

Congenital cricopharyngeal achalasia: a rare cause of dysphagia in an infant

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Dysphagia secondary to congenital cricopharyngeal achalasia (CCA) is a rare condition in pediatric patients. We report a case of CCA in a 10-month-old boy presented with dysphagia, choking and nasal reflux. The diagnosis was made by barium studies. The patient was successfully treated by cricopharyngeal myotomy.

Key words: cricopharyngeal achalasia, dysphagia, children.

Oropharyngeal and esophageal disorders produce dysphagia. Oropharyngeal dysphagia is caused by abnormalities affecting the neuromuscular mechanism of the oral cavity, pharynx, and upper esophageal sphincter (UES). The cricopharyngeus muscle is the major component of the UES. Primary cricopharyngeal achalasia resulting from failure of relaxation of the cricopharyngeus muscle is a rare cause of dysphagia in children¹. We herein present the case of a 10-month-old boy with cricopharyngeal achalasia treated by myotomy.

Case Report

A 10-month-old boy was referred to our hospital because of cyanosis and dysphagia. The patient had been delivered full-term, through spontaneous vaginal delivery after an uncomplicated pregnancy. He had been hospitalized several times because of cyanosis, dysphagia, nasopharyngeal reflux and repeated episodes of aspiration pneumonia since birth and required tube feeding.

On admission, the patient weighed 6300 g (<3rd percentile). He had peroral cyanosis. Physical examination was unremarkable except systolic murmur heard along the second right intercostal space with radiation to the back. Choking, nasopharyngeal reflux and cyanosis persisted at every feeding during

his hospitalization. Serum electrolyte analysis and thyroid functions were normal, as were the remainder of his laboratory tests. The patient was diagnosed as tetralogy of Fallot and major aortopulmonary collateral arteries after cardiologic evaluation. Cine-esophagogram with fluoroscopic observation of the swallowing showed regular posterior narrowing on the posterior wall of the esophagus at the level of 4th-5th cervical vertebrae with enlargement of the hypopharynx (Fig. 1). Therefore, he underwent cricopharyngeal myotomy and pharyngeal plexus neurectomy. He tolerated feeding very well on the 3rd postoperative day and was discharged after two weeks. He is presently 20 months old, has no feeding problems and an operation to correct congenital heart disease is planned.

Discussion

Congenital cricopharyngeal achalasia (CCA) is an important but relatively seldom cause of dysphagia resulting from a functional narrowing at the level of the UES^{1,2}. This is a rare entity in childhood and fewer than 50 pediatric cases have been reported¹⁰.

Symptoms of CCA include failure to thrive, regurgitation of food, choking, cyanosis, nasal reflux, coughing and recurrent aspiration pneumonia. These symptoms may develop weeks after birth. The age of initial presentation ranges from birth to six months¹⁻⁶.

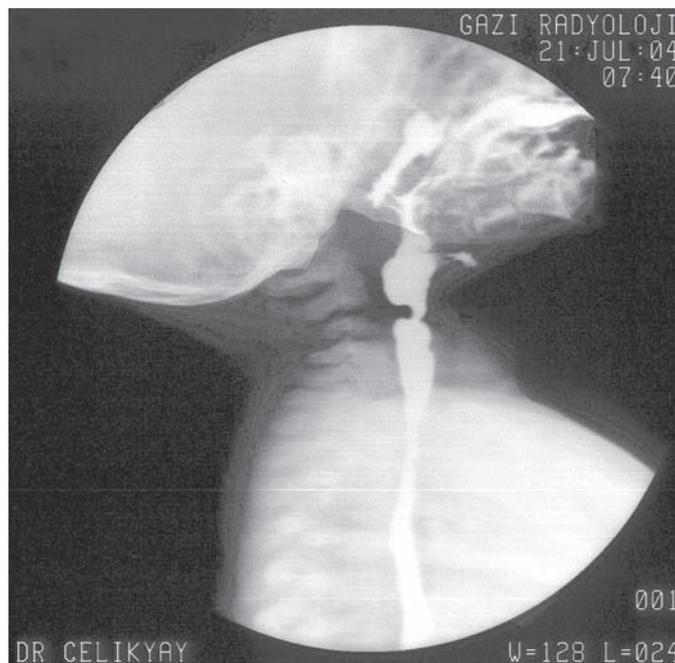


Fig. 1. Posterior narrowing (posterior bar) at the level of 4th-5th cervical vertebrae.

There may be delay in the diagnosis of disease, probably related to the fact that the condition of CCA is not widely recognized and the significance of early feeding difficulties in an affected child may be misinterpreted. In most cases, the symptoms are thought to be related to tracheoesophageal fistula, vascular ring or laryngeal cleft. A thorough history and observation of the patient while feeding are the essential points for diagnosis.

Congenital cricopharyngeal achalasia can be associated with Arnold-Chiari malformation, meningomyelocele or cerebral palsy. In children, however, isolated CCA is an unusual condition⁴.

The diagnostic workup includes an upper gastrointestinal series and manometric study. If there is a clinical suspicion of CCA, the radiologist must be informed. Cine-radiography can show posterior indentation (posterior bar). Aspiration and nasal reflux may be noted. Manometric study reveals the presence of a hypertensive UES, but it is difficult to perform in neonates and infants because the distance from pharynx to cricopharyngeal muscle area is short and the manometric catheter is prone to slip out at swallowing^{1,3,7,8}. For these reasons, video fluoroscopy is sufficient to show disturbance of passage at pharynx and posterior bar in a patient with dysphagia.

Once the diagnosis of CCA is established, definitive treatment can be instituted. Treatment options include bougienage or balloon dilatation, endoscopic botulinum toxin injection or surgical myotomy^{1-6,8-13}. Apparently, no consensus exists among authors for the best management option for CCA. Some authors suggest that myotomy should be kept only for patients who have severe symptoms and/or respiratory compromise or who are unresponsive to other treatment modalities such as balloon dilatation or bougienage. However, others state that with the safe and very effective cricopharyngeus myotomy available, these alternatives cannot be recommended in children because these procedures have to be performed using sedation and general anesthesia and have to be repeated frequently^{1-5,9-11}. Surgical myotomy was performed in our patient. On the third day of the post-operative period, dysphagia and nasopharyngeal reflux had improved dramatically.

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