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Case Report

Unusual Cause of Stridor in a Preschool Child

Budensab A H[*@], Praveen S. Bagalkot[*], Venkatesh Annigere[*], Suhas N. Joshi[**]

[*]Associate Professor, [**]Professor
Department of Pediatrics, SDM College of Medical Sciences and Hospital, Dharwad, Karnataka

[@]Correspondence Email: budensab.sab@gmail.com

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ABSTRACT

Persistent stridor in a preschool child could be due to variety of reasons like laryngomalacia, laryngotracheomalacia, foreign body, subglottic tracheal stenosis, vascular ring, gastro esophageal reflux, mediastinal mass compressing trachea and achalasia. Achalasia presents as dysphagia for solids and liquids and may be accompanied by undernutrition or respiratory symptoms; usually presents in school age children however rarely it can present in preschool children. We report a 4.5 year old child who had persistent stridor since the age of 6 months and was subsequently diagnosed to have achalasia.

Key words: persistent stridor, achalasia, esophagus,child

INTRODUCTION

Achalasia is a primary esophageal motility disorder characterized by loss of lower esophageal sphincter (LES) relaxation and loss of esophageal peristalsis, both contributing to a functional obstruction of the distal esophagus.1It could be primary in which there is absence of ganglion cells in the myenteric plexus or it could be secondary to inflammatory or autoimmune diseases where there is progressive degeneration of ganglion cells. [1] It usually presents with dysphagia, regurgitation and heartburn. Rarely, it may be complicated by acute airway compromise causing stridor. [2] We report a 4.5 year old child who presented with persistent stridor and on work up found to be having hugely dilated lower esophagus compressing trachea causing persistent stridor.

CASE REPORT

4.5 year old boy who was born to consanguineous marriage presented with noisy breathing and persistent stridor since the age of 6 months. Stridor was gradual in onset and progressive. There was no history of dysphagia to solids or liquids, fever, regurgitation or foreign body. They consulted several doctors in the past for the same with no improvement. On examination child was conscious, afebrile, pulse rate 84/minute regular, RR 34/minute, weight

8.6kg height 80 cms which were below 3rd centile.

Respiratory system examination revealed inspiratory stridor with chest retractions, pectus excavatum, trachea central, vesicular breathing with inspiratory stridor and expiratory wheeze. Other systems were normal.

The child was admitted to our hospital for evaluation of stridor (Fig. 1).

CBC was normal. Serum calcium, phosphate and alkaline phosphotase normal.

Chest X ray showed air filled dilated esophagus (Fig. 2).

Barium swallow showed significant dilatation of the distal 2/3rd of the esophagus with smooth, narrowing at cardio-esophageal junction suggestive of Achlasia cardia (Fig. 3).

CT scan of neck and chest (Fig. 4)- plain and contrast revealed significant dilatation of entire length of oesophagus with abrupt narrowing at the gastrooesophageal junction and resultant hold-up of intraluminal air and oral contrast suggestive of Achlasia cardia.

Child's condition was treated by modified Heller's oesophagocardiomyotomy.The surgery and recovery were uneventful, and the child was relieved from respiratory symptoms after surgery and the patient began to gain weight.

Fig. 1. Child for evaluation of stridor.

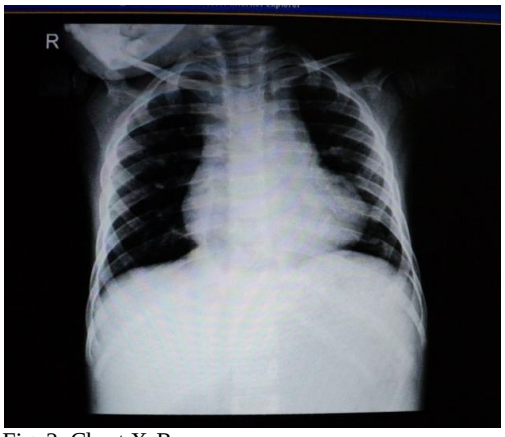

Fig. 2. Chest X-Ray

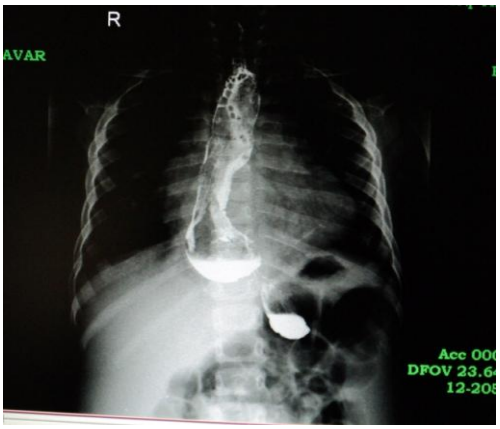

Fig. 3. Barium swallow examination

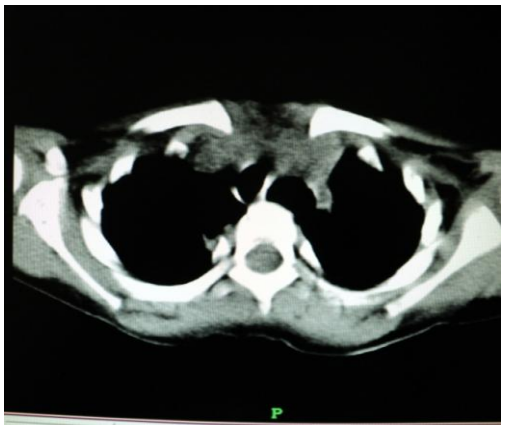

Fig. 4. CT scan chest

DISCUSSION

Primary esophageal achalasia is rare in pediatric patients. It commonly presents with gastrointestinal symptoms such as dysphagia, postprandial vomiting, failure to thrive and retrosternal chest pain. Pulmonary

symptoms, however, can occur and are mainly due to tracheal aspiration of the esophageal contents. Patients may present with chronic or nocturnal wet cough, episodes of persistent bronchospasm or recurrent pneumonia. [1, 2,4] Another cause of pulmonary symptoms in patients with achalasia is tracheal compression caused by a distended esophagus, which was initially reported by Bello et al [5] in 1950. Airway obstruction may manifest as dyspnea, wheeze and/or stridor. Very few cases of isolated persistent stridor due to achalasia in a preschool child are reported in literature. These symptoms can be insidious or acute at rest, after a meal, or during exertion and high respiratory demand.

Certain investigations are helpful in diagnosing achalasia1. A chest radiograph may show an air-fluid level in a dilated esophagus. A contrast esophogram may demonstrate retained food in an aperistaltic distal esophagus that exhibits a smooth taper leading to the closed lower esophageal sphincter. This radiological sign is known as 'bird's beak', and is usually indicative of esophageal achalasia. Esophageal manometry is the only test that confirms the diagnosis – evidenced by incomplete relaxation of a high-pressure lower esophageal sphincter during swallowing. Spirometry and the maximal expiratory and inspiratory flow-volume loop are important components of the pulmonary evaluation [6] in addition to radioimaging that includes chest radiograph and/or CT scan. Because of its rarity and nonspecific presentation, a diagnosis of achalasia may be delayed; the mean duration of symptoms before diagnosis can be up to 28 months. [1, 2,4]

The goal of treatment of achalasia is to relieve functional obstruction in the distal oesophagus and oesophagogastric junction. A modified Heller's oesophagocardiomyotomy is considered to be the gold standard in the management of achalasia cardia. [7] However, the addition of anti-reflux operation is still a matter of controversy. Some surgeons find an added anti-reflux procedure unnecessary, but because of the documented risks of Barrett's syndrome and severe GORD syndrome seen in 36% children, we feel that it's a useful adjunct, as long as it doesn't cause distal oesophageal obstruction. [8, 9] A modified Heller's oesophagocardiomyotomy with fundoplication can be done by open method or by laparoscopy. [4, 7] Other modalities of treatment are pharmacological or mechanical. Pharmacological treatment, in the form of calcium channel blockers (nifedipine) and phosphodiesterase inhibitors, offer temporary relief of dysphagia only. Botulinum toxin is an expensive option and is initially effective in treating the achalasia; however, the long-term results are disappointing. [10] Mechanical therapy includes forceful pneumatic dilatation of the esophagus; symptomatic improvement is mostly seen in older children. However esophageal perforation and recurrence of symptoms within six months are the main concerns of the procedure.

SUMMARY

Esophageal achalasia is a rare disease of childhood and may present with dysphagia, vomiting, recurrent wheezing, failure to thrive and rarely as stridor due to compression of trachea by dilated esophagus. Pediatricians and pediatric respirologists should consider the diagnosis in children with persistent stridor and failure to thrive.

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