Case Report

Achalasia Cardia in Infants: Report of Two Cases

DOI: http://dx.doi.org/10.5915/44-1-9260

Souvik Chatterjee, MBBS, MS (general surgery); Vishal Gajbhiye, MBBS, MS, MCh PDT; Avidip De, MBBS, MS; Sasanka Nath, MBBS, MS, MCh PDT; Dipak Ghosh, MBBS, MS, MCh; Sukanta Kumar Das, MBBS, MS, MCh

Department of Paediatric Surgery, Medical College and Hospital, Kolkata, West Bengal, India

Abstract

Achalasia cardia is a neuromuscular disorder of unknown etiology involving the body of the esophagus and lower esophageal sphincter (LES). It is characterized by aperistalsis of the body of the esophagus and failure of relaxation of lower esophageal sphincter. It usually affects patients between the ages of 30 and 60 years. It is unusual in childhood and extremely rare in infants. We report two cases of achalasia cardia in infants. Both cases were treated with open Heller's esophagocardiomyotomy with anti-reflux procedure.

Key words: Achalasia cardia, Infant

Introduction

Achalasia cardia is a primary motility disorder of the esophagus characterized by aperistalsis of the body of the esophagus and failure of relaxation of the lower esophageal sphincter (LES) appropriately in response to swallowing.[1] It is a chronic, benign disease of unknown etiology and is a common cause of dysphagia. The first case of this condition was reported by Sir Thomas Willis in 1674 in which he described esophageal dilation with whalebone in a patient who had dysphagia and a dilated esophagus. The term achalasia was coined by Hurst in the year of 1927, which means absence of relaxation, more specifically, inadequate LES relaxation in the absence of distal mechanical obstruction.[2] The mainstay of treatment of achalasia cardia is the relief of the functional obstruction at the level of the gastroesophageal junction. The condition is very unusual in pediatric age group, particularly in infancy. About 5% of all cases occur in children; while only a few are reported in infants.[3] Here we present our experience in two infants suffering from achalasia cardia, and both cases were treated successfully.

Correspondence should be directed to:

Souvik Chatterjee, MS (general surgery)
dr.souvikchatterjee@gmail.com

Case 1

A nine month-old female infant was admitted with complaints of regurgitation of feed, non-projectile vomiting, repeated fever and cough with occasional breathlessness for the last three months. The girl regurgitated every feed effortlessly even in upright position within minutes of ingestion of food. The baby was delivered normally at term having a birth weight of 2.8 kg. She was exclusively breast fed until six months of age. She had no family history of the same illness.

On examination, the baby was malnourished and anemic with loss of subcutaneous fat. She had fever with a respiratory rate of 58/min. Her weight was 2.4 kg, and her height was 52 cm. On examination of the respiratory system, bilateral fine crepitations were detected. Examination of other systems was unremarkable.

Complete hemogram revealed anemia (Hb-8.4gm %) with neutrophilic leukocytosis (totalleukocyte count (TLC) - 13,200/ml; neutrophil-72%). The serum total protein was 4 g/dl, with a serum albumin of 2 g/dl. Blood glucose, urea, creatinine levels and liver function test were within normal limits. The chest X-ray demonstrated bilateral patchy opacities suggesting bronchopneumonia and an absence of

the fundic gas shadow. Echocardiographic examination was within normal limits. She was diagnosed with bronchopneumonia and treated with intravenous antibiotics. She was treated conservatively for her regurgitation. Though bronchopneumonia responded to antibiotic therapy, her regurgitation persisted.

The presence of regurgitation of feeds and absence of fundic gas shadow guided us to perform barium swallow examination to rule out achalasia cardia. Barium swallow examination showed dilatation of the esophagus with hold-up of the contrast material and smooth narrowing of the distal esophagus with the typical bird's beak appearance (Figures 1 and 2). Esophageal manometric study was not performed due to technical difficulty. The presence of congenital causes of obstruction was ruled out by performing esophagoscopy. As symptoms and investigational findings suggested achalasia cardia, the infant underwent a modified Heller's esophagocardiomyotomy through open abdominal approach with antireflux procedure (Toupet posterior partial fundoplication). The nasogastric tube was removed on the fifth post-operative day, and oral feeding was resumed. The girl became asymptomatic in the post-operative period, and she started to gain weight with improvement of her general condition.

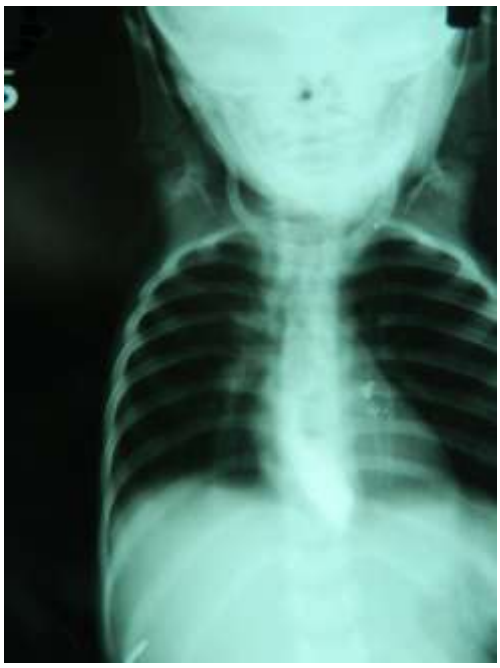

Figure 1. Case 1 - Anterior-posterior view of Barium esophagogram showing bird's beak appearance.

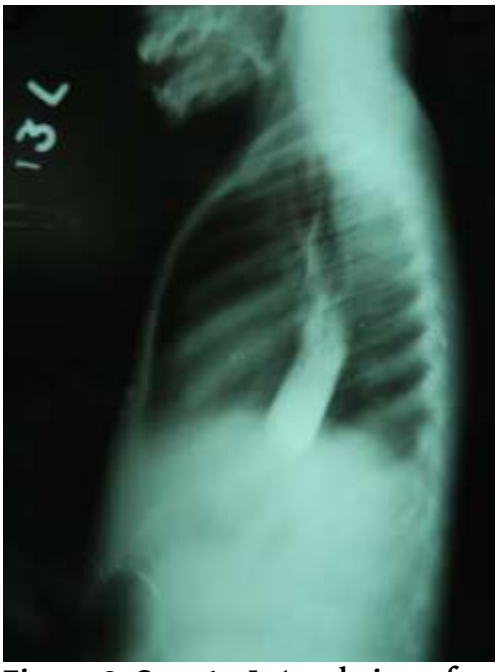

Figure 2. Case 1 - Lateral view of Barium esophagogram.

Case 2

An eleven month old severely malnourished girl was admitted to the pediatric medicine department, suffering from regurgitation of food after each feed with repeated pulmonary infections. On examination, the baby was severely malnourished and anemic. She weighed 2 kg. Her other systemic examination was unremarkable.

On investigation, complete hemogram revealed anemia (Hb- 7.2 gm %) with normal total leukocyte count. The serum total protein was 3.5 g/dl, with a serum albumin of 1.8 g/dl. Blood glucose, urea, creatinine levels and the level of liver enzymes were within normal limits. The chest X-ray demonstrates bilateral patchy pneumonitis. A barium swallow examination was performed. It showed features suggestive of achalasia cardia (Figure 3). Esophagoscopy was performed to rule out congenital anomalies causing upper gastrointestinal obstruction.

After correction of anemia and other parameters, the baby underwent open anterior esophagocardiomyotomy with Toupet posterior partial fundoplication operation. The post-operative period was uneventful. The nasogastric tube was removed on fourth post- operative day. With the symptomatic relief, her general condition improved, and she gained weight rapidly.

Both babies were followed for a year and are doing well and gaining weight without any

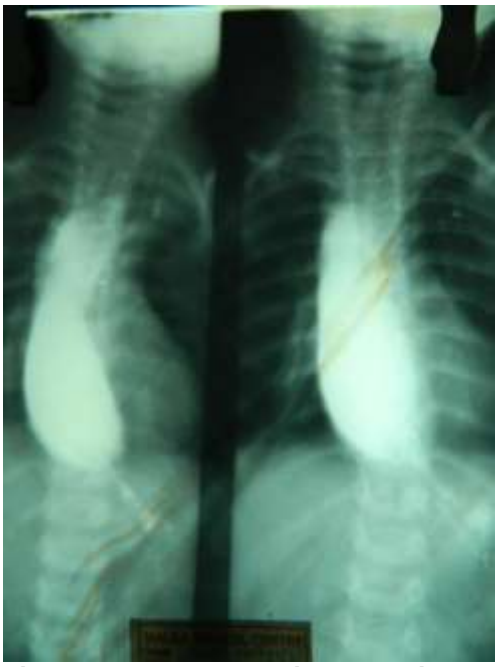

Figure 3. Case 2- Anterior-posterior view of the Barium esophagogram showing typical bird's beak appearance

problems.

Discussion

In the United Kingdom, the incidence of achalasia cardia has been estimated to be 0.5-1.0 in 100,000 persons per year, and the prevalence has been estimated to be around 8 per 100,000. During the period (from 1998 to 2008), the mean incidence of childhood achalasia in the U.K. was 0.18/10,000 children/year. The incidence increased in each of these successive years.[4] The incidence of achalasia cardia in Asian and African populations is lower than that of the British populations (0.3 per 100,000).[5] Racial variations have also been reported. Among the Asian populations, higher incidence is found among Chinese and Indians. In the United States, no difference in incidence is seen between whites and non-whites.[6] There is no sex predilection for the disease.[7]

The peak incidence of achalasia cardia is between the ages of 30 and 60 years and it is exceedingly rare in first two decades of life.[8] Very few patients (4-5%) with achalasia become symptomatic prior to 15 years of age.[9]

Achalasia cardia is a primary esophageal motility disorder. The main pathophysiology is the failure of LES to relax and the absence of esophageal peristalsis. These lead to a functional obstruction at the gastro-esophageal junction. According to the etiology, the disease can be classified into a primary neurogenic abnormality with failure of the inhibitory nerves supplying the sphincter and progressive degeneration of ganglion cells and a deficiency of the myenteric plexus ganglion cells, secondary to gastro-esophageal reflux disease, Chagas disease, or viral infection.[10] Allgrove and colleagues described a rare condition (Allgrove Syndrome) where achalasia cardia was associated with isolated glucocorticoid deficiency with alcrimia.[11,12]

Achalasia cardia is a very rare condition in pediatric age group particularly in infants. The first case in an infant was reported by King in 1953.[13] This child was diagnosed at the age of 6 months. After failure of anticholinergic therapy, Heller's esophagocardiomyotomy was performed at the age of 9 months which led to an excellent symptomatic improvement. Due to the rarity of the disease in infants, there is lack of extensive experience with the disease entity in this age group. The two large institutional series of achalasia cardia comprise only few patients in the age group below 1 year.[12,14] Paucity of experience about the disease in infancy leads to delay in the diagnosis and treatment of the condition.

The common presenting features of achalasia cardia are dysphagia, regurgitation of feeds, emesis, failure to thrive, weight loss, heart burn, chest pain and recurrent lower respiratory tract infections.[15] Because the most common characteristic feature of achalasia cardia is vomiting of uncurdled milk, which is the same symptom of gastro -esophageal reflux disease (GERD),[16] physicians should keep achalasia in mind when evaluating children with persistent vomiting or failure to thrive as occurred in our cases. Plain radiograph may demonstrate the absence of the fundic air bubble, and sometimes a mediastinal air fluid level may present. The barium swallow with cine-esophagogram may show the following features: dilated esophagus, body aperistalsis with smooth narrowing of the distal esophagus and esophago-gastric junction also described as "bird-beak" sign. Endoscopic assessment is essential to rule out other causes of esophageal obstruction, e.g. congenital membrane and acquired stricture. Endoscopy will also demonstrate retained saliva with frothy appearance which will obscure the mucosa and, in advanced cases, sigmoid esophagus with retained food debris can be found. Typical manometric findings of achalasia are high resting pressure of the lower esophageal sphincter (LES) with incomplete relaxation of the

LES with wet swallows and aperistalsis of esophageal body. The manomertic studies are, generally, not done in children due to technical difficulty.

Pharmacological (isosorbide dinitrate, calcium channel blockers, injection of botulinum toxin) and mechanical modalities of treatment are mainly used in older children and adults with varying success. Medical therapy has not provided long-term relief in children. Esophageal dilation with pneumatic dilators has been used in older children, but this procedure has not been used in infants due to its disadvantages and risks.[17] Patients with surgical risk factors are candidates for medical therapy. Sublingual nitrates are more effective than calcium channel blockers in terms of onset of action and esophageal emptying.[18]

Surgical esophagocardiomyotomy is reserved for the patients who failed the primary dilatation or had recurrence of the symptoms following dilatation. Laparoscopic cardiomyotomy not only decreases the morbidity but also causes relief of dysphagia in 80% of cases for 5 years following the procedure.[19,20] Different studies comparing the efficacy of graded pneumatic dilatation with that of laparoscopic cardiomyotomy show better remission rate with the laparoscopic procedure.[21,22,23] In the series reported by Cloud et al, 6 out of 7 patients who had undergone Heller's operation had an excellent result, and only one patient required post-operative dilatation.[24] The proximal esophageal dilatation returned to normal within 2 years and cineradiography showed return of peristalsis. Several authors have recommended the additional antireflux procedure to the myotomy.[25,26] Modified Heller's esophagocardiomyotomy with fundoplication by open or laparoscopic approaches is considered to be the gold standard to relieve the functional obstruction in the distal esophagus and gastro-esophageal junction. It is safe and has excellent long term results. Both the trans-abdominal and the transthoracic routes can be used.[27] The trans-abdominal route along with anti-reflux procedure has been reported to result in better outcome.[9] Though some surgeons prefer a transthoracic approach,[28] it is easier to perform an adequate antireflux procedure through the trans-abdominal route. Gavrilu advocated trans-abdominal myotomy and antireflux procedure using a flap of greater curvature of stomach to be sutured over esophageal mucosa through a left subcostal incision.[29] The operation resulted in dramatic relief of symptoms with disappearance of pulmonary symptoms and rapid weight gain. Most of the series however prefer Nissen-Rossetti total fundoplication after the myotomy , but it carries a higher incidence of dysphagia and gastroesophageal reflux than that of Toupet posterior partial fundoplication.[30,31] In our two cases, we had performed Toupet posterior partial fundoplication with successful outcome.

In conclusion, achalasia cardia is a rare entity in infants. A high index of suspicion is required to diagnose this condition in infants. Even though GERD is a more common presentation than achalasia cardia, the latter condition should be kept in mind in the differential diagnosis in infants with symptoms of intractable regurgitation, persistent vomiting, dysphagia and failure to thrive. An early diagnosis and prompt surgery are necessary to achieve a successful outcome.

Editor's Note: Additional figures are available as supplementary files on the JIMA web site version of the article.

References

1. Patti MG, Fisichella PM, Perretta S, et al . Impact of minimally invasive surgery on the treatment of esophageal achalasia: a decade of change. J Am Coll Surg. 2003;196:698-703. http://dx.doi.org/10.1016/S1072-7515(02)01837-9

2. Reynolds JC, Parkman HP. Achalasia. Gastroenterol Clin North Am. 1989;18:223-55. PubMed PMID: 2668168

3. Hamza AF, Awad HA, Hussein O. Cardiac Achalasia in Children. Dilatation or Surgery? Eur J Pediatr Surg. 1999;9:299-302. http://dx.doi.org/10.1055/s-2008-1072268

4. Marlais M, Fishman JR, Fell JME, et al. UK incidence of achalasia: an 11-year national epidemiological study. Arch Dis Child. 2011;96:192-194. http://dx.doi.org/10.1136/adc.2009.171975

5. Ho KY, Tay HH, Kang JY. A prospective study of the clinical features, manometric findings, incidence and prevalence of achalasia in Singapore. J Gastroenterol Hepatol. 1999;14:791-95. http://doi.org/cx2rw8

6. Sonnenberg A, Massey BT, McCarty DJ, et al. Epidemiology of hospitalization for achalasia in the United States. Dig Dis Sci. 1993;38:233-44. http://dx.doi.org/10.1007/BF01307540

7. Fisichella PM, Raz D, Palazzo F, et al. Clinical, radiological and manometric profile in 145 patients with untreated achalasia. World Journal of Surgery. 2008;32:1974-9.

http://dx.doi.org/10.1007/s00268-008-9656-z
8. Podas T, Eaden J, Mayberry M, et al. Achalasia: a critical review of epidemiological studies. Am J Gastroenterol. 1998;93:2345-7. http://doi.org/dthx5x
9. Olsen AM, Harrington SW, Moersch HJ et al. The treatment of cardiospasm: analysis of a twelve year experience. J Thorac Sur. 1951;22:164-87. PMID: 14861932
10. Vijay D Upadhyaya, AN Gangopadhyaya, DK Gupta et al. Esophageal achalasia of unknown etiology in infants. World J Pediatr. 2008;4:63-5. http://dx.doi.org/10.1007/s12519-008-0013-y
11. Allgrove J, Clayden GS, Grant DB. Familial glucocorticoid deficiency with achalasia of the cardia and deficient tear production. Lancet. 1978;1:1284-6. http://doi.org/ddf253
12. Azizkhan RG, Tapper D, Eraklis A. Achalasia in childhood: a 20-year experience. J Pediatr Surg. 1980,15:452-6. http://doi.org/fccbsw
13. King R. Surgical correction of achalasia of the esophagus in infants. J Med Assoc Ga. 1953;42:466-8. PMID: 12097050
14. Nihoul-Fékété C, Bawab F, Lortat-Jacob S, et al. Achalasia of the esophagus in childhood: Surgical treatment in 35 cases with special reference to familial cases and glucocorticoid deficiency association. J Pediatr Surg. 1989;24:1060-3. http://dx.doi.org/10.1016/S0022-3468(89)80215-5
15. Myers NA, Jolley SG, Taylor R. Achalasia of the cardia in children: a worldwide survey. J Pediatr Surg. 1994;29:1375-9. http://doi.org/b858zg
16. Thomas RJ, Sen S, Zachariah N, et al. Achalasia cardia in infancy and childhood : an Indian experience. J R Coll Surg Edinb. 1998;43:103-4. PMID: 9621534 http://bit.ly/Za6lZN
17. Jackson CA, Liu DC. Achalasia. In: Guandalini S (editor). Textbook of pediatric gastroenterology and nutrition. London and New York: Taylor & Francis; 2004: 61-72.
18. Gelfond M, Rozen P, Gilat T. Isosorbide dinitrate and nifedipine treatment of achalasia: a clinical, manometric and radionuclide evaluation. Gastroenterology. 1982;83:963-96. PMID: 6288509
19. Zaninotto G, Costantini M, Molena D, et al. Treatment of esophageal achalasia with laparoscopic Heller myotomy and Dor partial anterior fundoplication: prospective evaluation of 100 consecutive patients. J Gastrointest Surg. 2000;4:282-9. http://doi.org/cm9bq3
20. Bonatti H, Hinder RA, Klocker J, et al. Long-term results of laparoscopic Heller myotomy with partial fundoplication for the treatment of achalasia. Am J Surg. 2005;190:874-8. http://dx.doi.org/10.1016/j.amjsurg.2005.08.012
21. Wang L, Li YM, Li L. Meta-analysis of randomized and controlled treatment trials for achalasia. Dig Dis Sci. 2009;54:2303-11. http://dx.doi.org/10.1007/s10620-008-0637-8
22. Campos GM, Vittinghoff E, Rabl C, et al. Endoscopic and surgical treatments for achalasia: a systematic review and meta-analysis. Ann Surg. 2009;249:45-57. http://doi.org/dv5dxm
23. Boeckxstaens GE, Annese V, Bruley des Varannes S, et al. The European achalasia trial: a randomized multi-centre trial comparing endoscopic pneumodilation and laparoscopic Heller myotomy as primary treatment of idiopathic achalasia. Gastroenterology. 2010;138(Suppl 1):S53. http://doi.org/c2zh2v
24. Cloud DT, White RF, Linkner LM, Taylor LC. Surgical treatment of esophageal achalasia in children. J Pediatr Surg. 1966;1:137-44.
25. Murray G, Battaglini JW, Keagy BA, et al. Selective application of fundoplication in achalasia. Ann Thorac Surg. 1984;37:185-8. http://dx.doi.org/10.1016/S0003-4975(10)60321-9
26. Ballantine T, Fitzgerald JF, Grosfeld JL. Transabdominal esophagomyotomy for achalasia in children. J Pediatr Surg. 1980;15:457-61. http://dx.doi.org/10.1016/S0022-3468(80)80753-6
27. Tachovsky TJ, Lynn HB, Ellis FH Jr. The surgical approach to esophageal achalasia in children. J Pediatr Surg. 1968;3:225-31. http://doi.org/cq4x4r
28. Moazam F, Rodgers BM. Infantile achalasia. Brief clinical report. J Thorac Cardiovasc Surg. 1976;72:809-12. PMID: 979322
29. Gavriliu D. Aspects of esophageal surgery. Curr Probl Surg. 1975;12:29-36. http://doi.org/fw5846
30. Hunter JG, Trus TL, Branum GD, Waring JP. Laparoscopic Heller myotomy and fundoplication for achalasia. Ann Surg. 1997;225:655-64; discussion 664-65. http://doi.org/db33zn
31. Oddsdóttir M. Laparoscopic management of achalasia. Surg Clin North Am. 1996;76:451-8. http://dx.doi.org/10.1016/S0039-6109(05)70452-7