Cricopharyngeal achalasia – a rare cause of dysphagia

Raman Wadhera · S. P. Gulati · Ajay Garg · Anju Ghai

Abstract Cricopharyngeal achalasia is a rare clinical entity in which upper esophageal sphincter (cricopharyngeus) does not open adequately during swallowing leading to dysphagia. Barium swallow reveals a smooth posterior impression of cricopharyngeus at C6 level. We report cricopharyngeal achalasia in a 68 year male who was managed with dilatation.

Keywords Cricopharyngeal achalasia · Barium swallow · Cricopharyngeal myotomy

Introduction

Cricopharyngeal achalasia is a condition in which the upper esophageal sphincter (cricopharyngeus) does not open adequately during swallowing. Unlike the other muscles of pharyngeal constriction, the cricopharyngeus remains contracted between swallows, acting as an upper esophageal sphincter. It normally relaxes during deglutition to allow the passage of the food bolus. When the muscle fails

to completely relax, it can lead to dysphagia. Severity of dysphagia may be related to the degree of cricopharyngeal abnormality. The patients are usually above 40 years of age and most commonly over 60 [1]. Rarely, it may be present in infants also [2]. The patient complains that it takes two or more swallows to force the food out of the mouth or in severe cases, no matter how much the food is chewed, it goes “round and round but won’t go down”. Other symptoms like retrosternal burning, feeling of lump in throat, choking spells, aspiration and weight loss may be present. Barium swallow film shows a horizontal indentation or a bar on the posterior wall of a barium filled pharyngoesophageal area, at the level of C6 vertebra, caused by the unrelaxed cricopharyngeus muscle. Unlike most retropharyngeal masses, the cricopharyngeus does not exceed one cm in vertical dimension [3]. Endoscopic examination shows redundant folds of mucous membrane. The cricopharyngeus muscle itself is unduly prominent as a transverse ridge, which may or may not relax readily when touched with the tip of the esophagoscope. The raising of this ridge and the narrowing of the lumen by the contracted muscle gives a characteristic appearance at the entrance to the esophagus [1]. Manometric studies may help in showing the incoordination at the cricopharyngeal area. We are reporting a rare case of cricopharyngeal achalasia.

Case report

A 68-year-old male presented with the history of dysphagia to solids for last 10 years. The dysphagia varied in degree and at times he was free of symptoms for several days. There was no history of weight loss and general physical examination was normal. His haemoglobin was 14gm%. Barium swallow revealed a smooth posterior impression at the level of C6 vertebra (Fig. 1). Esophagoscopy was done under general anesthesia and it revealed redundant pharyngeal folds of mucosa with evidence of narrowing of the

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Fig. 1 Barium swallow showing unreleased cricopharyngeus muscle producing a prominent defect at C6 vertebra

cricopharyngeus yielding to firm pressure by the esophagoscope. Patient was discharged after one day. The patient is in our follow up for the last six months and he is having satisfactory relief from the symptoms of dysphagia.

Discussion

The reason for the lack of complete relaxation of the cricopharyngeus muscle is not fully understood. It may be because of either incomplete relaxation, paradoxical contraction, spasm of cricopharyngeus muscle or fibrosis and shortening of the cricopharyngeus muscle [4]. There may be degeneration of cricopharyngeus muscle with consequent loss of elasticity [5]. Gastroesophageal reflux disease can also lead to cricopharyngeal spasm.

The most frequent cause of cricopharyngeal achalasia is cerebrovascular disease. Other causes include pseudobulbar palsy, nasopharyngeal carcinoma, poliomyelitis, thyroid myopathy, cervical vagotomy, polymyositis, dermatomyositis, oculopharyngeal syndrome, amyotrophic

lateral sclerosis and hiatus hernia, but many cases are idiopathic [3]. We could not find any cause in our patient. Cricopharyngeal achalasia has been implicated in the development of Zenker's diverticula [3].

Botulinum toxin injections into the cricopharyngeus muscle have been used with limited success [2]. A trial of antireflux treatment may be given. Dilatation provides only temporary relief so cricopharyngeal myotomy is the treatment of choice in severe cases [4, 6]. Cricopharyngeal myotomy can be done either by lateral pharyngotomy approach or by endoscopic laser assisted technique using KTP-532 or CO₂ laser.

In our case, as the general condition of the patient was good, so only dilatation was done with rigid esophagoscope. Cricopharyngeal myotomy was not advised because symptoms were not severe enough to warrant operation.

References

1. Sutherland HD (1962) Cricopharyngeal achalasia. *J Thoracic & Cardiovas Surg* 43(1):114–126
2. Sewell RK, Bauman NM (2005) Congenital Cricopharyngeal Achalasia Management With Botulinum Toxin Before Myotomy. *Arch Otolaryngol Head Neck Surg* 131: 451–453
3. Branstetter BF (2004) Diagnostic imaging of the pharynx and oesophagus. In: Cummings CW, editor. *Otolaryngology Head and Neck Surgery*. Vol. 4. 4th edition. Elsevier Mosby: USA. pp.1785–1824
4. Goyal RK (1984) Disorders of the cricopharyngeus muscle. In: Fried MP, editor. *The Otolaryngologic Clinics of North America*. Vol. 17. WB Saunders Company: Philadelphia. pp.115–130
5. Russell RCG, Williams NS, Bulstord CJK (2004) Bailey and Love's. *Short Practice of Surgery*. 24th edition. Edward Arnold (Publishers) Ltd: London. pp. 989–1025
6. St. Guily J, Zhang KX, Perie S (1995) Improvement of dysphagia following cricopharyngeal myotomy in a group of elderly patients. *Ann Otol Rhinol Laryngol* 104:603–609