Case Report

Balloon dilation of epidermolysis bullosa-related esophageal strictures:
A report of two cases


A B S T R A C T

Epidermolysis bullosa (EB) is a very rare inherited disease featured with skin blistering resulting from minor trauma. Sometimes the esophageal mucosa could also be involved, which leads to esophageal strictures. Here we report two cases of EB-related esophageal strictures who were successfully treated with esophageal balloon dilations. The two cases with EB had severe dysphagia. Clinical examination showed signs of malnutrition, skin blisters and loss of toenails due to EB. They underwent careful fluoroscopic balloon dilation with 10- and 16-mm-sized balloon catheters, respectively. They could ingest soft and some solid foods after the procedure and maintained during the 20 months and 16 months follow-up periods.

Keywords: Balloon dilation; Epidermolysis bullosa; Esophageal stenosis; Fluoroscopy

Introduction

Epidermolysis bullosa (EB) is a rare inherited genetic disorder which is featured with recurrent mucotaneous blister formation caused by minor trauma. Sometimes, the esophagus could be involved and results in esophageal stricture, with its upper part being the most commonly affected. Management of the esophageal stricture usually starts from modification of diet texture to soft, puree and liquids. However, the stricture could get severe within years and requires treatment. In this study, we report two cases of EB-related esophageal strictures that were successfully treated with balloon dilation.

Case Report

Case 1

A 52-year-old man presented with dysphagia for over 30 years and complained of symptom worsening for 5 years. He started experiencing dysphagia after swallowing a piece of hot fish when he was 22 years old and he mostly consumed liquids during the recent 5 years. He had been diagnosed with dystrophic EB 14 years ago. On admission, he could not take any food and thus the dysphagia score was 4 according to the CIRSE guidelines. Although he was diagnosed with an upper esophageal stricture by esophagography two years ago in another hospital, balloon dilation had not been performed either under fluoroscopy or endoscopy before our admission. On physical examination, his body mass index was 16.1 kg/m² and he had blisters on the skin, loss of fingernails and toenails, and deformity of hand joints (Fig. 1). Prior to balloon dilation, an esophagography was performed and revealed two severe strictures at the upper esophagus (Fig. 2A). Balloon dilation was carried out in this patient by an interventional radiologist (Z.Q.Y.) with 18 years of relevant clinical experience. Midazolam (5 mg; Nhwa Pharma, Xuzhou, China) was given by intramuscular injection for pre-procedural sedation. Lidocaine hydrochloride (10 mL; Jumpcan Pharma, Taixin, China) was sprayed on the pharynx for topical aesthesia. Under fluoroscopic guidance, a 0.035-inch stiff guidewire (Terumo, Tokyo, Japan) was inserted through the patient’s mouth and then manipulated to cross the strictures. A 10-mm-sized balloon (Atlas, Bard, AZ, USA) was placed across the strictures over the guidewire and slowly inflated with contrast medium (Omnipaque 300; GE Shanghai, Shanghai, China) (Fig. 2B). The contrast pressure was sustained for 30 seconds before deflation (Fig. 2C). The procedure was completed with the obvious improvement of the

Department of Radiology, The First Affiliated Hospital, Nanjing Medical University, Nanjing, China
Received December 21, 2017; Revised February 2, 2018; Accepted February 2, 2018
* Corresponding author. Department of Radiology, The First Affiliated Hospital, Nanjing Medical University, 300 Guangzhou Road, Nanjing 210029, China.
E-mail address: yangzhengqiang@jsph.org.cn (Z.-Q. Yang). ORCID: https://orcid.org/0000-0001-9103-3534

© This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0) which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original work is properly cited.
esophageal stricture and without complications on esophagography (Fig. 2D). After the procedure, the patient could have soft and some solid foods (dysphagia score = 1). The patient was followed up by phone call and outpatient clinic interview. His dysphagia score remained the same during the 20-month follow-up period.

Case 2

A 27-year-old man was admitted to our hospital due to progressive dysphagia for three years. He complained of inability to swallow and the sensation of “food getting stuck in his throat”. On admission, he could only take liquid foods (dysphagia score = 3). He had had extensive skin blisters since birth and was diagnosed as dystrophic EB by the dermatologist in our institution. Physical examination revealed that he was ill nourished. He had skin blisters involving his head, neck, and body (Fig. 3A). His fingers were normal but many of his toenails and his umbilicus were missing (Fig. 3B, 3C). An esophagography was done after admission and showed a tight cervical esophageal stricture (Fig. 4A). This patient underwent careful fluoroscopic balloon dilation. A 16-mm-sized balloon catheter (Atlas) was used and fully dilated during the procedure (Fig. 4B–4D). He recovered uneventfully after the procedure and gradually started on soft and some solid foods (dysphagia score = 1). During a 16-month follow-up period, the patient could still ingest the soft and some solid foods.

Discussion

As an autosomal dominant or an autosomal recessive disease, EB is very rare with a reported prevalence of 8% to 49% per million in western countries. According to the level of cleavage in the skin layer, EB is classified into four major types; EB simplex, junctional EB, dystrophic EB, and Kindler syndrome. EB is characterized by skin fragility and blister formation. Apart from skin, other parts of the body such as gastrointestinal tracts could also be affected. When esophagus is involved, it can cause esophageal stricture, as the mechanical trauma to esophageal mucosa by solid or hot alimentation would lead to bulla formation, ulceration, and subsequent fibrosis of the esophagus. The upper esophagus is the most common part to be involved, since the diameter of upper part is smaller than the lower part.

The EB-related esophageal stricture could lead to a series of problems, such as nutritional failure, food bolus obstruction, esophagitis, and aspiration pneumonia. The current treatments
of the esophageal stricture include diet modification, conservative medical therapy, balloon dilation, and colonic interposition or transposition. The diet change to soft, puree and liquids added with multivitamins and minerals may decrease the injury to the esophagus; however, the caloric intake usually cannot meet the normal demand, which can result in malnutrition and growth retardation. The medical therapy with proton pump inhibitors is reported to be little effective, especially in tight weblike strictures. Because of the high mortality and morbidity rates, surgical repair is not the first line therapy unless the patients cannot be successfully treated with conservative and interventional therapies. Therefore, balloon dilation becomes the mainstay of treatment.

Previously, bougienage was also used for esophageal dilation. However, the tangential shearing forces during bougienage dilation can cause the detachment of the esophageal mucus membrane and may create a new esophageal stricture. Balloon dilation is considered to be safer than bougienage dilation, because it only produces longitudinal forces to the esophageal wall. The balloon dilation could be performed endoscopically or fluoroscopically. When comparing the two means, it seems that fluoroscopic guidance owns more advantages. Under fluoroscopic guidance, the fine hydrophilic guidewires and catheters not only can pass through very severe strictures that the endoscope could not, but also they make less damage to the esophageal wall owing to their small size and smoothness.

In this study, the two patients underwent successful balloon dilation under fluoroscopic guidance without severe complications such as massive esophageal bleeding or perforation. The improvement of dysphagia in both of them after the procedures showed the effectiveness of the treatment. The key point of balloon dilation is the selection of balloon size. As the cases reported in the literature was very limited, there is no consensus on it. However, sequential dilations starting from a reasonably small balloon was recommended in order to minimize the risk of esophageal perforation. In one study, the authors treated 19 patients with dystrophic EB and suggested that the commonly used balloon size ranged from 12 to 18 mm. In our study, 10- and 16-mm balloon catheters were chosen in the two patients respectively depending on the severity of the strictures. During the balloon dilation procedure, the blood staining on the balloon surface usually indicates the mechanical distention of the stricture and the procedure could be finished. In some patients, multiple sessions are required due to the recurrence of dysphagia. Although the reported recurrence rate after the initial balloon dilation was above 75% from the lim-
ited previous studies,12,13 around 30% patients could get a one or two years long dilation-free interval after multiple dilations.

In conclusion, fluoroscopic balloon dilation was generally safe and effective in the treatment with EB-related esophageal strictures. It should be considered the first line method to relieve the patient’s dysphagia owning to its mini-invasive advantage.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

References

Our Goals:

- **Multi-disciplinary Collaboration to promote world-wide Expertise**
  Establish a comprehensive GI intervention network among endoscopists, interventional radiologists and gastrointestinal surgeons for multidisciplinary collaboration and interaction

- **Sharing and advancing technological Innovations**
  Inform, promote and globalize the many outstanding technological innovations of each of the specialties

- **Foster future Specialists**
  Aid young brilliant doctors to make an early debut on the international stage through SGI

- **Become a Role Model**
  Showcasing the benefits of multi-disciplinary collaboration in science, education and clinical practice