Case Report

Esophageal diverticulum and high-grade esophageal stricture responsive to serial balloon dilation and nutritional therapy

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ABSTRACT

Aims: 16 year old boy with recessive dystrophic epidermolysis bullosa and previous history of esophageal strictures presents with progressive dysphagia and hematemesis. Patient was admitted to hospital to determine cause of hematemesis. Methods: Patient underwent endoscopy using Olympus GIF-XP 160 endoscope. Results: Endoscopy revealed active esophagitis and multiple blood clots. A high-grade esophageal stricture was noted at level 35cm from incisors, in which the endoscope (5.9 mm diameter insertion tube) could not pass. Endoscopy two days later revealed an esophageal diverticulum and bifurcation located 35cm from the incisors. Instilling a small amount of water revealed a blind ended sac on the left side and a lumen leading to the stomach on the right side (see image). The true esophageal lumen was dilated using a 6-7-8mm CRE™ Wire guided Balloon Dilator (Boston Scientific). He was treated with nutritional therapy (TPN, then enteral feedings following gastrostomy placement by interventional radiology), sucalafate, ranitidine, and zinc. Three subsequent endoscopic dilations increased the true lumen diameter to 15 mm. Follow-up esophagram revealed a notably smaller esophageal diverticulum and a small amount of mediastinal air, suggesting microscopic esophageal perforation treated conservatively with IV antibiotics. Conclusion: EGD three months later showed no residual esophageal stricture and complete resolution of the esophageal diverticulum. Balloon dilation along with management of nutrition have been shown to be effective and safe methods of treating esophageal strictures in children, and in this case has been effective in managing an esophageal diverticulum.

Key words: Eidermolysisbullosa, Esophageal stricture, Esophageal diverticulum

INTRODUCTION

The patient is a 16 year old boy with past medical history significant for recessive dystrophic epidermolysis bullosa, esophageal strictures, and esophagitis presenting with one day of dysphagia and hematemesis. Epidermolysis bullosa is a group of disorders caused by a genetic mutation in the genes coding for structural proteins of the skin. Characteristics of epidermolysis bullosa include: fragile skin, profound anemia, failure to thrive and airway compromise. Epidermolysis bullosa has been reported to be associated with anatomical lesions of the esophagus such as esophageal webs. He was in his usual state of health one day prior to admission, when he had difficulty swallowing food. One hour later he developed esophageal spasms and began having multiple episodes of bloody emesis, prompting his family to bring him into the emergency room. He was hemodynamically stable, and gastroenterology was consulted. He was placed on intravenous fluids with intravenous ranitidine and admitted to the intensive care unit to undergo endoscopy the following morning. He remained stable overnight.

METHODS

The patient underwent serial endoscopies using Olympus GIF-XP 160 endoscope, to determine cause of hematemesis and dysphagia.
RESULTS

On day one of admission, endoscopy revealed esophagitis, erythema, and friability of the mucosa of the esophagus. At levels twenty-five and twenty-seven centimeters there were blood clots attached to the esophageal lumen. A tight esophageal stricture was noted at level thirty-five, which the endoscope could not bypass. Active bleeding was found at the time, and dilation was held off secondary to presence of marked esophagitis. Patient tolerated procedure well and started on Carafate four times daily to alleviate the discomfort from the noted esophagitis. Overnight the patient continued to have epigastric pain. He underwent second endoscopy on day three of admission. Endoscopy revealed improved esophagitis and no further blood clots. A bifurcation appeared at level thirty-five, which was not present on the previous day’s endoscopy. A small amount of water was instilled revealing a blind ended sac on the right and a tract passing into the stomach on the left. A balloon dilator was inserted and expanded to six millimeters and then to seven millimeters. At eight millimeters, significant resistance was felt, and dilator removed. Upon removal, the true tract leading to the stomach was more dilated than the false tract. Patient was continued on Carafate, zantac, and zinc, and by day four of admission his pain was significantly better. He underwent third endoscopy on fifth day of admission. Esophagitis was improved. The bifurcation was still present with each lumen appearing the same diameter. Balloon dilator was inserted and dilated to eight millimeters, nine millimeters, and ten millimeters, with the ten millimeter dilation held for two minutes. The next day patient reported no further pain. The fourth endoscopy was performed on seventh day of admission. The true lumen appeared slightly wider than the false tract. Balloon dilator was inserted and inflated to ten millimeters and then to twelve millimeters and held for thirty seconds. Some pressure was felt and there was a small amount of bleeding at dilation site.

On hospital day ten, he had his next endoscopy. Balloon dilator was inserted and dilated from twelve millimeters to thirteen and then to fifteen millimeters and held for one minute. Esophagram was performed that same day revealing, the notable esophageal diverticulum and also a small amount of mediastinal air, suggesting microscopic esophageal perforation. Patient had thus far been asymptomatic without any history of fever, pain, or tachypnea. He was advanced to clear liquid diet by day eleven of admission and advanced to full liquids by day thirteen of admission which he tolerated well. Gastrostomy tube was placed on admission day fourteen secondary to patient’s malnutrition. In addition to receiving gastrostomy tube feeds, he eventually tolerated soft diet prior to his discharge on day eighteen. He was seen in clinic eight days after discharge without any dysphagia, hematemesis, and tolerating his soft diet well.

DISCUSSION

Epidermolysis Bullosa is a rare disorder involving blistering at sites caused by minimal trauma. In some forms of Epidermolysis Bullosa there are also gastrointestinal manifestations. A retrospective study looked at 35 patients with EB who were evaluated consecutively revealing 30 of those patients had some type of upper GI complaint with painful swallowing. Twenty of those thirty-five patients had Recessive dystrophic EB like our patient and all of them had an upper GI complaint. Twenty one of those patients had dysphagia so they underwent barium swallow revealing 62% of them with strictures. Endoscopy was performed revealing esophageal strictures and one patient with esophageal web. None of these patients were reported to have findings of a esophageal diverticulum such as our patient did. There is no cure for epidermolysisbullosa, but management in symptoms. There have been different ways of management in children and adults with esophageal strictures involving dilation, nutrition, or medication. In a study involving 9 children with dysphagia and sensation of food stuck in their esophagus, many of them had poor solid food intake, and a barium swallow revealed esophageal stricture. These children underwent intensive nutritional therapy of 120% Recommended dietary allowance and underwent dilation under fluoroscopy with seven of those children having successful dilations without requiring repeat dilations. These children were able to tolerate solid foods without any serious complications. Another study involved 22 patients with Recessive dystrophic epidermolysisbullosa after endoscopic placement of guidewire and dilation under fluoroscopic assistance. Following the procedure, dysphagia was improved and all children were able to tolerate normal solid foods within six hours of the procedure. Complications were minimal and managed medically, and a child as young as 30 months old tolerated the fluoroscopic guided balloon dilation. In a study involving 49 patient with strictures, all children with EB related strictures had a successful endoscopic balloon dilation without any of them having complication of perforation compared to the other groups with strictures secondary to causes such as erosion or esophageal atresia who had had unsuccessful procedures as well as perforations. All these studies demonstrate the success of esophageal strictures via balloon dilation whether it be done fluoroscopically or endoscopically; however, a procedure like this does involve risks such as perforation, which in our case was managed medically without serious complication. Medical management of esophageal complications secondary to EB must be considered too. Oral budesonide has been studied in children with moderate to severe esophageal strictures secondary to EB. The children were prescribed four months of oral budesonide nebulizer to be administered twice a day. The follow up esophagram showed significant improvement of the strictures as well as increased food intake, which in fact would decrease number of dilations or other invasive treatments needed for these children. Nutrition is another important aspect involved in the care of all children with EB. Many children with EB have malnutrition because of decreased intake secondary to problems involving the esophagus as well as an increased demand of nutrients for wound healing and loss of nutrients via the skin. These patients are also at greater risk of infection and loss of protein by way of epithelial repair, requiring increased protein intake for both that could be upwards of 200% RDA. Our
patient underwent nutritional therapy involving insertion of a gastrostomy tube which would help with overall growth and repair of wounds. Patients with EB have low levels of zinc, which our patient received while hospitalized. Esophageal strictures have been shown to be corrected by way of medical management, balloon dilation whether endoscopically or fluoroscopically and with nutritional optimization revealing good results and increased quality of life allowing the patients to tolerate eating. Although none of these studies involved esophageal diverticum, our patient benefitted from similar approach with great success and improvement of daily life.

Figure 1: Above: Bifurcation seen at 35 cm from the incisors. Left side is a blind-end sac, Right side true lumen

Figure 2: After third balloon dilation up to 15 mm

Figure 3: Three month follow up revealing no esophageal stricture or diverticulum

REFERENCES