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An 18-Month-Old Child Suffering From Achalasia Cardia Successfully Treated With Pneumatic Dilation

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Abstract: Achalasia cardia is an idiopathic esophageal motility disorder. It is rare in children and infrequent in below school-going age groups. The "bird's beak" appearance of the lower esophagus on the esophagogram (barium swallow) is a classical radiological finding in the cases of esophageal Achalasia. The goals of achalasia therapy are symptom relief and improvement of esophageal emptying to prevent megaesophagus. The most effective treatment options are pneumatic dilation and surgical myotomy (Heller's myotomy). Pneumatic dilation is the initial treatment of choice and does not preclude myotomy. Here, we present our experience with a young child with achalasia cardia that was successfully treated with pneumatic dilation.

Key Words: achalasia cardia, dysphagia, motility disorder, pneumatic dilation

INTRODUCTION

Achalasia cardia is an idiopathic esophageal motility disorder characterized by failure of the lower esophageal sphincter (LES) to relax during the deglutition (1,2). It is a chronic but benign disease of an unknown etiology and is a common cause of dysphagia (3). The most common symptoms of Achalasia are regurgitation, vomiting, dysphagia, cough, recurrent pneumonia, and poor weight gain or weight loss (1,2,4). The "bird's beak" appearance of the lower esophagus on the esophagogram (barium swallow) is a classical radiological finding in the cases of esophageal Achalasia (1,2,4). Achalasia is rare in children, with an estimated annual incidence of only 0.02–0.11 cases per 100 000 children, and it is more infrequent in the below 5-year age group (1,2,4). The goal of treating achalasia cardia is the relief of the functional obstruction at the level of the gastroesophageal junction (3).

CASE PRESENTATION

An 18-month-old female child was admitted with the complaints of regurgitation of feed, nonprojectile vomiting, repeated fever, and cough with occasional breathlessness for last 1 year. The girl regurgitated every feed effortlessly immediately after feeding. Her mother also complained that the baby was not growing well like her peers. The child was delivered by normal vaginal delivery at term at a hospital with average birth weight without any postnatal complications. She was exclusively breastfed up to 6 months of her age. Then weaning was started with suji (a kind of semolina porridge) and khichuri (a South-Asian dish made of rice and lentils). Since then, the child started regurgitation and vomiting following every feed. She had no history of prolonged fever and contact with tubercular patients.

On physical examination, the child was malnourished with loss of subcutaneous fat. Her weight was 5.5 kg, height was 69 cm, and Mid-Upper Arm Circumference was 95 mm. She was severely wasted and stunted. Her Weight-for-Age Z-score was –5.8, Height-for-Age Z-score was –11, and Weight-for-Height Z-score was –6.2. She was mildly pale and anicteric with normal vitals. The other systemic examinations were unremarkable.

On investigation, a complete hemogram revealed anemia (Hemoglobin 7.9 gm%) with normal white blood cell count and platelet count. Serum total protein was 3.3 g/dL, with serum albumin

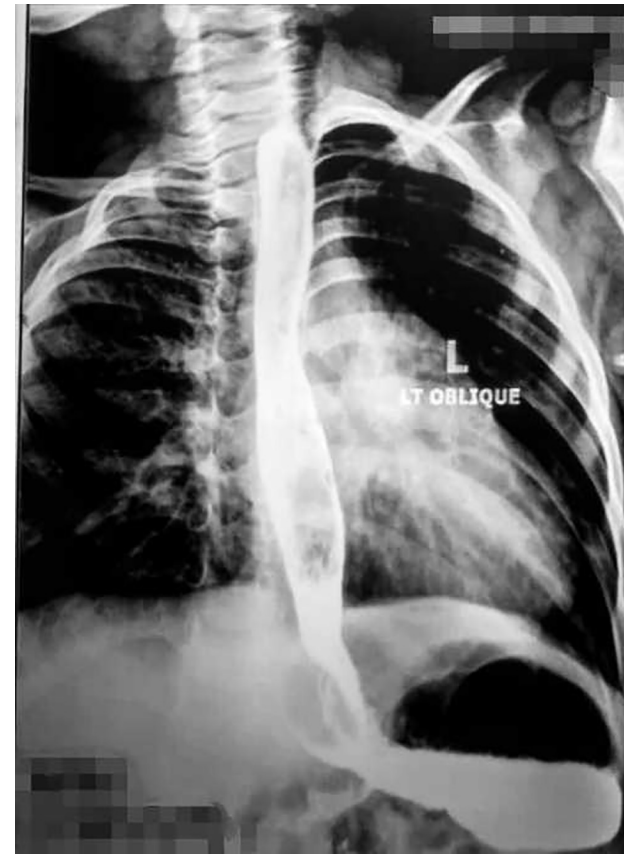

FIGURE 1. The barium swallow showed the "bird's beak" appearance of the lower end of the esophagus. LT, Left (Left Oblique Position).

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The authors report no conflicts of interest.
Parents and guardians of the studied child were informed about this case study, and written consent was obtained.
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of 1.9 g/dL. Blood glucose, urea, creatinine levels, and the levels of liver enzymes were within the normal limits. The chest x-ray and ultrasonography of the whole abdomen were unremarkable. Then a Barium swallow examination was performed, which showed the typical "bird's beak" appearance of the lower end of the esophagus (Fig. 1).

Upper gastrointestinal endoscopy revealed the esophagus was hugely dilated and full of fluid. LES was 20 cm from the incisor teeth, tighter than usual, and almost occluded. However, a 5.5 mm scope could be passed with extra efforts, suggestive of achalasia cardia. The mucosa of the cardia, fundus, body, and antrum appeared normal. The bulb, post-bulbar area, and second part of the duodenum appeared normal (Fig. 2).

After nutritional rehabilitation and correction of anemia, the baby underwent esophageal dilatation using a 10 mm Controlled radial expansion balloon dilator (Fig. 3). The procedure was uneventful.

The baby was well immediately after the procedure, and her symptom was relieved afterward. In a telephonic follow-up after 3 months of the procedure, the mother informed that her general condition was improved, and she had gained weight satisfactorily. Due to the COVID-19 outbreak, the patient did not come to the hospital for a physical follow-up.

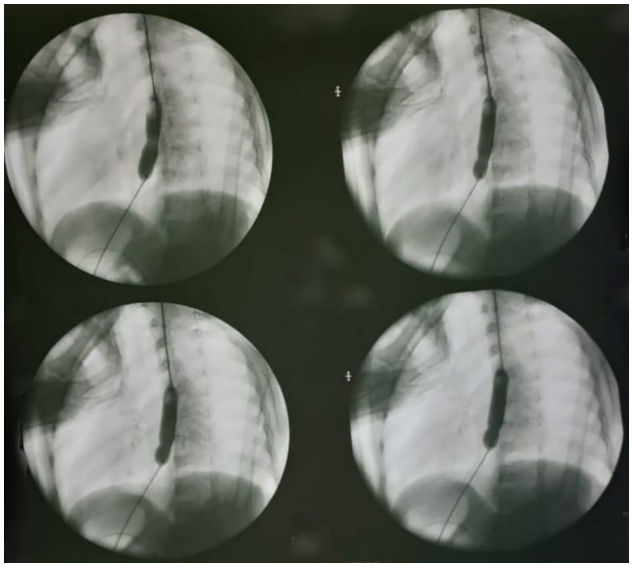

FIGURE 2. Fluoroscopic images of esophageal dilatation of the patient.

DISCUSSION

Achalasia is a primary esophageal motor disorder characterized by loss of LES relaxation leading to functional obstruction of the distal esophagus. Possible etiologies are degenerative, autoimmune, and infectious. In rare cases, Achalasia is familial or part of the triple A syndrome (Achalasia, alacrimia, adrenal insufficiency) (5). The pathological basis of Achalasia is selective loss of postganglionic inhibitory neurons, which leads to sphincter relaxation. Hence, postganglionic cholinergic neurons remain unopposed (5). Therefore high basal LES pressure is produced, and insufficient LES relaxation occurs (5).

Achalasia manifests with regurgitation and dysphagia for solids and liquids and may be accompanied by undernutrition or chronic cough (5). Our case was also presented with dysphagia and failure to thrive. It is uncommon for school-aged children (5), but the presenting case was only 18 months old. The loss of primary peristalsis in the distal esophagus with retained food and poor emptying is often present (5). In Achalasia patients, barium fluoroscopy reveals a smooth tapering of the lower esophagus resembling a bird's beak. Our patient also had a fluoroscopic feature of distal narrowing of the esophagus with proximal dilatation.

Endoscopy can be a diagnostic tool for Achalasia and helps characterize esophageal disease (ie, distinguishing between

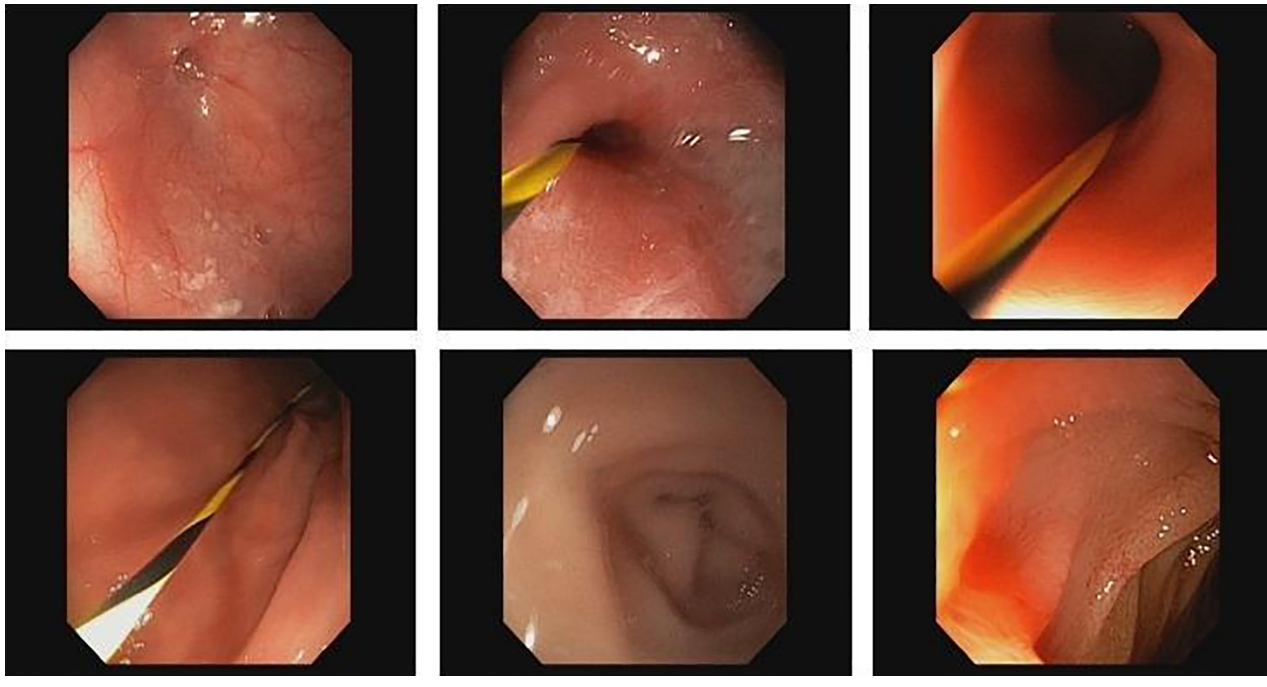

FIGURE 3. Different stages of the endoscopic procedure of the patient.

Gastroesophageal reflux disease and eosinophilic esophagitis) (6). In Achalasia, esophageal dilatation and food stasis are frequent findings at endoscopy, but it is not uncommon to be normal (6). However, in our case, we found that the LES is tighter than usual, and the lower part of the esophagus was almost occluded, suggestive of achalasia cardia. Manometry is the most sensitive diagnostic test and helps differentiate types of Achalasia (5). Unfortunately, manometry is unavailable in our country (Bangladesh).

The goals of achalasia treatment are relief of symptoms, improvement of esophageal emptying, and preventing megaesophagus. The most effective treatment options are pneumatic dilation (PD) and surgical myotomy (Heller's myotomy) (5). PD produces a controlled tear of the LES, resulting in relief of the distal esophageal obstruction and clinical improvement (6). Although laparoscopic myotomy is an effective procedure for young adults, peroral endoscopic myotomy remains a more attractive alternative (5).

Treatment options depend on patient's willingness to undergo an invasive procedure and their physical ability to endure it (7). The medical treatment includes calcium channel blockers, which inhibit the transmembrane influx of calcium in cardiac and smooth muscle; used primarily in adults (7). However, its use in children is not well studied. Other medical treatment options include endoscopic injection of Botox into the LES (7). It acts on the terminal nerve endings and prevents the release of acetylcholine (7). Nevertheless, its optimal dose and frequency are not determined in children. Eventually, most of the cases require either dilation or surgery (5).

We successfully treated our patient with PD using a 10 mm Controlled radial expansion balloon dilator. Careful preparation for this procedure is important to reduce the risk of esophageal perforation and aspiration (6). Pre-procedure preparation includes a clear liquid diet for a few days and a 12-hour fast before the procedure (6).

The results of PD in children with Achalasia are variable and difficult to compare because of variations in the techniques used (6). The most common complication of PD is the risk of esophageal perforation, which ranges from 0.5% to 6% of PD procedures for Achalasia, with a mortality rate of less than 1% (8–10). Our patient experienced no such complication after the procedure.

CONCLUSIONS

Although Achalasia in children is significantly rare, we suggest keeping it in the differential diagnosis of patients with a history of regurgitation of food and dysphagia. There are multiple treatment options available, including both invasive and noninvasive options. However, the choice depends mainly on the availability of that treatment and the patient's willingness.

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REFERENCES

1. Tovar JA. Disorders of esophageal function. In: Coran AG, Adzick NS, Krummel TM, eds. *Pediatric Surgery*. 7th ed. Philadelphia, PA: Elsevier Saunders; 2012:936–946.
2. Franklin AL, Petrosyan M, Kane TD. Childhood achalasia: a comprehensive review of disease, diagnosis and therapeutic management. *World J Gastrointest Endosc*. 2014;6:105–111.
3. Chatterjee S, Gajbhiye V, De A, et al. Achalasia cardia in infants: report of two cases. *J IMA*. 2012;44:44–41.
4. Wakhlu A, Kureel S, Singh S, et al. Retrospective analysis of paediatric achalasia in India: single centre experience. *Afr J Paediatr Surg*. 2012;9:117–21.
5. Khan S, Matta SKR. Upper esophageal and upper esophageal sphincter dysmotility (striated muscle). In: Kliegman RM, St Geme III JW, Blum NJ, et al, eds. *Nelson Textbook of Pediatrics*. 21st ed. Philadelphia, PA: Elsevier; 2020;117–121.
6. Gold BD. Other esophageal neuromuscular disorders. In: Kleinman RE, Goulet OJ, Mieli-Vergani G, et al, eds. *Walker's Pediatric Gastrointestinal Disease*. 6th ed. Raleigh, NC: PMPH-USA; 2018:325–391.
7. Khan N, Liebenberg C, Suleman F. Achalasia cardia in children: a report of two cases. *SAJCH*. 2015;9:65–66.
8. Katsinelos P, Kountouras J, Paroutoglou G, et al. Long-term results of pneumatic dilation for achalasia: a 15 years' experience. *World J Gastroenterol*. 2005;11:5701–5705.
9. Khan AA, Shah SW, Alam A, et al. Sixteen years follow up of achalasia: a prospective study of graded dilatation using Rigiflex balloon. *Dis Esophagus*. 2005;18:41–45.
10. Kim H. Gastroplasty for esophageal perforation after endoscopic balloon dilatation for achalasia: two cases. *J Korean Med Sci*. 2014;29:739–742.