly preoperatively with successful differential diagnosis using a high-frequency catheter probe EUS in the current cases. This method may show the structural abnormalities of the esophageal wall, even in small children, and may be useful for planning a therapeutic strategy for CES.

REFERENCES

1. Takamizawa S, Tsugawa C, Mouri N, et al: Congenital esophageal stenosis: Therapeutic strategy based on etiology. J Pediatr Surg 37:197-201, 2002

2. Nihoul-Fékété C, DeBacker A, Loatat-Jacob S, et al: Congenital esophageal stenosis: A review of 20 cases. Pediatr Surg Int 2:86-92, 1987

3. Kawahara H, Imura K, Yagi M, et al: Clinical characteristics of congenital esophageal stenosis distal to associated esophageal atresia. Surgery 129:29-38, 2001

4. McNally LC, Lemon JC, Goff JS, et al: Congenital esophageal stenosis presenting as noncardiac, esophageal chest pain. Dig Dis Sci 38:369-373, 1993

5. Katzka DA, Levine MS, Ginsberg GG, et al: Congenital esophageal stenosis in adults. Am J Gastroenterol 95:32-36, 2000

6. Das A, Sivak MV, Chak A, et al: High-resolution endoscopic imaging of the GI tract: A comparative study of optical coherence tomography versus high-frequency catheter probe EUS. Gastrointest Endosc 54:219-224, 2001

7. Schembre D, Chak A, Stevenss P, et al: Prospective evaluation of balloon-sheathed catheter US system. Gastrointest Endosc 53:758-763, 2001

8. Baba M, Sekine Y, Suzuki M, et al: Correlation between endobronchial ultrasonography (EBUS) images and histologic findings in normal and tumor-invaded bronchial wall. Lung Cancer 35:65-71, 2002