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Case Report

Management of cricopharyngeal achalasia in an 8-month child using endoscopic cricopharyngeal myotomy

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ABSTRACT

A term baby was transferred to our tertiary care center with desaturations and inability to manage upper airway secretions. Rigid bronchoscopy and swallowing study revealed cricopharyngeal (CP) achalasia. A gastrostomy tube insertion and Botulinum Toxin-A injection were performed at 6 weeks of age. Improvement of symptoms was observed, however were short-lived requiring recurrent injections. Given the symptom severity, at 8 months, a successful endoscopic CP myotomy was performed. Patient was able to tolerate oral feeds as early as 2 months post-operatively. This is the youngest patient, to our knowledge, treated with endoscopic CP myotomy. Intraoperative pictures and video are presented.

1. Introduction

Cricopharyngeal (CP) achalasia occurs secondary to a poor relaxation of the upper esophageal sphincter leading to an obstruction of the pharyngeal phase of swallowing [1,2]. It is characterized by symptoms of choking, dysphagia and nasopharyngeal regurgitation, poor weight gain and respiratory illnesses secondary to aspiration of poorly managed secretions [2]. CP muscle achalasia diagnosis can be made with barium swallow studies depicting a characteristic “bar” in the region of the CP muscle [2].

The primary management modality of CP achalasia is gastroesophageal reflux(GER) treatment, as GER irritation may aggravate muscle spasms [1–3]. Treatment options for CP muscle achalasia involve endoscopic mechanical dilation techniques [4], as well as endoscopic botulinum toxin(BT) injection into the CP muscle [2,5]. In the first instance, there has been limited benefit described for balloon dilation, with reported recurrence and need for surgical CP myotomy in a significant proportion of cases [6,7]. Furthermore, although BT injection is a minimally invasive technique, it must be tailored appropriately, with its temporary effect resulting in the requirement of multiple treatment injections. It may not suffice for the more severe cases of achalasia [2,5,7].

Surgical options include the open and more recently, endoscopic CP myotomy. The endoscopic approach, being well-established in adults, has also been suggested to be an effective treatment option in the pediatric population. Additional studies are needed, however, to clearly establish its safety. In a study by Chun and colleagues [1], endoscopic CP myotomy was used to treat a 13-month old child who presented with CP achalasia and failed initial management with BT injection.

Recent reports have advocated for earlier treatment of CP achalasia in order to avoid prolonged tube feedings, whether nasogastric or gastrostomy tube(GT), as both fail to provide appropriate oral feeding behaviors leading to poorly developing swallowing mechanisms.

In this case report, we present a child with CP achalasia who underwent endoscopic CP myotomy at the age of 8 months. This is, to our knowledge, now the youngest patient reported to have undergone endoscopic myotomy successfully. This case report will discuss the presentation, management and outcomes of this patient.

2. Methods

This study consists of a case report conducted in the tertiary healthcare center of the Montreal Children's Hospital, a McGill

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University Health Center (MUHC). Ethical approval by the ethics boards (REB) of the MUHC was obtained. After appropriate consent of the patient as well as legal guardians (patients' parents), a review of the patient's medical records, radiological imaging, lab tests, and operative reports was performed. A comprehensive literature review, evaluating endoscopic cricopharyngeal muscle myotomy in the pediatric population was performed.

3. Case report

A term baby boy was transferred to our tertiary care center on the third day of life with a history of desaturations and inability to manage upper airway secretions. The infant was born vigorous with APGAR's 8–9. Shortly after the initiation of feeds, frequent desaturations and duskiness associated to feeding were noted, in addition to significant upper airway secretions, requiring abundant suctioning as well as oxygen (30% O2 via nasal prongs). The otolaryngology team was thus consulted to rule out an upper airway obstruction. Initial evaluation demonstrated a patient with no facial dysmorphisms or retrognathia. Flexible laryngoscopy demonstrated a bilateral mobile vocal cords, pooling of secretions in the supraglottic area with mild laryngomalacia. A rigid bronchoscopy was then performed to evaluate the upper aerodigestive tract and revealed pooling in the post-cricoid region. Inspection of the trachea revealed mild tracheomalacia without any evidence of tracheoesophageal fistula. Further investigations included an abdominal ultrasound which was normal. In addition, a barium esophagogram demonstrated indentation of the left esophagus suggesting an aberrant right subclavian artery with left aortic arch, confirmed with a Computed Tomography as well as echocardiogram.

In order to assess swallowing and aspiration risk, a Videofluoroscopy (VFS) was performed evaluating two textures: clear and nectar-thickened fluids. Findings demonstrated nasopharyngeal reflux, CP spasm, incomplete clearance of contrast from lower pharynx, and laryngeal penetrations and aspiration with both textures (Fig. 1). The patient was deemed unsafe for any oral feeding. Salivagram also demonstrated salivary aspiration in both mainstem bronchi. Brain was normal on MRI.

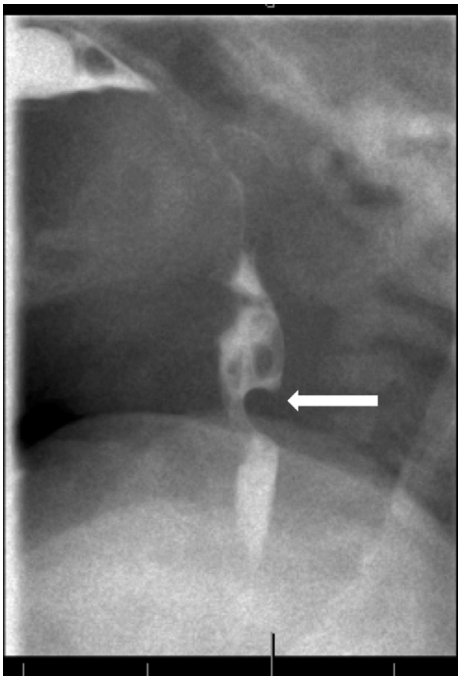

Fig. 1. Preoperative videofluoroscopy (VFS) demonstrating cricopharyngeal bar (white arrow).

The discontinuation of per os (PO) feeding and initiation of naso-duodenal tube feeds resolved the desaturation episodes. The patient, however, continued to show clinical signs of severe GER, requiring prevacid and domperidone. After discussion between our interdisciplinary team, the patient at 6 weeks of age was brought to the operating room for an endoscopic injection of BT-A into the CP muscle. A laparoscopic Nissen fundoplication as well as laparoscopic GT insertion was carried out by General Surgery at the same time. A significant improvement in clinical status with a decreased amount of upper airway secretions following BT injections was reported. A repeat VFS showed no evidence of aspiration, with very few episodes of laryngeal penetration with thin liquids only.

Despite continued use of pro-motility agents and anti-reflux medications, the patient's initial symptoms reappeared 4 months following the injection and continued worsening over time. He was admitted to hospital for respiratory distress, sialorrhea and chocking on his own secretions on two occasions. BT-A was injected to the salivary glands with moderate improvement only. At the age of 8 months, the patient was brought to the OR for endoscopic CO2 laser CP myotomy.

The endoscopic CP myotomy was performed under suspension using the pediatric Weerda diverticular scope. With the suspension in place, the CP muscle was identified. Under microscopic view, the CO2 laser was used (setting of 100 MiliJoules of Energy, 1 Watt). Dissection of the mucosa and CP muscle was carried out layer by layer until the bucco-pharyngeal fascia was encountered and left intact (Fig. 2A–C). The pocket that was formed from the incision of the CP muscle was then filled with FLOSEAL®, a hemostatic matrix to minimize mucosal edge and muscle bleeding (see video in Appendix).

Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.ijporl.2017.07.037.

In the immediate peri-operative course, the patient remained in the recovery room for airway monitoring overnight. He developed one episode of desaturation to 92% which resolved after suctioning accumulated thick nasal secretions. He was also given nebulized epinephrine mask. Tylenol(12.5mg/kg), Advil (10mg/kg) and Morphine (0.05mg/kg) PT were prescribed for post-operative pain management, however patient only required Tylenol during the hospitalization. Post-operative antibiotics were also initiated. Feeds were progressively restarted via G-tube and advanced to the pre-operative flow rates as tolerated. A postoperative gastrograffin study was performed as well to ensure no esophageal perforation was present. Oral feeds were not initiated at this point as the decision was made to withhold oral feeds until oral secretions were deemed to be better managed by the patient.

Once discharged from hospital, the patient was followed by the occupational therapist, the clinical nutritionist as well as GI and ENT. Within three weeks following the CP myotomy, parents began to notice a marked decreased in oro-nasal secretions. Feeds were initially attempted using smooth puree consistencies.

Two months following surgery, the patient demonstrated excellent weight gain with GT and PO purees. A repeat VFS was within normal limits (see Fig. 3). Within a period of 6 months post-operatively, a functional endoscopic evaluation of swallowing demonstrated normal vocal cord movement, with no aspiration or penetration.

4. Discussion

Cricopharyngeal (CP) muscle achalasia is a rare disorder seen in children, which is characterized by the failure of the CP muscle to