

Endoscopic Botulinum Toxin Injection for Infantile Cricopharyngeal Achalasia: Long Term Effects of a Single Procedure - A Case Report

Swathi Y K¹, Shantanu Tandon², Vijay S³

¹Associate Consultant (DLO, DNB ENT), Parivar Super Speciality Hospital, Gwalior

²Senior Consultant (MS, DNB), Sakra World Hospital, Bangalore.

³MD Community Medicine, OIC Station Health Organisation, Air Force Station, Gwalior

Corresponding Author: Swathi Y K

DOI: <https://doi.org/10.52403/ijshr.20230237>

ABSTRACT

Background: The cricopharyngeus muscle forms the Upper Esophageal Sphincter and separates the esophagus from the hypopharynx. Its main function is to prevent reflux of gastric contents causing Laryngo-Pharyngeal Reflux. Achalasia secondary to cricopharyngeal muscle dysfunction is more common in adults, the treatment of which would be a cricopharyngeal myotomy. However when this is encountered in the pediatric age group, the challenge lies in accurately diagnosing it and choosing the appropriate management option. We would like to report one such case of cricopharyngeal spasm causing achalasia in an infant presenting with failure to thrive and aspiration pneumonitis and was diagnosed and treated with endoscopic botulinum toxin injection, a minimally invasive procedure and associated with less morbidity, early recovery times and significant improvement of symptoms. Positive response to the minimally invasive method of Inj. Botulinum Toxin with minimal morbidity can always be reassuring of a successful cricopharyngeal myotomy, if required, at a later date. Hence, a trial of Inj. Botulinum Toxin in an otherwise rare condition with a difficult diagnosis can be a safe approach in the pediatric age group and can help prognosticate the role of second stage cricopharyngeal myotomy, if required.

Objectives: Minimally invasive procedure on an infant with achalasia secondary to cricopharyngeal spasm presenting with failure to thrive and aspiration pneumonia.

Methods: Injecting Botulinum toxin into cricopharyngeal sphincter endoscopically.

Results: Positive response seen in the infant evidenced by weight gain and tolerance to solid feeds.

Conclusion: Positive response to the minimally invasive method of Inj. Botulinum Toxin with minimal morbidity can always be reassuring of a successful cricopharyngeal myotomy, if required, at a later date.

Keywords: Cricopharyngeus, Achalasia, Infant, Endoscopic Botulinum toxin injection

INTRODUCTION

The cricopharyngeus muscle forms the Upper Esophageal Sphincter (UES) and separates the esophagus from the hypopharynx. Its main function is to prevent reflux of gastric contents causing Laryngo-Pharyngeal Reflux (LPR). At times there is hyperfunctioning of the muscle causing distressing symptoms.

Achalasia secondary to cricopharyngeal muscle dysfunction (CPA) is more common in adults, the treatment of which would be a cricopharyngeal myotomy (CPM). In the pediatric age group, the challenge lies in accurately diagnosing it and choosing the appropriate management option. It presents as choking, dysphagia and nasopharyngeal regurgitation, poor weight gain and respiratory illnesses secondary to aspiration of poorly managed secretions. ^[1]

Hyperfunctioning of the cricopharyngeus muscle causing symptoms have been treated in the past with mechanical dilation of UES, pharyngeal plexus neurectomy, and transcervical cricopharyngeal myotomy (CPM). Wound infections, hematoma formation, post-operative respiratory distress, injury to recurrent laryngeal nerve, pharyngo-esophageal diverticulum is some of the complications of transcervical CPM. Endoscopic CPM avoids the above said complications; it is minimally invasive but has a risk of retropharyngeal space violation and infection.

Spontaneous resolution of Cricopharyngeal achalasia has been reported in children upto one year of age and adequately treated with nutritional and supportive management. [2] Botulinum toxin has been successfully used for the treatment of blepharospasm, torticollis, spasmodic dysphonia, and other regional dystonias. Local infiltration of Botox has been used in some cases of CPA but has often required multiple sittings and sometimes a follow-up myotomy. We would like to report one such case of cricopharyngeal spasm causing achalasia in an infant presenting with failure to thrive and aspiration pneumonitis and was diagnosed and treated with endoscopic botulinum toxin injection and followed up for 1 year post op with no recurrence of symptoms.

CASE REPORT

An 11 month old male child was referred to the department of ENT with history of cough on giving feeds since the age of 3.5 months. The child was born at term to a non-consanguinously married couple by a vacuum assisted vaginal delivery and had good APGAR scores. The child was breastfeeding well for about 2.5 months when during the routine visit to the pediatrician, it was noted that he wasn't gaining weight and hence was started on top- feeds. Over a month, he developed aspiration to all oral feeds, had nasal regurgitation of feeds and was diagnosed to have aspiration pneumonia. Oral feeds were

stopped, and Nasogastric (NG) tube was inserted and the child was kept on full feeds through the NG tube. A neurological evaluation proved normal and a brain MRI showed no pathology. Electromyography studies were conducted and proved normal. A videofluoroscopic study was done at an outside institution showed no evidence of aspiration.

The patient was evaluated by the gastroenterology and paediatric department of our institution and an upper GI endoscopy was done which ruled out strictures/ web in the oesophagus. A more detailed evaluation of the videofluoroscopic images showed a subtle narrowing at the upper end of the oesophagus. A detailed ENT evaluation including a fiberoptic laryngoscopy showed normal laryngeal anatomy and function but pooling of saliva in the pyriform fossa and vallecula. The supraglottic and glottis sensations appeared normal.

Based on the videofluoroscopic study and the clinical picture, a collective diagnosis of cricopharyngeal achalasia was made. After counseling the patients parents about the treatment options, given the age, we opted to wait- to improve the nutritional status of the child and to observe for any spontaneous resolution of symptoms.

Over the next 2 months, there was no clinical improvement, so a plan for injection of botulinum toxin was made. Under general anesthesia, a direct laryngoscope was positioned and the cricopharyngeal sphincter was exposed. The type A botulinum toxin was diluted in saline to target dosages and injected at multiple points: A total of 30 IU (3.5IU/kg) of Botulinum toxin was injected into the cricopharyngeus muscle. Post operatively, the vitals were monitored and the child developed transient respiratory distress and decreased breath sounds in both basal lung fields. The child was started on Levosalbutamol and Budesonide nebulisation 2nd hourly with continuous oxygen through nasal prongs. The complaints seemed to settle on day-2 post

op and he was discharged the following day with the naso-gastric tube.

Trial of oral feeds beginning with small amounts of clear fluids was started from post op day 10 and there were no signs of aspiration. Over the next 10 days various liquid feeds with subsequent semisolids were given. The naso-gastric tube was removed on post op day 15. Over the next one week, the child was able to tolerate adequate oral feeds. The child has gained weight adequately (Pre-procedure weight of the child being 9.7Kg and the child weighed 10.8 Kg ,11.3 Kg ,11.5 Kg, 12.2 Kg and 13.5Kg at 1,2 ,3, 4 and 12 months post operatively). Regular telephonic follow ups were taken from the patients' attenders and found good results with adequate feeding.

DISCUSSION

The Upper Esophageal Sphincter (UES) is mainly (but not exclusively) composed of the cricopharyngeus muscle, which has a constant basal tone and causes luminal occlusion at rest, enabling rapid relaxations during swallowing. Cricopharyngeal achalasia is an extremely rare diagnosis in infants, more commonly seen in adults above 60 years of age. Very few cases of pediatric cricopharyngeal achalasia have been reported [3] primarily of idiopathic etiology, and all cases have required surgical intervention at some point of time. Diagnosis in the pediatric age group may often be difficult, radiological finding may be scarce, often subtle and a trial with Inj.Botulinum Toxin in such cases may even assist to underline the diagnosis. Upper esophageal endoscopy usually shows no positive findings in CP dysfunction.

Treatment for this condition is mechanical dilatation of the UES or the cricopharyngeal myotomy which is considered to be the gold standard. Endoscopic Crico-Pharyngeal Myotomy has been the practiced approach according to literature. Botulinum toxin injection of the upper esophageal sphincter has been described for the treatment of dysphagia caused by hypertonicity or faulty relaxation of cricopharyngeal muscle. The

botulinum A neurotoxin, acts at the neuromuscular junction by binding to presynaptic cholinergic receptors and inhibiting the release of acetylcholine which accumulates at the motor-end plate causing paralysis and virtually complete loss of miniature endplate potentials.

The potential role of Botulinum toxin to chemically ablate the cricopharyngeus muscle is appealing, it is rapid acting, minimally invasive, less costly than surgery, and can also help in the identification of patients in whom surgery can be a success. However, the drawback being limited duration of action of a maximum of 3.5 months and needing repeat injections.[4] Difficulties in deciding dosing of Inj. Botulinum Toxin in children is always a concern [5] and the safe recommended dosage (1-20 IU/ Kg body weight) has been reached through a consensus.

Therapeutic effects are usually evident by 3 days after injection and last for nearly 4 months³. Literature says, if the toxin fails to improve cricopharyngeal spasm, then myotomy is also unlikely to be helpful. Although Inj. Botulinum toxin is a minimally invasive technique, it must be tailored appropriately and requires multiple sessions due to its temporary effect; it may not suffice for severe cases of cricopharyngeal achalasia. [5]

CONCLUSION

Injecting Botulinum toxin A into the cricopharyngeus muscle in cases of pediatric CPA is minimally invasive and associated with less morbidity, early recovery times and significant improvement of symptoms. Positive response to the minimally invasive method of Inj. Botulinum Toxin with minimal morbidity can always be reassuring of a successful cricopharyngeal myotomy, if required, at a later date. Hence, a trial of Inj. Botulinum Toxin in an otherwise rare condition with a difficult diagnosis can be a safe and dependable solution in the pediatric age group and can also help prognosticate the role of second stage CPM, if required.

Declaration by Authors

Acknowledgement: None

Source of Funding: None

Conflict of Interest: All authors affirm that there is no actual or potential conflict of interest including any financial, personal or other relationships with other people or organizations that could inappropriately influence, or be perceived to influence, the authors work.

REFERENCES

1. Huoh KC, Messner AH. Cricopharyngeal achalasia in children: indications for treatment and management options. *Curr Opin Otolaryngol Head Neck Surg.* 2013 Dec;21(6):576-80.
2. Seema Khan, Sravan Kumar Reddy Matta. Dysmotility- Upper Esophageal and Upper Esophageal Sphincter Dysmotility (Striated Muscle). In: Robert. M. Kleigman, Joseph W ST Geme III, Karen M Wilson et al, eds. *Nelson Textbook Of Pediatrics.* 21st edition. Canada:Elsevier publications; 2020. p7675

3. Barnes MA, Ho AS, Malhotra PS, Koltai PJ, Messner A. The use of botulinum toxin for pediatric cricopharyngeal achalasia. *Int J Pediatr Otorhinolaryngol.* 2011 Sep;75(9): 1210-4
4. Scholes MA, McEvoy T, Mousa H, Wiet GJ. Cricopharyngeal achalasia in children: botulinum toxin injection as a tool for diagnosis and treatment. *Laryngoscope.* 2014 Jun;124(6):1475-80.
5. Heinen F, Desloovere K, Schroeder AS, Berweck S, Borggraefe I, van Campenhout A, et al. The updated European Consensus 2009 on the use of Botulinum toxin for children with cerebral palsy. *Eur J Paediatr Neurol.* 2010 Jan;14(1):45-66.

How to cite this article: Swathi Y K, Shantanu Tandon, Vijay S. Endoscopic botulinum toxin injection for infantile cricopharyngeal achalasia: long term effects of a single procedure - a case report. *International Journal of Science & Healthcare Research.* 2023; 8(2): 297-300. DOI: <https://doi.org/10.52403/ijshr.20230237>
