

Case Reports

Achalasia cardia in an infant

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Sri Lanka Journal of Child Health, 2010; **40**: 74-75

(Key words: Achalasia cardia; infant; reflux; dysphagia)

Abstract

Achalasia cardia is a neuromuscular disorder of the oesophagus, characterized by abnormal oesophageal motility and failure of lower esophageal sphincter relaxation. It usually manifests in the fourth and fifth decades of life. It is unusual in childhood and is extremely rare under the age of one year. We report a 7-month-old child with oesophageal achalasia treated with laparoscopic Heller's oesophagomyotomy.

Case report

A 6-month-old first born male infant of non-consanguineous parents presented to us with regurgitation of feeds and poor weight gain since 3 months of age. He used to regurgitate unaltered feeds immediately after ingestion. He was hospitalized previously twice in the last two months for pneumonia and got treated for gastroesophageal reflux but with no improvement. There was no significant birth / neonatal /family history related to his illness. On examination, the infant was malnourished and anaemic with a weight of 4.6 kg (below the 5th percentile) and a length of 62 cm (below the 50th percentile). Examination of other systems was unremarkable.

Routine blood investigations were within normal limits. Barium swallow with cine-oesophagogram revealed dilated oesophagus and body aperistalsis, failure of lower oesophageal sphincter to relax to the passage of the barium bolus (Figures 1-4).

Motility study could not be done because of technical difficulty in performing oesophageal manometry in

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(Received on 22 April 2010: Accepted on 20 May 2010)

this child. Diagnosis of achalasia cardia was based on clinical grounds and barium cine-oesophagogram. The infant underwent a laparoscopic modified Heller's anterior oesophagocardiomyotomy with fundoplication. Post operative period was uneventful. His follow-up examination has revealed dramatic relief of symptoms and satisfactory weight gain.



Figure 1

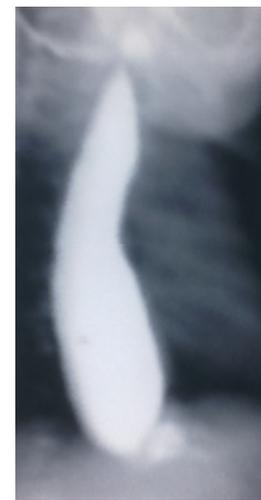


Figure 2



Figure 3



Figure 4

Discussion

Oesophageal achalasia is a relatively rare problem in children. Thomas Willis first described achalasia cardia in 1674. Achalasia is a primary esophageal motility disorder caused by failure of the lower esophageal sphincter to relax and an absence of oesophageal peristalsis leading to a functional obstruction at the gastro-oesophageal junction.

The aetiology of the disease is unknown and there is an almost equal sex incidence. Proposed theories include: (a) a primary neurogenic abnormality with a failure of the inhibitory nerves and progressive degeneration of ganglion cells and (b) a deficiency of the myenteric plexus ganglion cells, secondary to gastro-oesophageal reflux disease, Chagas disease, or viral process¹.

Incidence of achalasia has been estimated as 1 in 10,000. Only 4-5% of patients with achalasia are symptomatic prior to 15 years of age². Achalasia cardia can occur as part of Allgrove syndrome, where it is associated with alacrima and adrenocorticotrophic hormone insensitivity³. Regurgitation of feeds, emesis, dysphagia, recurrent lower respiratory tract infections, weight loss, failure to thrive, chest pain and heart burn are the common modes of presentation⁴. Vomiting of uncurdled milk is characteristic of achalasia⁵. The vomiting may be mistaken for gastro-oesophageal reflux and may delay diagnosis. Retardation of growth and development, as well as the severity of pulmonary symptoms, are profound in infancy and hence early diagnosis and treatment are important.

Plain radiograph features include the absence of the fundic air bubble and a mediastinal air fluid level. In most infants, a barium swallow with cine-oesophagogram will reliably provide the diagnosis. Features include dilated oesophagus, body aperistalsis with smooth narrowing of the distal oesophagus and oesophagogastric junction also described as "bird-beak" sign. Although motility studies are desirable for an absolute evidence for the diagnosis, manometry is performed very infrequently in children due to technical difficulty.

The goal of treatment is to relieve functional obstruction in the distal oesophagus and oesophagogastric junction. A modified Heller's oesophagocardiomyotomy with fundoplication by open or laparoscopic surgery is considered to be the gold standard in the management of achalasia cardia.

Other modalities of treatment described in older children and adults with varying success rate are pharmacological and mechanical. Pharmacotherapy includes the use of smooth muscle relaxants like isosorbide dinitrate or calcium channel blockers (nifedipine) or local injection of botulinum toxin. Mechanical therapy includes pneumatic dilatation of the oesophagus⁶.

In conclusion, achalasia cardia presents a diagnostic challenge in infancy. High clinical suspicion is necessary as the symptoms are easily mistaken for that of the more common GER. Infants may predominantly have respiratory symptoms which may overshadow the vomiting. The children where there was considerable delay in diagnosis continue to have respiratory symptoms. In suspected intractable GER, it is important to rule out obstructive lesions in the oesophagus and upper gastro-intestinal tract by a cine-oesophagogram. Early diagnosis and surgical oesophagomyotomy leads to a successful outcome.

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