Esophageal achalasia of unknown etiology in infants

Vijay D Upadhyaya, AN Gangopadhyaya, DK Gupta, SP Sharma, Vijayendra Kumar, SC Gopal

Varanasi, India

Background: **Achalasia cardia is an uncommon disease in children particularly in infants. We present 8 cases of achalasia who were encountered over a 12-year period. In infantile achalasia, respiratory symptoms predominate and vomiting may easily be mistaken for gastroesophageal reflux (GER). Vomiting of uncurdled milk is characteristic of achalasia.**

Methods: **In this retrospective study, the data were obtained from records of the Department of Pediatric Surgery IMS, BHU Varanasi, India. The patients were diagnosed by clinical examination and barium study. Any other associated anomalies were noted in these patients. The patients underwent esophagocardiomyotomy with antireflux procedure via the abdominal route.**

Results: **In the present series, 7 patients survived. The follow-up study after operation showed remarkable relief of symptoms with satisfactory weight gain. No post-operative death occurred in the patients. Achalasia associated with alacrimia was noted in one infant who was lost to follow up. One infant was initially diagnosed as having gastric volvulus, but exploration revealed achalasia cardia.**

Conclusions: **Esophageal achalasia is a rare disease in children and its origin is generally indeterminable. Achalasia cardia should be emphasized in the differential diagnosis of an infant presenting with signs and symptoms of esophageal obstruction.**

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Author Affiliations: Department of Pediatric Surgery, IMS, BHU, Varanasi, India (Upadhyaya VD, Gangopadhyaya AN, Gupta DK, Sharma SP, Kumar V, Gopal SC)

Corresponding Author: Vijai Upadhyaya, MD, Senior Resident Pediatric Surgery, Department of Pediatric Surgery, IMS, BHU, Varanasi, 221005, U. P. India (Tel: 91-11-0542-9257; Email: upadhyayavj@rediffmail.com)

Introduction

Thomas Willis first described achalasia in 1674. He successfully treated a patient by dilating the lower esophageal sphincter (LES) with cork-tipped whalebone. Esophageal achalasia of unknown etiology is a rare disease, affecting 1:100 000 of the general population and just 4% to 5% of these cases are described in children,[1] and less than 1% cases in infants. Achalasia involves failure of the LES in conjunction with reduced motility in esophagus itself. The passage of food via the esophagogastric junction is extremely difficult without true organic stenosis or extrinsic compression.

Symptoms of achalasia vary from regurgitation to recurrent aspiration pneumonia. Older children present with symptoms of progressive dysphagia. For these reasons, the disease is often confused with gastroesophageal reflux disease (GERD), delaying correct diagnosis and treatment.[2,3] We present 8 infants with achalasia cardia, aged from 5 months to 9 months (meadian, 5.9 months).

Clinical summary

In this retrospective study, the data of 8 infants from records were analyzed. From January 1990 to December 2002, 8 infants (5 males and 3 females) with primary achalasia cardia were treated at the Institute of Medical Sciences, BHU, India. They were subjected to plain chest X-ray, showing mediastinal air-fluid level in 4 patients and absence of gastric bubble in 3. The patients were confirmed by barium swallow study which showed dilatation of the lowermost esophagus and ruled out other pathological changes. Seven patients underwent operation, and one patient with alacrima refused surgical intervention. All patients were treated by Heller's esophagocardiomyotomy along with partial fundoplication via the abdominal route. The patients took diet on the 6th and 7th post-operative days. They were followed up at an interval of 3 weeks for the first 2 months and then at an interval of 3 months for 2 years.

Most of the patients in this series presented with failure to thrive, followed by vomiting. Some patients presented with recurrent chest infection and respiratory distress (Table). One patient had alacrimia and none of

them had adrenal malfunction. All patients were treated by transabdominal esophagocardiomyotomy along with the antireflux procedure. The operations resulted in remarkable relief of symptoms and satisfactory weight gain. Cough and symptoms of pneumonia were improved within 7 days after operation. Vomiting ceased in all patients at the first follow up, and all patients took diet properly. Weight gain was observed in all patients on subsequent follow up. Recurrent chest infection was improved in all patients except one. During the follow up, 2 patients presented with features of esophagitis which responded to medical treatment and 1 patient presented with acute gastric volvulus, which was explored and treated by anterior gastropexy (Fig. 1, 2).

Table. The symptoms of patients on examination

Symptoms	No. of patients (%)
Failure to thrive	6 (75)
Vomiting	4 (62.5)
Recurrent chest infection	4 (50)
Associated anomaly (alacrimia)*	1 (12.5)

*: patient refusing the operation and lost to follow up.

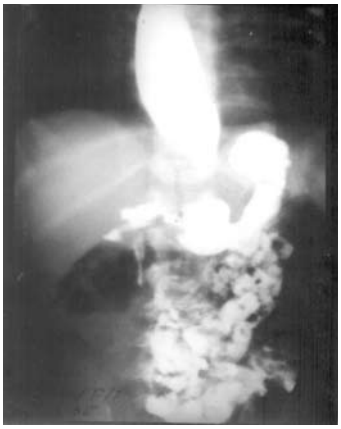

Fig. 1. Barium examination showing dilated lower esophagus.

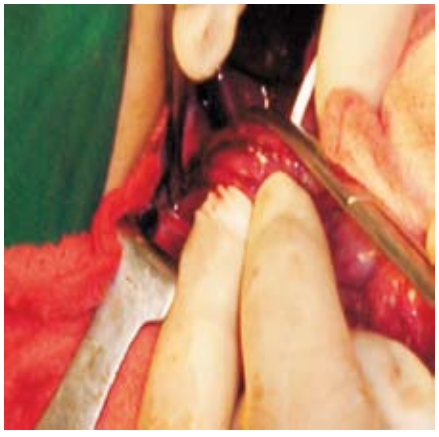

Fig. 2. Per-operative photograph showing a nasogastric tube bulging beneath the mucosa after modified Heller's cardiomyotomy.

There was no post-operative death, but one patient (the case associated with alacrimia) was lost to follow up.

Discussion

Esophageal achalasia is a failure of the coordinated muscle relaxation mechanism of the LES after a peristaltic contraction of the esophageal body, stimulated by a deglutition.[2] The etiology of the disease is unknown and it is rarely seen during childhood. The disease is not family-related, but it affects more male children (6:1) than female.[2-5]

The following theories can explain the onset of the disease: (1) a primary neurogenic abnormality with a failure of the inhibitory nerves and progressive degeneration of ganglion cells; (2) an acquired deficiency of the myenteric plexus ganglion cells, secondary to GERD, Chagas disease, or viral process.[1,4] Clinical presentations include regurgitation of ingested food described as vomiting immediately after meals, a deficit in growth or weight loss, repeated aspiration pneumonia in older children, and complaints of dysphagia.[1-3] Approximately 25%-50% of patients with dysphagia also had episodes of chest pain, which are frequently induced by eating. Adult patients may present with symptoms of heartburn, weight loss and nocturnal cough. Because these symptoms are related to GERD, diagnoses of achalasia of the esophagus are often underestimated.

In our series, 62.5% of the patients presented with vomiting, 75% had failure to thrive, and 50% had a history of recurrent chest infection with respiratory distress, and one had associated alacrimia. As the clinical manifestations of achalasia cardia mimic other causes of esophageal obstruction, it challenges the clinical acumen of the attending physician.

Plain chest radiographs occasionally offer clues in the diagnosis of achalasia. A double mediastinal stripe is occasionally depicted. An air-fluid level can be seen in the esophagus, and this is frequently retrocardiac. In our series, air-fluid level in the mediastinum was observed in 4 patients and gastric bubble was absent in 3.

The classical esophagographic finding for diagnosis of achalasia cardia is bird's beak appearance[6] (Fig. 1). In our series, all patients had typical changes suggestive of achalasia cardia.

Upper gastrointestinal endoscopy is helpful in the diagnosis of achalasia cardia as well ruling out severe esophagitis and malignancy. It was conducted in all our patients. Scintillographic studies and esophageal pH tests are useful to diagnose or rule out GERD,[2-4] but we failed because of lack of the resources. pH monitoring of the lower esophagus may also be required to exclude gastroesophageal reflux that often occurs with achalasia. Esophageal manometry is optional to confirm

the presence of achalasia,[7,8] but we were unable to do this examination on our patients owing to lack of this facility. It may be difficult to differentiate achalasia from congenital esophageal stenosis since both of them may have similar symptoms and radiological appearance, but it was possible to place a nasogastric tube into the stomach to rule out the chances of esophageal stenosis.

Although there is no definite treatment for achalasia, the management should be focused on: relieving the patient's symptoms, improving esophageal emptying, and preventing development of a mega-esophagus. The optimal treatment for achalasia includes several options and presents a challenge for most gastroenterologists. Achalasia can be treated by botulinum toxin injection, pneumatic dilatation or esophagomyotomy, but the most effective treatment options are graded pneumatic dilatation and surgical myotomy.

Classically, esophageal myotomy either by laparatomy or by thoracotomy has been the treatment of choice for the last 40 years.[1,4] Other modalities of treatment with varying success rates are pharmacological and mechanical. Pharmacotherapy includes the use of smooth muscle relaxants like isosorbide dinitrate or calcium channel blockers like nifedipine or local injection of botulin toxin which has been tried with some success in adults.[1,4] Mechanical therapy includes forceful pneumatic dilatation of the esophagus;[1,9] however, symptomatic improvement is mostly seen in older children. Recurrence of symptoms within 6 months and esophageal perforation are the main concerns of the procedure.[9,10]

The classical surgical approach is myotomy for the esophagus and cardia. This extends from the LES to 2 or 3 cm above the cardia, generally associated with anti-reflux procedures, and it is done with a view to the fact that cardiomyotomy alone favors gastroesophageal reflux.[2,6] These children exhibit good postoperative results, and certain series have demonstrated satisfactory results over long-term follow-up.[11,12]

In recent years, laparoscopy has made surgical treatment of esophageal achalasia less invasive. The approach can be either abdominal or thoracic. According to reports,[10,13] the so-called minimally invasive surgery (laparoscopic) does not just represent the most effective treatment for the disease, it also reduces the time of hospitalization (an average of 2 to 3 days) and hospital costs, allowing the child to recover more quickly. But unfortunately this facility is not available at our center.

In the present series, 7 patients were operated on via the abdominal route. Esophagocardiomyotomy (Fig. 2) was done along with the antireflux procedure (floppy Nissen fundoplication), which is considered as a treatment of choice for achalasia cardia. The results of the treatment were satisfactory with no mortality.

In conclusion, achalasia cardia should be stressed in the differential diagnosis of infants presenting with signs and symptoms of esophageal obstruction, and promptly confirmed by upper gastrointestinal contrast examination. Esophageal manometry can be used to confirm the diagnosis. Modified Heller cardiomyotomy is the treatment modality of choice in combination with the antireflux procedure.

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References

1 Hamza AF, Awad HA, Hussein O. Cardiac achalasia in children. Dilatation or surgery? Eur J Pediatr Surg 1999;9:299-302.
2 Jolley SG, Baron HI. Disorders of esophageal function. In: O'Neill JA, eds. Pediatric Surgery, 5th ed. Boston: Mosby, 2000: 997-1007.
3 Ashcraft KW. The esophagus. In: O'Neill JA, eds. Pediatric Surgery, 5th ed. Boston: Mosby, 2000: 325-347.
4 Patti MG, Albanese CT, Holcomb GW 3rd, Molena D, Fisichella PM, Perretta S, et al. Laparoscopic Heller myotomy and Dor fundoplication for esophageal achalasia in children. J Pediatr Surg 2001;36:1248-1251.
5 Porras G, Hernández MH, Izundegui G, Porras JD. Selecting the surgical procedure for simple and complicated esophageal achalasia in children. J Pediatr Surg 1997;32:1595-1597.
6 Nakajima K, Wasa M, Kawahara H, Soh H, Taniguchi E, Ohashi S, et al. Laparoscopic esophagomiotomy with Dor anterior fundoplicacion in a child with achalasia. Surg Laparosc Endosc Percutan Tech 1999;10:236-239.
7 Chelimsky G, Hupertz V, Blanchard T. Manometric progression of achalasia. J Pediatr Gastroenterol Nutr 2000;31:303-306.
8 Tovar JA, Prieto G, Molina M, Arana J. Esophageal function in achalasia: preoperative and postoperative manometric studies. J Pediatr Surg 1998;33:834-838.
9 Lan LC, Wong KK, Lin SC, Sprigg A, Clarke S, Johnson PR, et al. Endoscopic balloon dilatation of eshophageal strictures in infants and children: 17 years' experience and a literature review. J Pediatr Surg 2003;38:1712-1715.
10 Rothenberg SS, Patrick DA, Bealer JF, Chang JH. Evaluation of minimally invasive approaches to achalasia in children. J Pediatr Surg 2001;36:808-810.
11 Morris-Stiff G, Khan R, Foster ME, Lari J. Long-term results of surgery for childhood achalasia. Ann R Coll Surg Engl 1997;79:432-434.
12 Liu JF, Zhang J, Tian ZQ, Wang QZ, Li BQ, Wang FS, et al. Long-term outcome of esophageal myotomy for achalasia. World J Gastroenterol 2004;10:287-291.
13 Mattioli G, Esposito C, Pini Prato A, Doldo P, Castagnetti M, Barabino A, et al. Results of the laparoscopy Heller-Dor procedure for pediatric esophageal achalasia. Surg Endosc 2003;17:1650-1652.

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