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Case Report

Radiology Section

An Infant with Nasal Regurgitation Since Birth and Failure to Thrive

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ABSTRACT

The condition achalasia cardia is rare in paediatric age group, especially in infants. An 11-month-old female infant presented with complaints of oronasal regurgitation since birth and failure to thrive. Upper GI contrast study was conducted which demonstrated massive dilatation of lower 2/3rd of oesophagus with abrupt narrowing at lower oesophageal sphincter and positive 'bird beak sign'. On the basis of radiological findings infantile achalasia cardia was diagnosed and patient underwent modified Heller's Oesophagocardiomyotomy with anti reflux procedure. Post operatively the symptoms subsided and weight gain was noted after six month follow up. Although functional infant regurgitation and Gastro-oesophageal reflux (GER) is common in infancy, uncommon causes like achalasia cardia should also be considered as a differential when symptoms are persisting.

Keywords: Achalasia cardia, Heller's myotomy, Upper GI contrast study

CASE REPORT

An 11-month-old female infant presented with complaints of oronasal regurgitation since birth and failure to thrive. The child also had complaints of recurrent respiratory distress, fever and increased frequency of regurgitation since seven months of age and constipation from last one month. Upon examination, pulse was 150/min, Respiratory rate was 35/min and temperature was examined to be 101°F. The routine pathological investigation revealed normal parameters. On examination we noticed that infant had effortless regurgitation of uncurdled milk after every feed, even in upright position.

The motor and cognitive milestones were found to be at par with age. The weight of the infant was noted to be 7 kg (3rd percentile for 11 months) and head circumference was 44 cm. Examination of other system was unremarkable. The family history was found to be insignificant. X-ray Chest of the patient [Table/Fig-1] revealed a large patch of consolidation in mid and lower zone of right lung with mediastinal widening possibly due to oesophageal dilatation and absent fundal gas in stomach. Upper GI contrast study was performed using non ionic water soluble contrast agent. The contrast was given with a breast feeding like technique using a plastic bottle and a nipple. The study revealed massive dilatation of lower 2/3rd of oesophagus with pooling of contrast and air fluid level [Table/Fig-2]. There was abrupt smooth tapering at the level of lower oesophageal sphincter [Table/Fig-3a,b]. The narrowed lower oesophageal sphincter (LOS) gives 'Bird's Beak' like appearance which is characteristic finding of Achalasia Cardia. Oesophagoscopy was done and possibility of stricture, diverticulum and obstruction was ruled out [Table/Fig-4].

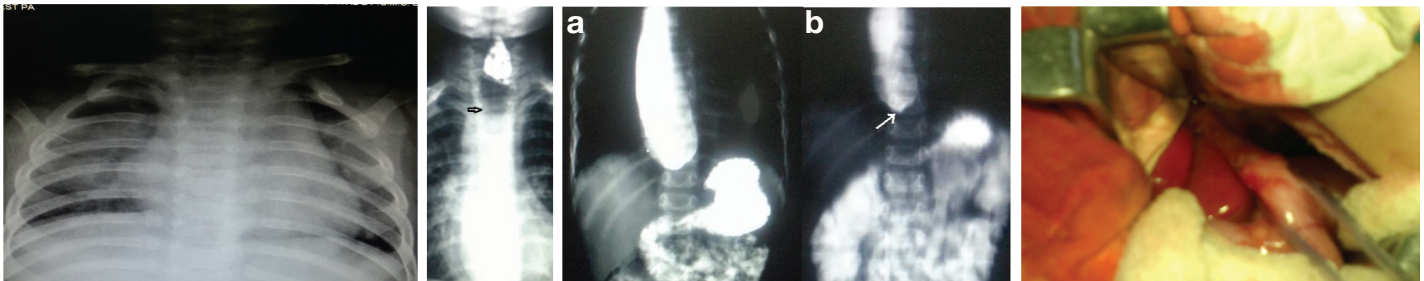
Oesophageal manometry could not be performed because of non-availability of oesophageal manometry facility for infant at our institution and unaffordability of patient's parents for particular investigation at other institute. Infant underwent a Modified Heller's Oesophagocardiomyotomy through sub costal route with anti reflux procedure, which has been reported to give the best results for achalasia [1]. Other associated congenital abnormalities like narrow oesophageal hiatus were ruled out by visualization. Asymptomatic ladd's band/malrotation was specifically looked for during exploration. The nasogastric tube was removed on 4th postoperative day and child was able to resume normal feeding. Feeding in prop up position and solidification of diet was advised post operatively to prevent reflux. In 6 month postoperative follow up the infant was symptom free and started gaining weight.

DISCUSSION

The word Achalasia Cardia comes from a Greek word which means "Does not relax". Thomas Wills first described achalasia in 1674. He successfully treated a patient by dilating the lower oesophageal sphincter (LOS) with cork-tipped apparatus. Oesophageal achalasia of unknown aetiology is a rare disease, affecting 1 per 100,000 of general population and just 4-5% in children [2] amongst it less than 1% cases in infants. The usual presentation of Achalasia in adults is frequently reported between fourth to sixth decades of life and rare in first and second decade [3]. 1st case in an infant was reported by king R in 1953 [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]. Among the Asian population, higher incidences have been reported among Indians and Chinese. Sporadic cases have been reported by few authors worldwide till last year. Very few patients from the age group below one year accounted for the total population in two large institutional series of achalasia cardia [6,7].

Oesophageal achalasia characteristically leads to failure of the coordinate muscle relaxation mechanism of the LOS after a peristaltic



[Table/Fig-1]: X ray chest PA view reveals a large patch of consolidation in right mid and lower zone with Mediastinal widening possibly due to dilated oesophagus and absent fundal gas
[Table/Fig-2]: Upper GI contrast study reveals massive dilatation of lower 2/3 rd of oesophagus with pooling of contrast and air fluid level [arrow]
[Table/Fig-3a,b]: Upper GI contrast study reveals massive dilatation of visualized oesophagus with abrupt narrowing at lower oesophageal sphincter and positive bird beak sign [arrow]
[Table/Fig-4]: Intra operative image showing abruptly narrowed oesophageal sphincter

contraction of the oesophageal body, stimulated by deglutition [8]. Dysmotility of lower oesophageal sphincter and abnormal peristalsis of the oesophagus to the act of swallowing comes out as a result of neuromuscular disorder i.e. ganglionic cells degeneration. The peristalsis in distal segment of the oesophagus may be due to an abnormality of Auerbach's plexus which is responsible for smooth muscle relaxation, leads to weak contractions that are uncoordinated and as a result, non propulsive. The aftermath of oesophageal dilation leads to the food stasis leading to oesophagitis, regurgitation, aspiration pneumonia and bronchial spasm. The LOS fails to relax, either partially or completely with elevated pressures which can be explained manometrically. Oesophageal manometry is optional to confirm the presence of achalasia [9,10].

Non surgical options of treatment for achalasia cardia are pharmacological and mechanical which gives only temporary relief of symptoms with varying success rate and some adverse effects. Medical therapy 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 but in infant it is not suitable because they don't give satisfactory and long term outcome [1,11]. The use of dilators to treat achalasia is as old as the disease. In 1664, Thomas Willis described mega esophagus and used the first rudimentary dilator made of whale bone. Since then, many mechanical, hydrostatic and pneumatic dilators have been used however symptomatic improvement is mostly seen in older children and adults. Younger children do not have much relief of symptoms (only 30%) and suffer many complications (20% perforations) [1]. Despite apparent advantages, dilatation also presents many risks such as perforation, gastro oesophageal reflux & recurrence and furthermore, children are uncooperative and general anaesthesia is required for these maneuvers [1,11]. So we did not considered for dilatation, pharmacotherapy & botulinum toxin and decided to go for surgery.

Since the most common characteristic feature of infantile achalasia is vomiting of uncurdled milk, which is also a feature of gastro-oesophageal reflux disease (GERD), it can mimic GERD and hence, can be misdiagnosed [12]. To rule out the other possibilities, foremost radiological evaluation of underlying cause by upper GI study for persistent ongoing vomiting/regurgitation is obligatory.

Differential diagnosis of nasal regurgitation include; defect in soft palate, pharyngeal stricture, diverticulum, oesophageal obstruction and functional swallowing disorder which can be ruled out by clinical examination, upper GI endoscopy/contrast study and clinical history. Regurgitation often mistaken for vomiting and regurgitate stomach contents look alike vomitus but some classical features of vomiting can differentiate it from regurgitation e.g. force, discomfort which is usually preceded by nausea, retching and gagging. The international group of expert, the paediatric Rome working team has defined functional infant regurgitation as with symptoms consisting of i) at least three week of regurgitation of stomach content, ii) at least twice daily during infancy and iii) the first year of life. Presence of other associated symptoms like retching, bleeding, refusal to feed or breathing problems suggests other disease rather than a functional disorder [13].

It is also important to know difference between similar type of terms e.g. gastroesophageal reflux (GRE), gastroesophageal reflux disease (GERD) and functional infant regurgitation. Gastroesophageal reflux episode occur when contents of stomach move backward up into oesophagus. Regurgitation episode occur when contents of stomach reaches up to mouth.

GERD refers to consequence or harmful effect that happen due to reflux. GERD is not common in otherwise healthy infant but it is commonly present in infant with preterm birth, cerebral palsy and chronic lung disease. Frequently occurring complications of GERD are aspiration pneumonia, oesophagitis, midline chest pain, food refusal, failure to gain weight and anaemia.

This case clearly demonstrates the significance of upper GI contrast study for diagnosis. Modified Heller's Oesophagocardiomyotomy with anti reflux procedure is the treatment of choice for infantile achalasia. Good to excellent clinical response has been reported in long term survey after Heller's Oesophagocardiomyotomy and no further surgery or dialatation was required in most of the cases [14-16].

CONCLUSION

Although gastro-oesophageal reflux is very common in infancy but other causes of oronasal regurgitation should also be considered if problem is persisting and causing complications. Thorough clinical examination, upper GI contrast study and endoscopy are indicated if regurgitation is persisting and causing failure to thrive, weight loss, refusal to feed and respiratory symptoms. Achalasia is rare in infancy but should be kept in mind if regurgitation is persisting and causing complications. It can be diagnosed easily by upper GI contrast study. Heller's Oesophagocardiomyotomy is the treatment of choice and has been proved to give good result in long term survey.

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