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Case Report

Congenital pharyngeal webs: Treatment of a rare clinical entity by endoscopic CO_2 laser approach

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ABSTRACT

Importance: Oesophageal inlet stenosis can promote dysphagia and aspiration. We report the cases of syndromic children with congenital pharyngeal webs successfully treated with endoscopic CO2 laser.
Observations: Pharyngeal webs were excised with CO2 laser (Ultrapulse mode) and resurfaced using mucosal advancement flaps to avoid restenosis and/or formation of secondary synechia. This led to a significant enlargement of the oesophageal inlet, which was documented immediately postoperatively and the clinical improvement of dysphagia and decreased aspiration persisted at distant follow-up.
Conclusion and relevance: Pharyngeal webs are congenital anomalies that can be safely and effectively corrected with endoscopic treatment.

1. Introduction

Pharyngeal webs are extremely rare congenital anomalies characterized by a mucosal band-like extension from the posterior pharyngeal wall anteriorly to the glottis. The first two cases were reported by Gerson et al. in 1983 [1]. Only two subsequent cases were reported thereafter with slight variation in their clinical presentation and management [2,3]. We report two cases of pharyngeal webs associated with other laryngo-tracheo-oesophageal anomalies occurring in syndromic children. We discuss the potential functional and clinical implications of this anomaly, notably dysphagia and aspiration. We also present their endoscopic laser treatment and review the differential diagnosis with the available literature and embryology relevant to this malformation.

2. Report of cases

2.1. Case 1

A 2-year old girl was referred to our airway center for further management of a laryngeal web. She was known for the Frasier's syndrome with heterozygous mutations of exon 15 and exon 74 of the *FRAS1* gene [4]. Frasier syndrome is a rare syndromic entity associated with cryptophtalmia, syndactily, anal stenosis and renal agenesis. After an uneventful postnatal period, the infant developed over the next few months episodes of biphasic stridor that was attributed to laryngotracheitis. The laryngeal web was first noticed during an intubation attempt for abdominal surgery. The infant was subsequently tracheotomized before being referred to our center. The parents mentioned progressive dysphagia characterized by coughing with feeding. After full review of medical history and physical examination, the child was planned for an endoscopic assessment.

During the endoscopy we noted a congenital laryngeal web type 3 (involving 70% of the glottic length) according to Cohen [5], a minor grade II subglottic stenosis according to Myer-Cotton with 60% subglottic obstruction [6], and a severe suprastomal collapse. In addition, there were two pharyngeal webs, extending from the posterior pharyngeal wall to the post-cricoid region with consequent narrowing of the oesophageal inlet (Fig. 1A and B). We decided to transect the webs in order to facilitate bolus passage and diminish secondary aspiration.

The pharyngeal webs were exposed using a Parson spatula in suspension micropharyngoscopy (Fig. 1C) and incised using the carbon dioxide (CO2) laser set in ultrapulse mode, 150 mJ/cm2, 250u spot size and 10Hz repetition rate [7]. These parameters allow for a clean bloodless surgery, yet without charring of the tissues

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List of abbreviations

CO_2	carbon dioxide
LTR	laryngotracheal reconstruction
MPS	suspension micropharyngoscopy
PAHT	pulmonary artery hypertension
VACTER association	vertebral anomalies, anal defect, cardiac anomaly, trachea-oesophageal fistula, renal anomalies
TCA	trichloroacetic acid

(Fig. 1D). The underlying cricopharyngeal muscle was untouched. To avoid re-stenosis and/or synechia, the wound borders were mobilised submucosally and sutured endoscopically in the latero-medial axis with braided absorbable 5-0 Vicryl sutures (Fig. 1E). The child was put on prophylactic regimen of amoxicillin with clavulanic acid for 5 days and discharged the same day.

The parents reported much improved peroral feeding with cessation of coughing episodes. About 6 weeks thereafter, double-stage laryngotracheal reconstruction (LTR) with anterior and posterior costal cartilage grafts was performed. Augmentation by an anterior or posterior midline incision of the cricoid and insertion of cartilage graft was performed in order to expand the laryngo-tracheal airway. As well, suprastomal collapse was corrected and the child was successfully decannulated in the following 2 months. The endoscopic control after 3 months showed a supple and superficial scar healing with an enlarged oesophageal inlet (Fig. 1F).

2.2. Case 2

A 10-year old girl with VACTER association was referred to our center from abroad for persistent trachea-oesophageal fistula after multiple endoscopic repairs of a type III laryngeal cleft [8]. VACTER association is the acronym for vertebral, anal, cardiac, tracheo-oesophageal, and renal anomalies [8]. The child had a tricuspid valve insufficiency with right ventricular hypertrophy with development of severe pulmonary artery hypertension (PAHT). The child had a normal voice, and was fed partially by the mouth and gastrostomy tube in her parent country. She categorically reported blockage while swallowing her saliva and had intermittent moderate degree aspiration of liquids into the airway. The child had been tracheotomised about 18 months prior to her referral to our institution to prevent aspirations. After full medical history and clinical examination, she was planned for an endoscopic assessment. She showed a status post endoscopic repair of the laryngeal cleft with a small distal residual fistula at the level of first tracheal ring - that was treated endoscopically using a cytology brush and 50% trichloroacetic acid. Specifically, the cytology brush was used to de-epithelialized the fistula tract. Together with trichloroacetic acid, application of cytology brush around the fistula tract induce scar formation and closure of the fistula. Similar to case 1, we noticed pharyngeal webs obstructing the oesophageal inlet (Fig. 2A and B). The webs were situated more superiorly and directly in contact with both arytenoids on each side (Fig. 2B). They were mucosal bands not attributing their origin to any iatrogenic cause secondary to previous interventions. The dynamic examination of the vocal cords movement under spontaneous respiration revealed incomplete adduction of the arytenoid cartilages due to the

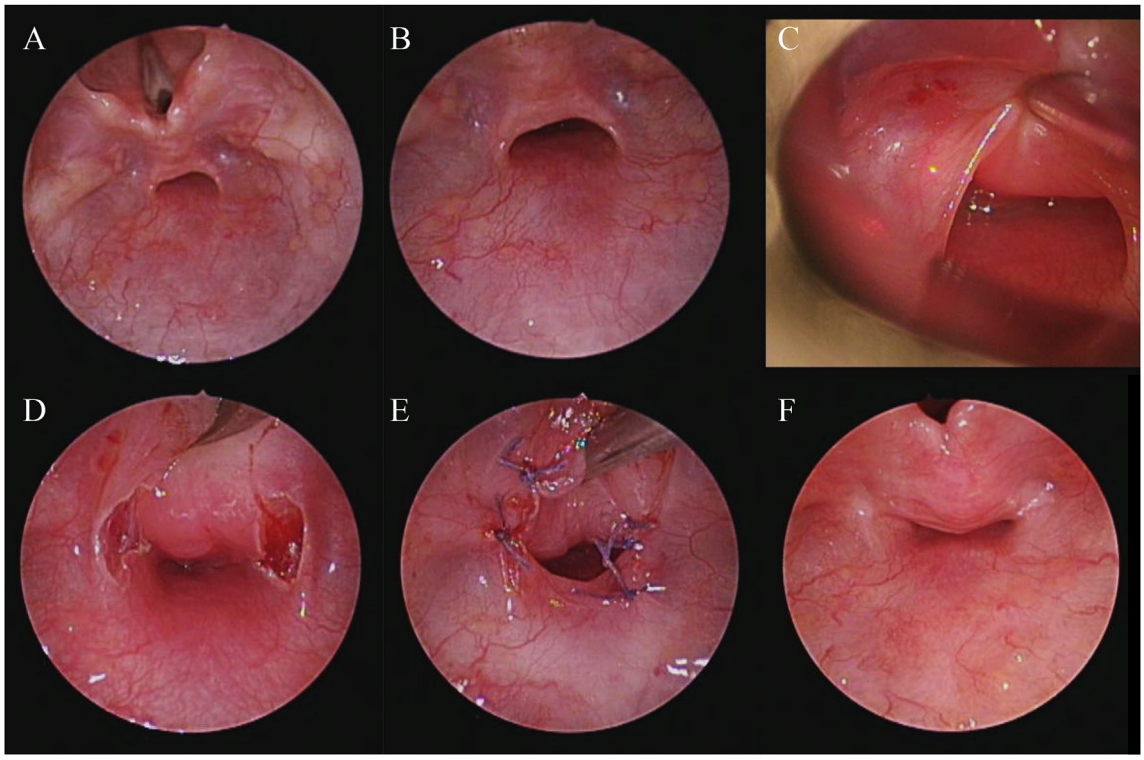

Fig. 1. A: Overview of laryngopharynx with a bare 0-degree 4 mm-diameter sinuscope. Note the concomitant type 3 laryngeal web according to Cohen.
B: Close up showing oesophageal inlet with bilateral web formations in the hypopharyx.
C: Exposition of the left pharyngeal web in suspension micropharyngoscopy (MPS) with a pediatric Parsons' laryngoscope.
D: Immediate postoperative view after laser incision of the pharyngeal webs.
E: Endoscopic suture of mucosal flaps.
F: Endoscopic control at 3 months following decannulation.