Journal of Neonatal Surgery 2018; 7:37
DOI: 10.21699/jns.v7i3.764

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CASE REPORT

Congenital Complete Esophageal Diaphragm: A Rare Variant of Esophageal Stenosis

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How to cite: Nahla K, Farhani R, Hmida B, Lamiri R, Ezzi A, Mosbehi S, *et al.* Congenital complete esophageal diaphragm: a rare variant of esophageal stenosis. J Neonatal Surg. 2018;7:37.

ABSTRACT

Congenital esophageal web is a rare disorder that presents a diagnostic and management challenge. In a female infant born at 31 weeks of gestation, significant secretions and respiratory distress were noted at birth. Chest X-ray demonstrated the nasogastric tube in the esogastric junction with no distal bowel gas. Esophagogram showed a congenital web near the esogastric junction. An endoscopic examination under general anesthesia showed a complete, thick membrane on the distal esophageal lumen. Endoscopic incision and cauterization of the web through the midline were performed, improving the clinical symptoms and esophageal stenosis.

Key words: Balloon dilatation; Child; Congenital esophageal web; Electrocauterization

INTRODUCTION

Congenital esophageal stenosis (CES) is a rare anomaly, with an incidence of about one per 25,000–50,000 live births [1]. It is characterized by intrinsic narrowing of the esophagus secondary to congenital malformation of the esophageal wall architecture [1]. CES is divided into three types: Tracheobronchial remnants (TBR), fibromuscular thickening (FMT), and membranous webbing (MW) [1,3]. Congenital esophageal web is a rare disorder that corresponds to a thin diaphragm of tissue usually forming a concentric lumen in the lower esophageal lumen reported only sporadically [2-4]. It presents a diagnostic and management challenge.

CASE REPORT

A female infant was born at 31 weeks of gestation through cesarean delivery after pregnancy complicated by polyhydramnios. Body weight at birth was 1900 g. Apgar scores were 3, 6, and 9 at 1, 5, and 10 min, respectively. Significant secretions and respiratory distress were noted at birth. An attempt was made to pass a nasogastric tube. Chest X-ray demonstrated the nasogastric tube in the esogastric junction without distal bowel gas (Figure 1).

Gross Type A atresia was suspected, and gastrostomy was performed on the 2nd day of life.

A barium esophagogram through the gastrostomy and the esophageal tube revealed an intact, circumferential web at the distal esophagus with dilation of the proximal esophagus (Figure 2a and b).

Under general anesthesia, the patient was placed in the supine position. The endoscopic examination was performed with a pediatric gastrointestinal fiberscope. At a distance of 23 cm from the mouth, the distal esophageal lumen was obstructed by a thick, non-transparent, and complete membrane.

Endoscopic incision and cauterization of the web through the midline were performed using diathermy on three different portions. No significant bleeding occurred. An esophageal balloon dilator was placed through a nostril into the esophagus, and a subsequent dilatation with a balloon dilator was implemented slowly. The procedure was completed by

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Submitted: 02-05-2018 Accepted: 08-05-2018
Conflict of interest: None Source of Support: Nil

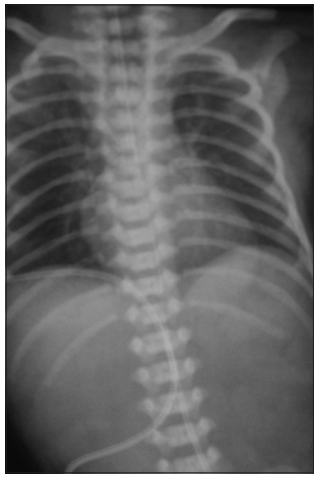

Figure 1: G-tube in the esogastric junction, no distal bowel gas.

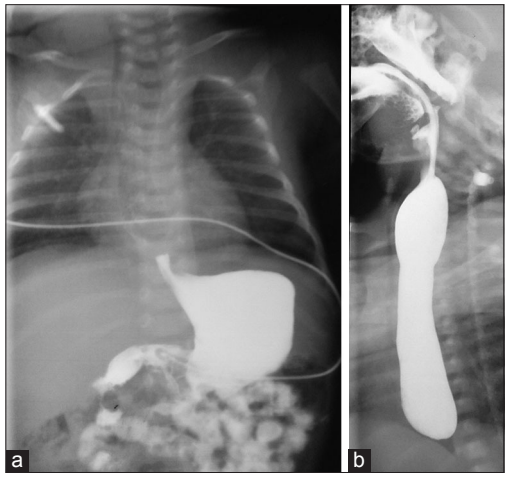

Figure 2: (a) Esophagogram showing a congenital web near the esogastric junction. (b) The proximal esophagus is dilated.

the insertion of a gastric tube, which remained for 10 days.

The patient did well. He had no difficulty in swallowing liquid food. The barium swallow was normal, and he had an ideal weight gain at the 20-month follow-up.

DISCUSSION

CES is a rare malformation which is observed with a relatively high incidence, as are other intestinal obstructions. Using available data, the overall frequencies of FMT, TBR, and MW were 53.8%, 29.9%, and 16.2%, respectively [3].

The congenital membranous diaphragm is exceedingly rare; it is believed to be produced by incomplete reformation of the esophageal lumen on recanalization of the esophagus observed in the 6–8th week of gestation. It is a thin diaphragm of tissue that could be complete or incomplete.

The diagnostic approach should be started with a barium swallow. The esophagoscopy is considered the most reliable diagnostic examination for an esophageal web [5].

Appropriate therapeutic strategies, including endoscopic treatment and surgical repair, still remain controversial. Resection of the membrane at thoracotomy is usually the only treatment available for a complete esophageal membrane [6-8]. The development of gastrointestinal fiberscopes suitable for infants and children enabled a second way of treatment, which was performed in our patient: The membrane was incised endoscopically. Electrocauterization involves burning or destroying tissue using a small probe with an electric current running through it. It has become a routine surgical means of burning unwanted or harmful tissue and is used to control hemorrhage [6]. Successful electrocauterization of an esophageal web had been reported in many studies. The performance of therapeutic modality is justified to avoid intervention by thoracic surgery.

The indication for this endoscopic treatment, however, should be restricted to patients with clear anatomic situation. It should be emphasized, however, that optimal technical equipment and skilled endoscopic technique are essential; otherwise, the risk of complications such as perforation and mediastinitis is too high. For incomplete membranes, esophageal dilatation or endoscopic incision of the membrane using cautery or a laser is possible [9]. Dilatation of the web with various instruments has been reported [10].

CONCLUSION

Congenital esophageal web is a rare condition in the pediatric population that is difficult to definitively diagnose. Although there is no consensus on how to manage these lesions, endoscopic treatment should be considered the first line of treatment. Nevertheless, its use should be weighed carefully against alternatives to minimize patient discomfort and adverse events.

Author's contribution

All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

Consent statement

Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that

every effort will be made to conceal the identity of the patient however it cannot be guaranteed.

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