Case Report

Double Trouble: A Rare Case of Dual Esophageal Stenosis in a 4-month-old

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Abstract

Esophageal stenosis is an uncommon clinical condition defined as a fixed narrowing of the esophagus. It can be congenital or acquired. The incidence of congenital esophageal stenosis (CES) is 1 in 25,000–50,000 live births. Most of these patients present in early infancy and many of them require surgical intervention. We report a very interesting case of a 4-month-old child with esophageal stenosis involving two different segments of the esophagus who was successfully treated surgically. This is one of the rarest variants of CES which involves two different segments of the esophagus.

Keywords: *Child, congenital esophageal stenosis, two segments*

Introduction

Esophageal stenosis is an uncommon clinical condition defined as a fixed narrowing of the esophagus. Congenial esophageal stenosis (CES) occurs in 1 in 25000 – 50000 live births. It comprises 10 - 15 % of all cases of esophageal stenosis.[1,2] The common types of congenital esophageal stenosis (CES) are (i) esophageal webs and (ii) esophageal stenosis due to bronchial remnants or congenital muscular hypertrophy. Most of these children present in early infancy and often require surgical intervention. Delayed diagnosis may lead to problems in swallowing, failure to thrive and complications such as recurrent cough, aspiration pneumonia, and malnutrition. CES may involve any part of the esophagus and is mostly isolated. We report an interesting case of a 4-month-old boy who had CES involving two different segments of the esophagus and required surgery for resolution of his symptoms.

Case Report

A 4-month-old boy born by normal delivery to nonconsanguineous parents presented with a history of difficulty in breast feeding since the age of 20 days. This was accompanied with episodes of recurrent cough. The antenatal history and scans were normal. The child also had two episodes of aspiration pneumonitis requiring hospitalization in the past 2 months. His weight was below the third percentile. In view of his symptoms, a contrast swallow was done which showed two areas of esophageal stenosis involving the upper esophagus and mid esophagus which were about 2 cm apart. The proximal esophagus was dilated [Figure 1]. The stomach was normal and there was no gastroesophageal reflux noted during the study. An upper gastrointestinal (GI) endoscopy was performed under general anesthesia. The first area of stenosis was at 10 cm from the oral cavity. The esophageal mucosa above the stenotic area was normal. The esophageal opening was just wide enough to accommodate a 0.89 mm sized flexible guide wire. The guide wire could not be blindly negotiated through the distal stenotic area. Esophageal dilatation was deemed risky and hence abandoned. Surgical excision of the strictures and esophageal anastomosis was planned. A right posterolateral thoracotomy was performed, and the esophagus dissected. There were two stenotic areas were 2.5 cm apart. Both the stenotic segments and normal esophagus in between was excised (total esophageal length excised 3 cm). The upper and lower esophageal ends were mobilized and end to end anastomosis done over an 8 F nasogastric tube using 5-0

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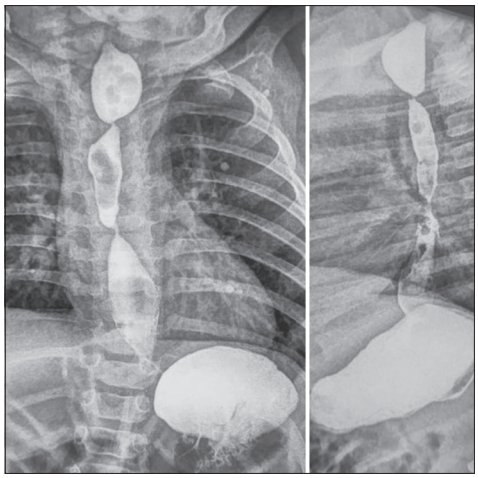

Figure 1: Oral contrast study showing two segments of esophageal stenosis in the upper and mid esophagus

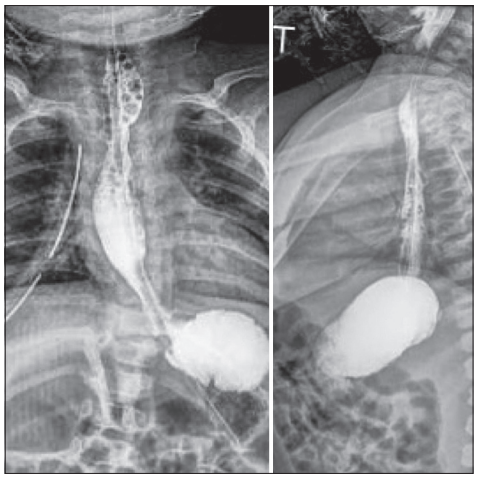

Figure 2: Postoperative oral contrast study showing smooth passage of the contrast through the reconstructed esophagus

monofilament polydioxanone sutures. The postoperative course was uneventful, and the child was commenced on oral feeds from the 6th postoperative day after a normal contrast swallow [Figure 2]. Histopathology of the stenotic segments showed thickened muscular and submucosal layers with diffuse fibrosis. The child is asymptomatic and thriving 4 months after surgery. In view of the esophageal mobilization, the child has been placed on oral lanzoprazole which we plan to continue for 6 months after surgery. The parents have also been advised to nurse him in an antireflux position. A check esophagoscopy done 2 months after the surgery showed a well-healed anastomosis, normal lower esophagus, and stomach. We plan to do a check upper GI contrast study to document presence or absence of gastroesophageal reflux before stopping oral lansoprazole.

Discussion

The first case of CES associated with esophageal atresia was reported in 1958. Only a few cases of multiple CES have been reported in world literature.[3,4] In 2001, Ramesh *et al*. suggested a new classification based on the type of stenosis and the association of esophageal stenosis with other anomalies of foregut separation.[5] By this classification, multiple stenoses were included as the rarest type of CES as was our case.

Presentation of symptoms depends on the severity of the stenosis. Infants with CES usually tolerate breast feeding and present with dysphagia or vomiting when weaning is commenced. Delayed diagnosis may lead to complications such as chronic cough, respiratory distress, aspiration pneumonia, and malnutrition. An oral contrast study shows a concentric, segmental short and smooth narrowing of the esophagus with dilatation of the proximal part. Mild varieties of CES can be missed or misinterpreted as transient spasm, dysmotility, or secondary narrowing due to gastroesophageal reflux and additional diagnostic work up like a computed tomography scan or an upper GI scopy may be required to confirm the clinical suspicion.

CES is divided into three histopathological types: (1) esophageal membrane (EM) which commonly involves the upper or middle third of the esophagus, (2) fibromuscular stenosis (FMS) which involves the middle or lower third, and (3) tracheobronchial remnants (TBR) which involve mainly the lower third of the esophagus within 1 cm of the gastroesophageal junction.[6] Histologically, EM has a normal squamous epithelium and muscular layer. FMS shows segmental hypertrophy of the muscular and submucosal layers with diffuse fibrosis whereas TBR may show cartilage, respiratory epithelium, or seromucous glands.

Both endoscopic dilatation and surgery can be offered for treatment of CES. However, the success of the chosen treatment depends on the severity and the histopathological features of the stenosis. Children with EM are successfully treated with dilatation. Michaud *et al*.[7] reported 38% success rates in patients of EM treated with endoscopic dilatations only. In patients of TBR endoscopic dilatation may be ineffective and resection of the stenotic esophageal segment and end-to-end anastomosis is the treatment of choice. Surgery for stenosis involving the lower esophagus may also require an additional fundoplication to prevent gastroesophageal reflux in the future. In the present case, we attempted an upper GI endoscopy but dilatation was risky as the guide wire could not be negotiated beyond the second area of stenosis which was almost >2 cm distal to the first area. Surgery was hence the only option in our case.

Conclusion

The diagnosis of CES is commonly delayed because of the rarity of the disease. The possibility of CES should be kept in mind when an infant suffers from repeated vomiting, dysphagia, respiratory distress, or failure to thrive. In such cases, a very low threshold should be kept for doing an oral contrast study as it is invariably helpful in arriving at a diagnosis. Nowadays, the treatment of CES mostly depends on the experience and personal opinion of the treating specialist. Individual approach could be applied in some exceptions, but standard protocol for early diagnosing and treatment of CES could improve the patient's condition after the treatment. Multicenter systematic analysis of CES, from diagnosis to treatment, with detailed description of follow-up period, could help in standardization of protocol for the most successful treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Brzački V, Mladenović B, Jeremić L, Živanović D, Govedarović N, Dimić D, *et al.* Congenital esophageal stenosis: A rare malformation of the foregut. Nagoya J Med Sci 2019;81:535-47.
2. Vandenplas Y. Management of benign esophageal strictures in children. Pediatr Gastroenterol Hepatol Nutr 2017;20:211-5.
3. Takamizawa S, Tsugawa C, Mouri N, Satoh S, Kanegawa K, Nishijima E, *et al.* Congenital esophageal stenosis: Therapeutic strategy based on etiology. J Pediatr Surg 2002;37:197-201.
4. Yoo HJ, Kim WS, Cheon JE, Yoo SY, Park KW, Jung SE, *et al.* Congenital esophageal stenosis associated with esophageal atresia/tracheoesophageal fistula: Clinical and radiologic features. Pediatr Radiol 2010;40:1353-9.
5. Ramesh JC, Ramanujam TM, Jayaram G. Congenital esophageal stenosis: Report of three cases, literature review, and a proposed classification. Pediatr Surg Int 2001;17:188-92.
6. Romeo E, Foschia F, de Angelis P, Caldaro T, Federici di Abriola G, Gambitta R, *et al.* Endoscopic management of congenital esophageal stenosis. J Pediatr Surg 2011;46:838-41.
7. Michaud L, Coutenier F, Podevin G, Bonnard A, Becmeur F, Khen-Dunlop N, *et al.* Characteristics and management of congenital esophageal stenosis: Findings from a multicenter study. Orphanet J Rare Dis 2013;8:186.