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CASE REPORT

Primary cricopharyngeal achalasia in a newborn treated by balloon dilatation: A case report and review of the literature

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Summary Primary crycopharyngeal achalasia consists of a failure of the crycopharyngeus muscle to relax at the appropriate time during the swallowing process in the abscence of other motor abnormalities. The principle treatment options for cricopharyngeal achalasia include either dilatation of the upper esophagus or surgical myotomy of the cricopharyngeus muscle. We report a primary cricopharyngeal achalasia case, who was diagnosed early in 1st week of life and treated successfully by balloon dilatation, and discuss the treatment options for this very rare disease. The presented case is the first successfully treated newborn case during the neonatal period in the literature with respect to balloon dilatation.

1. Introduction

Pharyngeal peristalsis propels the bolus of food toward the esophagus, and as the bolus approaches the upper esophageal sphincter (UES), there is a reflex relaxation of the cricopharyngeus muscle, and the bolus passes in to the esophagus. Any anatomic or functional abnormalities at this point may cause swallowing disorder [1]. Primary crycopharyngeal achalasia consists of a failure of the crycopharyngeus muscle to relax at the appropriate time during the swallowing process in the absence of other motor abnormalities [2]. It is a rare condition, in which a newborn presents with dysphagia, choking, salivation and nasal reflux on feeding [3].

The principle treatment options for cricopharyngeal achalasia include either dilatation of the upper esophagus or surgical myotomy of the cricopharyngeus muscle. The efficacy of these treatment methods has not been compared in any series. The long-term success of dilatation as a treatment has been detailed in several reports but many of them were bougienage trials [2,4—6].

We report a primary cricopharyngeal achalasia case who was diagnosed early in 1st week of life and treated successfully by balloon dilatation, and discuss the treatment options for this very rare

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disease. The presented case is the fifth case diagnosed in the neonatal period and, with respect to the treatment modality chosen, is the first successfully treated newborn case by balloon dilatation during the neonatal period in the literature.

2. Case

A 1-day-old female newborn was admitted to the Neonatal Intensive Care Unit for the episodes of regurgitation that began just after the first feeding. She was born from the first pregnancy of a 37-year-old mother and delivered by cesarian section because of late decelerations after an uncomplicated pregnancy. Her birth weight was 3200 g and Apgar scores were recorded as 8 and 10 at 1st and 5th min, respectively. She was roomed-in with her mother just after the birth and bottle feeding was started because of mother's mammoplasty history. After two unsuccessful attempts of feeding, the patient was evaluated for esophagus atresia, and it was ruled out by X-ray. Her clinical presentation with excess salivation and cyanosis during attempts of feeding required further evaluation, and the patient was hospitalized 4 h after the birth.

Physical examination on admission including neurological assessment was unremarkable. A chest X-ray with a nasogastric tube placed into the stomach excluded esophagus atresia. The persistent symptoms at every feeding attempt led us to evaluate the patient for gastroesophageal reflux (GER) and pH monitorization revealed GER. Medical treatment with domperidon and Na arginic acid did not resolve the symptoms. Barium swallow was performed to define whether there was an anatomic or functional obstruction, and it showed almost complete esophageal obstruction at the level of UES, with accumulation of contrast medium in the nasopharynx (Fig. 1A). Manometric study showed mean UES pressure of 60 mmHg, which was two-fold above the upper range of normals (N; 5—25 mmHg). Bronchoscopy under general anestesia was performed to evaluate the patient for other additional anomalies like tracheoesophageal fistula (TOF). It revealed neither an air-way obstruction nor TOF. The esophagoscopy revealed a slight shelf at the level of the UES, which confirmed the diagnosis of cricopharyngeal achalasia.

Balloon dilatation to widen the esophagus was successfully performed with a balloon of 15 mm diameter. It was conducted by a constant low pressure (1—1.5 atm) for 15 s at the level of UES. The patient was followed by a nasogastric tube placed for feeding, and it was removed after 1 week. A second balloon dilatation was performed depending on the barium swallow, which showed consistency of the obstruction. The patient was fed successfully

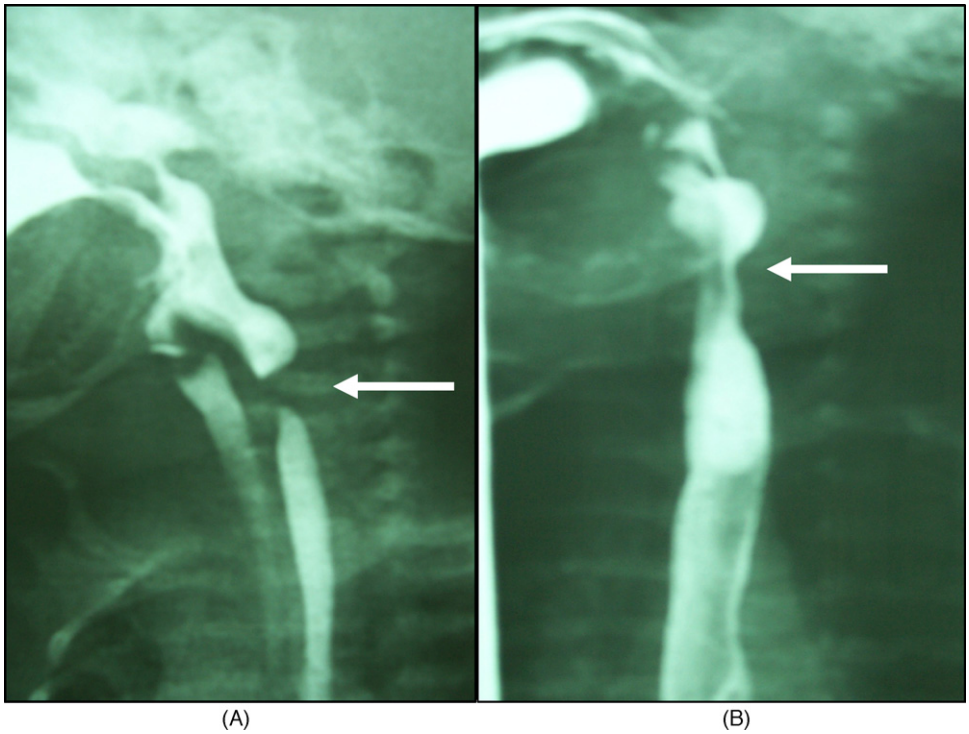

Fig. 1 (A) Left: barium swallow showing almost complete absence of of cricopharyngeal muscle relaxation and filling defect, and (B) right: control barium swallow showing resolution of achalasia after the treatment by balloon dilatation.