CASE REPORT

Congenital Cricopharyngeal Achalasia in an Infant Treated by External Cricopharyngeal Myotomy: A Case Report

Vivek Soman[1], Jayakumar R Menon[2], Manju E Issac[3], Jayanthy Pavithran[4]

Abstract

Congenital cricopharyngeal atresia is a rare but serious cause of dysphagia in infants. The failure of the cricopharyngeus muscle (CPM) to adequately relax on food intake is considered as the cause. This condition usually presents early in life with symptoms that include regurgitation, nasopharyngeal reflux, aspiration, choking episodes, and failure to thrive. Videofluoroscopy helps to clinch the diagnosis, demonstrating a prominent cricopharyngeal bar in most cases. This describes the case report of a male infant who presented with cricopharyngeal achalasia (CPA) and underwent open cricopharyngeal myotomy at 6 months of age, leading to the resolution of symptoms.

Keywords: Achalasia, Aspiration, Cricopharyngeus, Dysphagia, Myotomy.

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Introduction

Cricopharyngeal achalasia (CPA) is a rare cause of dysphagia in children and is often diagnosed late. It can cause significant morbidity in the pediatric population. In 1915 Jackson had described a condition in adults characterized by dysphagia due to spasm of the upper esophageal sphincter, which was secondary to a neurological disease.[1] In 1967, Utian and Thomas gave the first description of CPA in infancy.[2]

The cricopharyngeus muscle (CPM) is the major functional component of the upper esophageal sphincter. It is a C-shaped striated muscle situated between the inferior pharyngeal constrictor and the esophagus. It extends around the pharynx from one end of the cricoid arch to the other. CPM remains contracted at rest which protects the airway from gastric reflux and prevents aerophagia. The relaxation of CPM leads to the opening of the upper esophageal sphincter, facilitating the bolus into the food pipe. Any problem affecting the relaxation of CPM can lead to dysphagia, regurgitation, and aspiration into the airway. The exact pathogenesis of this condition remains obscure. In children, it occurs as an isolated finding in contrast to adults, which is linked to neurological processes.[3] Infants usually present in the perinatal period with feeding difficulties, choking episodes, nasopharyngeal reflux of feeds, and features of aspiration into the lower airway. Nearly 90% of reported cases are diagnosed in children under 1-year of age, but only 15% are diagnosed within 1-month of life.[4]

High index of suspicion of this condition should be maintained by clinicians while diagnosing infants with dysphagia. Early diagnosis is imperative because of the potential for surgical correction and to avoid complications due to aspiration.

Case Description

A 2-month-old male baby was referred to us with complaints of choking and regurgitation of feeds since birth, and poor weight gain. The child was not tolerating breastfeeding. He was evaluated in an outside hospital and was suspected of having a laryngeal cleft with tracheoesophageal fistula. The child has been on nasogastric (NG) tube feeding since then. On evaluation, the child was found to be emaciated with weight below the 3rd centile. The child appeared to cough and sputter up the breastmilk on feeding. Videofluoroscopy showed obstruction to the flow of barium with a persistent narrowing at the C6 level (cricopharyngeal bar) (Fig. 1). The pharynx above the CPM was distended in contrast with secondary aspiration into the trachea. Examination under anesthesia revealed a normal larynx. The interarytenoid area was normal on palpation and hence laryngeal cleft was ruled out. Esophagoscopy was done, which revealed a spasm of the upper esophageal sphincter, and the esophageal lumen was normal. CT neck with the chest was performed to rule out any external compression of the esophagus. CT showed aspiration pneumonia of the right upper and lower lobe and left lower lobe with no evidence of external compression. The child was treated with oral antibiotics, nebulization, and chest physiotherapy. During the stay in the hospital, Ryles tube feed was escalated, and a steady weight gain was ensured. After 4 months, when adequate weight gain (Weight = 6.8 kg) was achieved child underwent external cricopharyngeal myotomy under GA. Before the procedure NG tube was secured to aid in the identification of the esophagus and for postoperative feeding. A transverse cervical incision, typically on the left was put to access the CPM.

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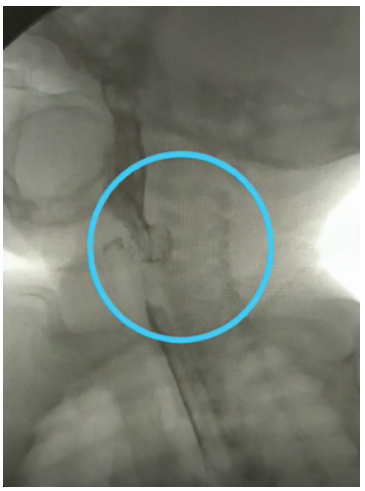


Fig. 1: Videofluoroscopic swallowing study performed at the time of presentation showing hold up of barium in the pharynx with little passage into the esophagus

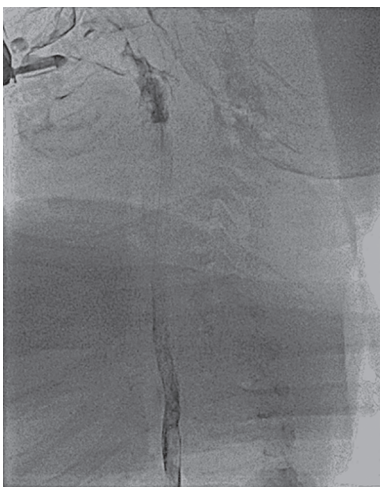


Fig. 2: Videofluoroscopic swallowing study performed 2 weeks after cricopharyngeal myotomy showing the passage of barium into the esophagus with no aspiration into the trachea

The sternocleidomastoid muscle and carotid sheath were then retracted laterally. Meanwhile, the larynx was rotated to expose the transverse muscle fibers of the CPM. A rigid esophagoscope was introduced to distend the esophagus, and the muscle was sharply divided in the midline without damaging the pharyngeal and esophageal mucosa. Nasogastric tube was left in place for 2 weeks for feeding in the postoperative period. After 2 weeks of the procedure, fluoroscopy was repeated, which showed resolution of the obstruction without any aspiration into the airway (Fig. 2). Hence Ryles tube was removed, and oral feeding was initiated.

Discussion

Although swallowing problems arising in the pharyngoesophageal segment may be due to various factors, the term CPA specifically denotes the failure of the CPM to relax appropriately during the pharyngeal phase of swallowing. Apart from problems related to dysphagia, regurgitation, and aspiration, some infants can present with inadequate weight gain and failure to thrive.[3] This condition may go unnoticed in some cases and can lead to sudden death.[5] Several congenital malformations such as esophageal atresia or stenosis, tracheoesophageal fistula, vascular rings/slings, and laryngeal cleft should be considered in the differential diagnoses of this condition.

A proper clinical history with a meticulous head and neck and neurological examination is quite helpful in arriving at a diagnosis. The practice of observing children while feeding is essential. This should be further augmented with the application of specific swallowing techniques.[6]

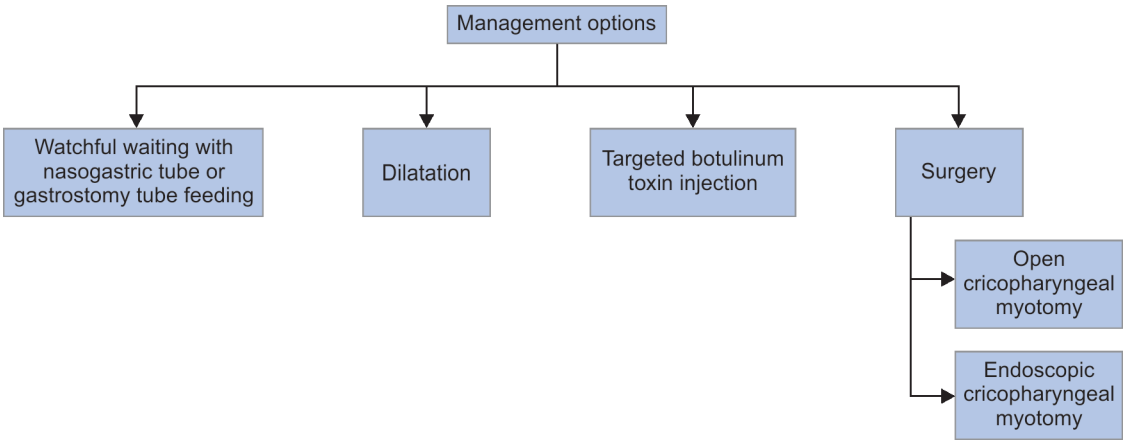
The gold standard for diagnosing cricopharyngeal achalasia is the videofluoroscopic swallow study (VFSS) which demonstrates a cricopharyngeal muscle bar. The VFSS also allows for estimating the efficiency of pharyngeal contraction, hyolaryngeal excursion, and presence of penetration (up to level of true vocal cords), aspiration (beyond true vocal cords), nasopharyngeal reflux, or gastroesophageal reflux.[7] Flexible endoscopic evaluation of swallowing (FEES) often compliments VFSS as it is often conducive for ruling out other causes for dysphagia or aspiration, such as obstructive masses or vocal fold immobility.[3] Manometry is not routinely employed for diagnosing CPA in the pediatric population because of the lack of normative data on UES pressure in this age group and interinstitutional variation of measuring technique employed.[8,9] In addition, the probe may move naturally during deglutition making it challenging to pinpoint the area of dysfunction exactly.

The role of gastroesophageal reflux (GER) in pediatric CPA is still debated with opposing evidence in the literature. Comparisons of UES pressure *via* manometry in infants with and without GER shows no difference in mean pressure or coordination of UES relaxation with swallowing.[10] However, Scholes et al. in their series of four infants with CPA who underwent endoscopy with biopsy, three were found to have esophagitis.[11] GER needs to be addressed simultaneously or before treating cricopharyngeal achalasia as the clinical manifestations of GER may flare up after cricopharyngeal myotomy.[12]

This condition resolves in some neonates suggesting that it may be attributable to a relative state of neuromuscular immaturity.[4] However, the literature lacks information about which patients are likely to resolve and how long one should wait, making early intervention the best option to avoid failure to thrive and complications of aspiration. In 1972 Blank first reported the use of bougienage dilatation for a child with CPA.[13] Some have reported successful dilatation of CPM using high-pressure balloons.[14,15] Skinner advocates an initial trial of dilatation before proceeding to more invasive procedures like myotomy.[16] Botulinum toxin injection, which inhibits acetylcholine release at the neuromuscular junction, can be used as a temporary measure before standard procedures like cricopharyngeal myotomy. In our case, open cricopharyngeal myotomy was done without resorting to botulinum toxin injection because it is a safe and effective technique, and the child can learn the act of swallowing early in life.

The cricopharyngeal myotomy is well tolerated with no significant complications, with long-term follow-up showing promising results in other studies.[17-19] Chun et al. report endoscopic cricopharyngeal myotomy for the management options for cricopharyngeal achlasia in an infant is represented in Flowchart 1.[20] Although the endoscopic approach can prevent

Flowchart 1: Management options of cricopharyngeal achalasia in infants



recurrent laryngeal nerve injury; it can increase the risk of salivary leak and mediastinitis.[3]

Conclusion

CPA is characterized by failure of the cricopharynx to relax during deglutition, preventing food entry into the esophagus. Although a rare cause of dysphagia in infants, it can cause significant morbidity because of coughing, choking, and recurrent chest infections. The diagnosis is made by the presence of a prominent cricopharyngeal bar on videofluoroscopy. Cricopharyngeal myotomy is a definitive procedure that is tolerated well in an infant and is associated with good results.

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