Case Report of Severe CES Cured by Replacement of Esophagus with Gastric Conduit

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Abstract

Congenital esophageal stenosis (CES) is a rare clinical entity that is often not diagnosed until months or sometimes many years after birth. Delayed treatment may lead to poor prognosis. Here we report one surgical procedure for curing delayed severe CES. The esophageal stenosis was resected and the gastric conduit was used to replace esophagus. The type of operation completely solved the patient’s dysphagia and acquired preferably effect.

Keywords: Congenital esophageal stenosis; Surgical treatment; Gastric conduit

Introduction

Congenital esophageal stenosis (CES) is defined as an intrinsic stenosis of the esophagus, which is caused by congenital malformations of the esophageal wall architecture that are present at birth, although it is not necessarily symptomatic in neonates. CES is a rare malformation with an incidence of approximately 1 in 25,000 to 50,000 live births [1] and can be classified based on the histologic type of the stenosis: 1) ectopic tracheobronchial remnants (TBR), 2) Fibromuscular thickening (FM), and 3) membranous diaphragm (MD) [2].

Herein, we present one case of severe CES cured by replacement of esophagus with gastric conduit.

Case Presentation

A three-year-old girl was hospitalized in our department for severe dysphagia, even water. The girl had been having episodes of vomiting discontinuously when eating since her birth. Over the next three years, she was noted to have poor weight gain and was hospitalized in other hospitals several times for full workup, but no diagnosis had been made. She continued to vomit four to six times per day. The other symptoms included solid food refusal, regurgitation and dysphagia. Dysphagia was progressively aggravated. The child’s weight was only 9 kg when she was hospitalized in our department 3 years ago. Esophagogram and CT scan showed marked narrowing of the middle thoracic esophagus, a bird’s-beak configuration of visualized contrast material, and dilation of the proximal esophagus (Figure A and B). Esophagoscopy confirmed the narrowing through which the gastroscope could not pass. The patient underwent open esophagectomy through three incisions (right chest, upper abdomen and left neck). First, the esophagus, including the narrowing was circumferentially mobilized from the esophageal hiatus up to the thoracic inlet during the thoracic operation. Secondly, the stomach was mobilized by dividing most ligaments and blood vessels around it, preserving the right gastroepiploic vessels during the abdominal operation. The gastric conduit (Figure C) was formed with multiple applications of the linear stapler along the lesser curvature from the right gastric vessels to gastric fundus, just as indicated in our pre-description [3]. Finally, the end-to-side anastomosis of the remaining esophagus and the gastric conduit was completed during the cervical operation. The patient was treated with broad-spectrum intravenous antibiotics and monitored in the pediatric intensive care unit three days. A repeated esophagogram showed resolution of the high-grade narrowing (Figure D and E). The postoperative pathology is FM of CES (Figure F and G). She had a gradual transition from liquid to normal diet after operation and has gained 12 kg for three years.
Discussion

The treatment of CES depends on pathological diagnosis. For example, cases of tracheobronchial remnants require surgical removal and reanastomosis [4]. A few cases of a membranous diaphragm may be successfully treated by balloon dilatation [5]. Fibromuscular hyperplasia of CES has been treated successfully by dilatation alone [6].

However, that causing severe dysphagia even water like this case, in spite of the pathological diagnosis, necessitates surgical removal and reanastomosis. The replacement organs may be stomach, jejunum or colon. The stomach may be unmodified or formed as gastric conduit before anastomosis. According to the primary stomach, the gastric conduit has the following benefits, such as less gastric juice retention, longer gastric conduit, better blood supply to gastric conduit, anastomosis lower on richer blood supply site, easier clearance of lymph nodes around the stomach, less dyspnea (no bulky stomach in chest), easier to pull up (pass hiatus/thoracic inlet).

For our case, the postoperative pathology is FM of CES. If the diagnosis of CES could be made in early stage, the patient may be cured by dilatation alone. While when the patient was hospitalized in our department, she couldn’t drink anything, even water. According to the condition of patients, the esophageal stenosis was resected and the gastric conduit was used to replace esophagus. The type of operation completely solved the patient’s dysphagia and acquired preferably effect.

Therefore, we declare that severe CES necessitates surgical removal and reanastomosis. The surgical procedure used in the case, esophagectomy with three incisions, is suitable for the treatment of CES with severe dysphagia.

Supportive Foundations: The Fundamental Research Funds for the Central University of China (Grant No.08143004), the Funds for Science and Technology Project of Shaanxi Province of China [grant No. 2014K11-02-03-07].

References