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CASE REPORT

Multiple Congenital Esophageal Stenosis: In a 14-Month-Old Girl

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Abstract

We aimed to share our very rare case with multiple congenital esophageal stenosis and its treatment. A 14-month-old girl presented with dysphagia, vomiting, and growth retardation, two esophageal strictures were detected in the esophagogram and esophagoscopic examination. While the proximal stenosis responded to balloon dilatation, the distal stenosis did not respond to dilatation and was perforated. Distal stenosis was treated with resection and anastomosis. Cases presenting with dysphagia and chronic vomiting should always be evaluated with esophagogram and endoscopy. Balloon dilatation should be the first choice in cases with stenosis. In cases that do not respond to dilatation, resection and anastomosis should be performed without insisting on dilatation.

Keywords: Congenital; esophageal stenosis; multiple.

Congenital esophageal stenosis is an intrinsic stricture of the esophagus present at birth. It occurs approximately once in 25000–50000 live births. It is very rare to have more than one congenital esophageal stenosis in one case. The exact cause is still unknown. It is usually seen in the middle or one-third lower part of the esophagus. One-third of the cases may associate with other anomalies. It is classified histologically in three main groups: fibromuscular thickening, tracheobronchial remnant, and membranous web. Fibromuscular thickening is the most common and the least membranous diaphragm type is seen[1-3].

Patients usually present with difficulty in swallowing, vomiting, growth failure and development retardation. Symptoms mostly begin to appear during transition to solid food. Diagnosis is often difficult as these symptoms may suggest many diseases such as gastroesophageal reflux, candidiasis, achalasia, and eosinophilic esophagitis. However, it can be easily visualized with an esophagogram[1,2,4].

In our study, we shared the case we treated due to the rare multiple congenital esophageal stenosis.

Case Report

A 14-month-old female patient was admitted to the pediatric gastroenterology outpatient clinic with complaints of vomiting and malnutrition for 3 months. It was learned from the mother that she could tolerate breast milk and vomiting is especially after solid foods. Vomiting has intensified for the past 5 days and has started to have difficulty in taking breast milk. The patient was under the 3rd per-

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centile with a weight of 7 kg and of 71 cm. There was no other pathology detected in the examination. She was admitted to the pediatric gastroenterology service due to nutritional difficulties, growth failure, and developmental delay. There were no abnormal findings in the patient's blood test. In the endoscopy performed on the patient, double esophageal stenoses were observed (15^{th} and 18^{th} cm from the mouth). The procedure was terminated by inserting an 8 Fr nasogastric tube (NGT) for feeding the patient. After endoscopy, the patient was transferred to the pediatric surgery service considering congenital esophageal stenosis in the council, which was performed with pediatric gastroenterology, pediatric surgery, and general pediatrics. In esophagoscopy performed by pediatric surgery (Fujifilm Co, Tokyo, Japan, distal end diameter: 5.8 mm), proximal stenosis could not be overcome. Balloon dilatation decision was made for the patient who had double stenosis in the esophagogram. Gastroesophageal reflux and additional pathology were not detected in the gastric duodenum radiography performed before dilatation. Thereupon, the esophageal balloon dilator (Micro-Tech Co., Ltd. 6 mm, 3 atm) was inserted into the stomach, the balloon was inflated in the area of the stenosis, visualized with scopy and dilatation was performed (Fig. 1).

It was completely dilated after 2 min in proximal stenosis, but no visible enlargement was observed in the lower stenosis, although it was waited for 12 min.

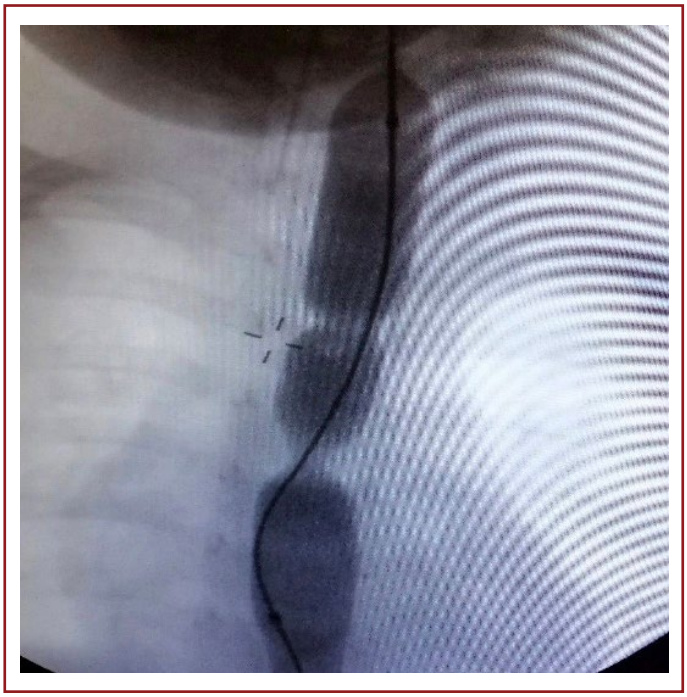

Figure 1. Esophageal strictures visualized by inflating the balloon with opaque material.

Thereupon, the endoscopy was repeated 2 weeks later. The endoscope passed through the proximal stenosis easily, minimal stenosis was detected, the endoscope did not pass through the distal stenosis, and stenosis continued. Balloon dilatation (Micro-Tech Co., Ltd. 6 mm, 3 atm) was applied to the patient again with fluoroscopy and endoscopy. There was no sandglass appearance in proximal stenosis. In the distal stenosis, balloon dilation was applied with a pressure of 3 atm for 10 min, but it was not dilated. On suspicion of perforation in the control endoscopy, it was observed that the contrast agent (omnipaq®) given through the NGT escaped into the thoracic cavity through the lower stenosis. The process was terminated by inserting NGT. Ten days later, the contrast-enhanced radiography showed that the perforation was healed, and the patient was started to take orally.

Resection anastomosis was planned, deciding that the stenosis might contain ectopic tissue, because the stenosis did not respond to dilatation and that recurrent dilatation would not be appropriate due to previous perforation.

One month after the perforation, thorocotomy was performed through the 5th intercostal space. On exploration, the esophagus and distal stenosis were determined by simultaneous esophagoscopy. An area of approximately 2 cm of lower esophageal stricture was resected and end-to-end anastomosis was performed. The patient, who was taken in the post-operative intensive care unit, was intubated for 14 days due to lung problems. On the post-operative 5^{th} day, anastomosis leakage was checked with contrast agent (omnipaq®). There was no leakage. On the 7^{th} post-operative day, the patient was started to feeding with NGT. The patient was extubated on the post-operative 14^{th} day and transferred to the pediatric surgery service on the 20^{th} post-operative day. The patient, who tolerated oral, was discharged on the 28^{th} post-operative day. No tracheobronchial remnant was found in the pathology and reported as fibromuscular thickening. In the esophagoscopy performed in the 2^{nd} post-operative month, a moderate stenosis was observed in the anastomosis line. Balloon dilatation was performed. At the post-operative 3rd month, proximal stenosis was normal in the endoscopy, and a moderate stenosis was detected in the resection area, which did not permit the passage of the 5.8 mm diameter endoscope. In the balloon (Micro-Tech Co., Ltd. 8 mm, 4 atm), dilatation performed with scopy the sandglass appearance completely improved in 1 min. No pathology was found in both stenoses in the endoscopy and esophagography performed 2 weeks later. There was not any pathology in old stenosis areas. However, there was an appearance at the esophagogastric junction that makes us achalasia think. There was not any pathology

in esophagoscopy. Moreover, the decision of follow-up was made because the patient had no complaints. The patient has no active complaints at 7 months postoperatively. Her weight gain is good and 9 kg.

Discussion

Since congenital esophageal stenosis is rare, related studies consist mostly of case reports and a small number of case series. Although it is defined as intrinsic stenosis resulting from congenital malformation of the esophageal wall structure, it cannot be easily diagnosed in the neonatal period. It is usually diagnosed after the 5th or 6th months in transition to supplementary food. Approximately in 33% of cases, esophageal atresia can be seen with many congenital anomalies, especially tracheoesophageal fistula. However, most patients have no problems other than congenital esophageal stricture[3,4]. As in our case, patients without additional anomalies are usually diagnosed late because they seem healthy. In our case, the family did not have any complaints until she was 11 months old, and she presented with persistent vomiting. There was growth and developmental delay due to vomiting.

It can be differentiated from other causes such as achalasia, esophagitis, and gastroesophageal reflux, which cause esophageal strictures, by the patient's history and imaging by esophagogram and esophagoscopy[4,5]. In our case, the diagnosis was made by esophagoscopy performed in terms of the etiology of vomiting. Two stenoses, one in the mid esophagus and the other in the distal esophagus, were observed in both esophagoscopy and esophagogram. In the series on congenital esophageal strictures, multiple esophageal strictures were found <10%[6,7]. In the study conducted by Lees et al.,[4] it was mentioned that a pediatric surgeon may encounter congenital esophageal stenosis only once or twice during his professional life. Accordingly, most pediatric surgeons never encounter with congenital esophageal strictures, which are multiple strictures in their professional life.

Studies have mentioned that dilatation will be sufficient in the treatment of congenital esophageal stricture, fibromuscular thickening, and membranous web type, but surgery in the type of tracheobronchial remnant will be required. In a study, in which tomography was performed in cases with congenital esophageal stenosis, esophageal stenosis was detected, while tracheobronchial remnant could not be evaluated in tomography. Esophagogram and endoscopic ultrasonography are used preoperatively to determine the histological type. However, no examination can fully distinguish the histological type. Therefore, it is not possible to determine the treatment according to the pathological type. The first treatment option is balloon dilatation. Perforation risk has been reported from 10% to 44% in balloon dilation[1,4,8,9]. Perforation usually occurs secondary to high pressure and prolonged dilatation. In our case, when there was no response to balloon dilatation (in the second dilatation), considering the risk of perforation, 4 atm pressure was applied with an 8 mm balloon dilatation catheter, waiting for 10 min and high pressure and long-term dilatation was applied. Considering the high risk of perforation, the patient was performed esophagoscopy and esophagogram after dilation. Thus, the perforation that occurred after the second dilatation had the chance to treat early diagnosis.

There have been authors who argue that 60–95% of cases respond to dilatation regardless of histological type. In cases that do not respond to dilatation, surgery is recommended especially after two dilations[8,9]. Surgical resection and end-to-end anastomosis were performed in our patient, because there was no response with two dilatations and there was perforation. The patient developed mild stenosis after surgery, oral intake was provided with dilatation and she started to gain weight.

In our study, which is rarely seen, we wanted to remind the case of multiple congenital esophageal stenosis and its diagnosis and treatment. Patients presenting with dysphagia and chronic vomiting should always be evaluated with esophagogram and endoscopy. Balloon dilatation should be the first choice in cases with stenosis. However, on patients who do not respond to dilatation should not be insistent. High pressure and prolonged dilatation should be avoided. Even If done, esophagoscopy and esophagogram should be performed after the dilatation procedure for the early diagnosis and treatment, considering the risk of perforation. Thus, complications of early diagnosis and treatment and late diagnosis and treatment are eliminated. It should be kept in mind that the success of the primary resection anastomosis with surgery will be high in these patients.

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