Original Research Article

DOI: http://dx.doi.org/10.18203/2349-2902.isj20164480

Evaluation and management of congenital esophageal stenosis: a rare entity

Jiledar Rawat¹, Anand Pandey¹*, Sudhir Singh¹, Sarita Singh²

¹Department of Pediatric Surgery, ²Department of Anesthesia, King George's Medical University, Lucknow, Uttar Pradesh, India

Received: 20 October 2016 **Accepted:** 17 November 2016

*Correspondence: Dr. Anand Pandey,

E-mail: drannad27@rediffmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Background: Congenital esophageal stenosis (CES) is an uncommon condition presenting with swallowing problems. It has been classified into three types. Clinically, the condition is manifested as dysphagia. This is more common after introduction of semisolid to solid foods. We present the management strategy of this uncommon entity. **Methods:** This study ranged from January 2009 to January 2015. Six patients of CES were included in the study. The patients were diagnosed by contrast swallow study and upper gastrointestinal endoscopy. The initial management included esophageal dilation under fluoroscopy guidance. In case of failed dilation, thoracotomy, resection of the stenotic segment and primary esophageal anastomosis was performed. If required, antireflux procedure was added.

Results: In 4 patients, the stenosis was in middle esophagus. Of these, one had associated esophageal diverticulum. In 2 patients, the stenosis was in the lower esophagus. These patients also underwent antireflux surgery. After the surgery, all patients were able to swallow properly. There was no problem in the follow up except in one patient, who needed dilation in the follow up. He responded to the dilation.

Conclusions: High index of suspicion is needed to diagnoses this condition. Dilation may not be successful to treat all type of CES, for whom surgery will be needed. Long term outcome may be satisfactory.

Keywords: Congenital esophageal stenosis, Esophageal dilation, Tracheobronchial remnants

INTRODUCTION

Congenital esophageal stenosis (CES) is a rare anomaly characterized by an intrinsic esophageal narrowing due to an abnormality of the esophageal wall. It is a variant of esophageal atresia (EA). It has been classified into three types: A membranous web or diaphragm, fibro-muscular thickening, stricture secondary to tracheobronchial remnants in the wall of the esophagus. 2

It may present with dysphagia, non-bilious vomiting, and recurrent respiratory tract infection on initiation of solid feeds.³ Since this is a rare anomaly, diagnosis may be delayed and comorbidities in form of repeated chest

infections may ensue. We present the diagnosis strategy and management of CES.

METHODS

This was a retrospective observational study conducted in the Department of Pediatric Surgery of the Medical University. The study period was from January 2009 to January 2016. All patients of CES were included in the study. The hospital records of all patients were evaluated. The patients were diagnosed by contrast swallow study and upper gastrointestinal (UGI) endoscopy. The initial management included esophageal dilation under fluoroscopy guidance. In case of failed dilation, thoracotomy, resection of the stenotic segment and

primary esophageal anastomosis was performed. If required, antireflux procedure was added. The patients were followed in the outpatient department (OPD). Any complication in the follow up was noted.

RESULTS

During the study period of seven years, there were six patients of CES (Table 1). The mean age of patients was

2.62 years (range 9 months- 5 years). Four patients were male and two female.

The complaints noticed were dysphagia mainly to solid food (6, 100%), repeated vomiting (6, 100%), respiratory tract infections (3, 50%), and failure to thrive (2, 33%). Five of them had been treated elsewhere for gastroesophageal reflux. One was later misdiagnosed as achalasia.

Table 1: Profile of patients operated for congenital esophageal stenosis.

Age	Sex	Clinical features	Site of involvement	Duration of hospital stay (days)	Follow up duration (years)
9 months	Male	Dysphagia, vomiting, RTI	Male	11	2
3 years	Male	Dysphagia, vomiting, failure to thrive	Female	12	1
2 years	Female	Dysphagia, vomiting	Female	14	2
5 years	Male	Dysphagia, vomiting, failure to thrive	Male	10	3
2 years	Male	Dysphagia, vomiting, RTI	Male	11	1
3 years	Female	Dysphagia, vomiting, RTI	Male	12	2

RTI- respiratory tract infection.

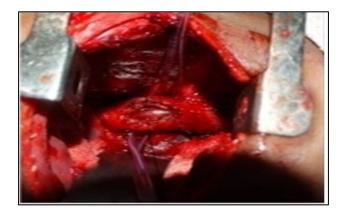


Figure 1: Dilated esophagus.



Figure 2: Stenosed segment being held in stay sutures. It was resected followed by anastomosis.

Contrast swallow study revealed abrupt esophageal tapering in all patients. It was located in mid esophagus in four patients and lower esophagus in remaining two patients. This was further confirmed by UGI endoscopy, which revealed stenosis in the esophagus. It also helped to rule out achalasia. Esophageal dilation provided partial response in two patients; however, none of them was completely relieved of the symptoms after five attempts of dilation. Hence, we considered it a failed dilation.

At the time of operation, the findings of UGI endoscopy were confirmed. In 4 patients, the stenosis was in middle esophagus. Of these, one had associated esophageal diverticulum.

In 2 patients, the stenosis was in the lower esophagus. These patients also underwent Thal's antireflux surgery. Resection of the involved stenotic segment and primary esophageal anastomosis was performed. Post-operative period was uneventful. There was no specific complication. The patients were allowed orally on 7th post-operative day and discharged on 10th day.

Histopathology revealed tracheobronchial remnants in four patients and fibromuscular thickening in two patients. The mean duration of follow up was 1.83 years (range 1-3 years). All patients were able to swallow properly. There was no problem in the follow up except in one patient, who needed dilation in the follow up. He responded to the dilation.

DISCUSSION

The incidence of CES has been reported to be about 1 in 25,000-50,000 live births.⁴ There may be associated anomalies with CES such as esophageal atresia, cardiac anomalies, intestinal atresia, midgut malrotation, anorectal malformations, and hypospadias etc.²

Clinically, the patients may present with progressive dysphagia, nonbilious vomiting, recurrent respiratory tract infection, and failure to thrive. These findings were also noticed in our patients. The condition may be difficult to differentiate from achalasia, gastroesophageal reflux (GER) and its resultant stricture. This was evident in this series also, where patients were inadvertently treated as GER or achalasia. The first investigation is to confirm the obstruction, and contrast swallow study may be helpful. However, there may be a doubt owing to the possibility of achalasia.

Hence, UGI endoscopy provides the information about the presence of stenosis and presence or absence of retained food material seen in achalasia. Besides this esophageal manometry and pH monitoring may be helpful.⁶ High-frequency endoscopic ultrasonography has been reported to be helpful in the diagnosis of CES.⁷ Careful interpretation is important to avoid misdiagnosis.

Esophageal dilation has been claimed to be effective in treating CES. It has been reported to be the most frequently used strategy in children and adults. Improvements in endoscopes and accessories have supported an increase in the number of patients who are conservatively treated with endoscopic dilations (ED) rather than surgical treatment. However, there may be some problems such as no consensus regarding the duration of inflation, problem of the optimum dilator is difficult to solve, because of the different esophageal size during the pediatric age, no consensus regarding the interval between repeated ED with either a balloon or a bougie etc. §

As noted in this study, the experience with esophageal dilation was not satisfactory and all patients underwent surgical intervention. Though it has been advocated to treat lower third of stenosis by laparotomy and middle third by thoracotomy, study were able to treat the lower third of stenosis by thoracotomy. Rather than myotomy, we performed resection of the stenotic segment as this was likely of a completely cure. Leaving the stenotic part had a possibility of recurrence. Since all of our patients had a satisfactory outcome in the follow up, our claim appears to be validated.

However, it is to be noticed that none of our patient has membranous variant, which may be more likely to be treated by dilation. ⁹⁻¹¹

CONCLUSION

CES is an uncommon congenital anomaly of the esophagus, which may be misdiagnosed. High degree of suspicion, proper evaluation, and appropriate treatment may provide optimal outcomes.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

REFERENCES

- Amae S, Nio M, Kamiyama T, Ishii T, Yoshida S, Hayashi Y. Clinical characteristics and management of congenital esophageal stenosis: a report on 14 cases. J Pediatr Surg. 2003;38:565-70.
- Fekete NC, Backer A, Jacob LS, Pellerin D. Congenital esophageal stenosis. Pediatr Surg Int. 1987;2:86-92.
- 3. Jain V, Yadav DK, Sharma S, Jana M, Gupta DK. Management of long segment congenital esophageal stenosis: A novel technique. J Indian Assoc Pediatr Surg. 2016;21:150-2.
- Valerio D, Jones PF, Stewart AM. Congenital oesophageal stenosis. Arch Dis Child. 1977;52:414-6.
- Ramareddy RS, Alladi A. Review of esophageal injuries and stenosis: lessons learn and current concepts of management. J Indian Assoc Pediatr Surg. 2016;21(3):139-4.
- 6. Harmon CM, Coran AG. Congenital anomalies of the esophagus. In: Coran AG, Adzick NS, Krummel TM, Laberge J, Shamberger RC, Caldmone AA (editors). Pediatric Surgery. 7th ed. Philadelphia: Elsevier Saunders; 2012:893-918.
- Usui N, Kamata S, Kawahara H, Sawai T, Nakajima K, Soh H, et al. Usefulness of endoscopic ultrasonography in the diagnosis of congenital esophageal stenosis. J Pediatr Surg. 2002;37:1744-6.
- Dalloglio L, Caldaro T, Foschia F, Faraci S, Abriola GF, Rea F et al. Endoscopic management of esophageal stenosis in children: New and traditional treatments. World J Gastrointest Endosc. 2016;8:212-9
- Grabowski ST, Andrews DA. Upper esophageal stenosis: two case reports. J Pediatr Surg. 1996;31:1438-9.
- 10. Feng FH, Kong MS. Congenital esophageal stenosis treated with endoscopic balloon dilation: report of one case. Acta Paediatr Taiwan. 1999;40:351-3.
- 11. Kurian JJ, Jehangir S, Varghese IT, Thomas RJ, Mathai J, Karl S. Clinical profile and management options of children with congenital esophageal stenosis: a single center experience. J Indian Assoc Pediatr Surg. 2016;21:106-9.

Cite this article as: Rawat J, Pandey A, Singh S, Singh S. Evaluation and management of congenital esophageal stenosis: a rare entity. Int Surg J 2017:4:408-10.