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CASE REPORT

Congenital esophageal stenosis due to fibromuscular thickening treated by circular myectomy - a case report

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ABSTRACT

Congenital esophageal stenosis (CES) is a rare congenital anomaly, with an estimated incidence of 1 in 25,000–50,000 live births. We report a unique case of CES caused by fibromuscular thickening in an 18-month-old girl. She presented with persistent vomiting that began at 4 months of age, coinciding with the introduction of solid foods. Contrast esophagography revealed a filiform stenosis in the lower third of the esophagus without evidence of gastroesophageal reflux disease (GERD). After GERD was excluded, the diagnosis of CES was confirmed by esophagogastroduodenoscopy. The patient underwent circular myectomy (extramucosal seromyotomy) combined with an anterior hemivalve anti-reflux procedure. The postoperative course was uneventful. Histopathological examination confirmed fibromuscular hypertrophy as the underlying cause of the stenosis. This case highlights the importance of considering CES in infants presenting with persistent vomiting after the introduction of solid foods. Early and accurate diagnosis, followed by appropriate surgical management, is essential for symptom resolution. It also demonstrates that circular myectomy is an effective primary treatment when endoscopic dilatation is not feasible.

Keywords: Congenital esophageal stenosis, fibromuscular thickening, Myectomy.

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1. INTRODUCTION

Congenital esophageal stenosis is a rare anomaly , with an incidence estimated at 1 per 25,000 to 50,000 live births [1]. It was initially reported by Frey and Duschl in 1936. In 1989, Nihoul-Fékété defined CES and categorized it based on three anatomopathological entities, stating that CES is caused by either fibromuscular thickening, tracheobronchial remnants, or membranous webs. Fibromuscular thickening is the second most frequent, after tracheobronchial remnants. It is most often located in the lower third of the esophagus and symptoms manifest when solids are introduced to the diet [1].

2. CASE REPORT

We report the case of an 18-month-old female patient who was delivered naturally. At the age of four months, she progressively began experiencing vomiting as soon as solid food was introduced without hematemesis. Her weight curve was appropriate for her age. Additionally, she had not suffered from any recurrent pneumonia. A per-oral contrast agent-supported chest X-ray showed a short, filiform stenosis of the lower third of the esophagus, with suprastenotic esophageal dilatation, but no signs of GERD (figure 1). Upper gastrointestinal endoscopy revealed an impassable stenosis at 18 cm. Achalasia was ruled out. The suprastenotic mucosa

appeared normal. A 24-hour pH monitoring study was performed and confirmed the absence of GERD. Therefore, the diagnosis of CES was established.

At the age of 20 months, she underwent surgery. A short esophageal stenosis, caused by a parietal fibrous ring, was described, surmounted by esophageal dilation. She underwent an extra-mucosal seromyotomy of the esophagus with subtotal resection of the parietal stricture ring. An anterior hemi-valve anti-reflux procedure was also performed. The postoperative course was uneventful. A per-oral contrast agent-supported chest X-ray showed the disappearance of the stenosis. Vomiting resolved, and the patient demonstrated appropriate weight gain. Histological examination of the surgical specimen revealed bundles of hypertrophied smooth muscle fibers, separated by dense fibrous tissue, consistent with fibromuscular hypertrophy; myenteric (Auerbach's) plexuses were identified and appeared normal in the resected specimen. The postoperative period proceeded without complications, and for more than five years now, there has been no recurrence or reflux. Normal growth in height and weight has been observed.

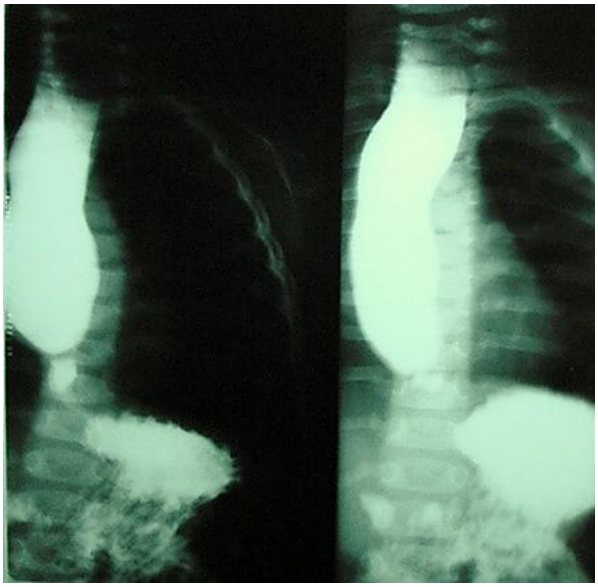

Figure 1. A per-oral contrast agent-supported chest X-ray showed filiform stenosis of the lower third of the esophagus.

3. DISCUSSION

Congenital esophageal stenosis is defined as the presence of intrinsic esophageal stenosis caused by an abnormality in the structure of the esophageal wall. The stenosis may or may not be symptomatic from the newborn period. Most cases have been described in the lower esophagus. The stenosis may be caused by ectopic tracheobronchial tissue, membrane webs, or hypertrophy of the muscular layer [2] as observed in our patient. Fibromuscular stenosis is less frequent than tracheobronchial remnants and involves segmental hypertrophy of the muscular and submucosal layers with diffuse fibrosis [1]. The stenotic segment often exhibits dysmotility. It is presumed that fibromuscular thickening may be linked to conditions such as achalasia of the esophagus or Hirschsprung disease [2].

There is no gender predisposition. A higher incidence has been observed in white populations [3]. Infants with CES usually tolerate breastfeeding, and start to present with regurgitation and vomiting upon the introduction of solid food. Symptoms typically begin at the age of 4 to 10 months but depend on the severity of the stenosis. A clinical sign of CES can be failure to thrive, recurrent respiratory infections, or during the follow-up of esophageal atresia surgery [2].

The first diagnostic step is to exclude the oropharyngeal causes of dysphagia by performing otorhinolaryngeal and neurologic evaluations. An indicative finding leads further to investigation through esophagography, endoscopy, pH probes, and esophageal manometry. Definitive diagnosis of CES and its types is possible through histopathology. In CES, contrast studies typically demonstrate segmental, concentric, short, and smooth narrowing of the esophagus with variable dilation of the proximal part [4].

The impassable nature of the stenosis confirms the stricture and rules out achalasia. The absence of signs of esophagitis excludes causes such as peptic stricture or stenosis by caustic ingestion. pH monitoring and esophageal manometry can differentiate stenosis

from peptic strictures. Endoscopic ultrasound (EUS) can be used to differentiate fibromuscular stenosis from tracheobronchial remnants stenosis [1]. Histopathology confirms the definitive diagnosis, revealing circumferential proliferation of smooth muscle fibers and fibromuscular thickening. The histological type is not always known preoperatively, hence the authors' uncertainty about whether to propose dilatation or surgery [5].

Surgery and dilatation are treatments for CES. The choice depends on the pathologic type and the severity of the stenosis. Surgery is recommended if dilations remain ineffective [6]. Surgical treatment involves myotomy and myectomy. Myectomy was performed for our patient. Thoracoscopic procedures have been reported, offering the benefits of minimally invasive surgery [7]. In our case, the impassable nature of the stenosis and its distal location supported primary surgical management rather than repeated dilations.

4. CONCLUSION

In infants presenting with recurrent vomiting, dysphagia, or respiratory distress, early consideration of congenital esophageal stenosis is crucial for timely and accurate diagnosis. Contrast studies of the upper gastrointestinal tract alongside endoscopy aid in establishing a differential diagnosis. This case highlights the role of circular myectomy as an effective primary treatment for fibromuscular CES when endoscopic dilation is not feasible. Minimally invasive surgery is currently recommended.

Consent: Written informed consent was obtained from the parents.

Competing interests: The authors declare that they have no competing interest.

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