



CLASSIC ACHALASIA CARDIA IN CHILDHOOD- A CASE REPORT

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ABSTRACT Achalasia is a neurodegenerative disorder of the lower oesophageal sphincter which occurs rarely in children when compared to adults. These children present with progressive dysphagia, cough, recurrent vomiting, regurgitation of feeds, failure to thrive or weight loss. Here we present our experience in a 7yr old boy with achalasia cardia with typical symptoms and imaging findings. He was successfully managed with pneumatic dilatation.

KEYWORDS : Achalasia cardia, lower oesophageal sphincter, high-resolution manometry.

Introduction:

Achalasia cardia is a neuromuscular disorder of unknown etiology characterized by peristalsis of the body of the oesophagus and failure of relaxation of lower oesophageal sphincter (LES). It usually manifests in the fourth and fifth decades of life and is very unusual in paediatric age group. These children most commonly present with vomiting, dysphagia, regurgitation, and weight loss. Definitive diagnosis is made with barium swallow study and esophageal manometry. The mainstay of treatment of achalasia cardia is the relief of the functional obstruction at the level of the gastroesophageal junction.

Case history:

A 7 year old boy presented to us with complaints of intermittent vomiting, regurgitation of feeds, non projectile vomiting since last one year. There was a history of regurgitation within minutes of ingestion of food. Physical examination and systemic examination was unremarkable. Complete haemogram, liver and renal function tests were normal. The possibility of oesophageal motility disorder was suspected. Barium swallow study done showed mild dilatation of distal cervical and thoracic esophagus with smooth narrowing at the level of the gastro-oesophageal junction with minimal hold up of contrast (Fig 1). No aspiration changes were noted in the lung fields in barium swallow study. Oesophageal manometry study performed demonstrated a stiff LES with resting pressure of >50 mm Hg with incomplete relaxation, absent oesophageal peristalsis suggestive of Achalasia cardia-Type I (Fig 2). As symptoms and investigational findings suggested achalasia cardia, the child underwent pneumatic balloon dilatation. The LES was dilated up to 30mm without any complication. Post procedure period was uneventful. At present, he is under regular follow-up without any symptoms with adequate growth.

Discussion:

Achalasia cardia is an oesophageal motility disorder characterized by failure of LES relaxation. The peak incidence of achalasia cardia is between 30-60 years of age and is rare in first two decades of life(1). Only 4-5% of the patients with achalasia become symptomatic prior to 15 years of age(2). The peak incidence of achalasia cardia is between 30-60 years of age and is rare in first two decades of life(2). The incidence of achalasia in childhood is 0.11/100000 children annually(3).

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 gastroesophageal reflux disease, Chagas disease, or viral infection(4).

The common presenting features of achalasia cardia in children are dysphagia, emesis, regurgitation of feeds, failure to thrive or weight loss, chest pain, heart burn and recurrent lower respiratory tract infections(5). The diagnosis of achalasia is on the basis of the results of gastroscopy, manometry and timed barium oesophagography. A plain radiograph may demonstrate the absence of the fundic air bubble and sometimes a mediastinal air fluid level. Barium swallow with oesophagogram may show dilated body of the oesophagus, with smooth narrowing of the distal oesophagus and gastro oesophageal junction also described as "bird-beak" sign. In our case typical bird-beak sign is noted on barium swallow radiography

With the introduction of high-resolution manometry (HRM) with pressure topography plotting it is possible to classify achalasia into three subtypes (Chicago classification)6. In type I achalasia (classic type), there is 100% failed contractions and no oesophageal pressurization. In type II achalasia (with compression), there will be pan oesophageal pressurization in at least 20% of swallows. Type III achalasia (spastic type), the presence of preserved fragments of distal peristalsis or premature contractions for at least 20% of the swallows is observed. HRM can be used to predict the outcome of each type of achalasia. Patients with type II are more likely to respond to therapies such as pneumatic dilation (PD), heller myotomy (HM), and botulinum toxin (BT) (overall, 70%-100%), followed by type I and type III(6). HRM may play an increasingly important role in the diagnosis of esophageal achalasia in the future, especially when the technique becomes more affordable.

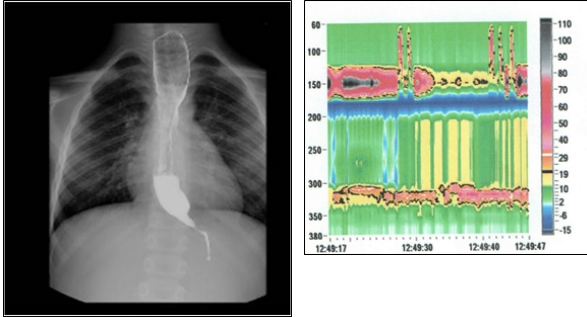
Treatment options depend on parents consent for their child to undergo the surgical procedure or to consider other modalities of treatment. Pharmacological treatment (isosorbide dinitrate, calcium channel blockers, injection of botulinum toxin) and mechanical modalities of treatment are mainly used in older children and adults with varying success. Medical therapy has not provided long-term relief in children. Pneumatic dilatation is generally

considered first-line nonsurgical treatment for achalasia, used in older children, but this procedure has not been used in infants due to its disadvantages and risks(7). In our case report parents opted for pneumatic dilatation though surgery (Heller's myotomy with or without fundoplication) is the most definite treatment of choice(8). At two years of follow-up of our case, there is complete resolution of symptoms. Among motility disorders, only achalasia cardia responds well to treatment.

Figures:

Fig 1: Classic 'bird beak' appearance of the distal oesophagus on barium swallow radiograph

Fig 2: HRM showing classic achalasia cardia type I



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