CASE REPORT

Zenker's diverticulum complicating achalasia: a 'cup-and-spill' oesophagus

Mark Sayles, Laura Harrison, Julian A McGlashan, David G Grant

Department of Otolaryngology—
Head and Neck Surgery,
Nottingham University Hospitals
NHS Trust, Nottingham, UK

Correspondence to
Mark Sayles,
sayles.m@gmail.com

SUMMARY

A 72-year-old woman presented with long-standing gastro-oesophageal reflux, regurgitation of swallowed food and worsening cervical dysphagia. Fluoroscopic barium oesophagography revealed a posterolateral pharyngeal pouch (Zenker's diverticulum (ZD)) complicating a 'cup and spill' oesophageal deformity with a smoothly tapered segment at the gastro-oesophageal junction. CT and high-resolution manometry confirmed that the underlying abnormality was a massively dilated oesophagus with aperistalsis and pan-oesophageal pressurisation, consistent with a diagnosis of oesophageal achalasia (type II). She underwent endoscopic stapled diverticulotomy, with good symptomatic relief. We discuss the aetiology of ZD, its management and the association here with oesophageal achalasia.

BACKGROUND

Zenker's diverticulum (ZD) is a relatively rare cause of dysphagia typically seen in elderly patients. The precise aetiology of these diverticulae remains uncertain. We found a diverticulum in the presence of an abnormally dilated oesophagus. Radiological investigation with fluoroscopic barium oesophagography and CT imaging, combined with high-resolution oesophageal manometry, supported a diagnosis of underlying type II oesophageal achalasia. The high intrabolus pressures generated in the oesophagus secondary to the non-relaxing lower oesophageal sphincter may have pre-disposed the patient to the development of a diverticulum at Killian's dehiscence. In the setting of achalasia we successfully managed the ZD with endoscopic stapled diverticulotomy.

CASE PRESENTATION

A 72-year-old Caucasian woman presented to the otolaryngology clinic at our academic tertiary referral centre with increasing symptoms of regurgitation of swallowed food and cervical dysphagia 9 months ago. She had previously been under the care of the otolaryngologists at another institution, where she was diagnosed with ZD on the basis of her symptoms and findings at barium oesophagography. She denied any weight loss or halitosis. She had undergone an unsuccessful attempt at endoscopic stapled diverticulotomy 5 years ago. This procedure was abandoned as the anatomy of the diverticulum was not favourable for an endoscopic approach at that time. Her medical history was otherwise unremarkable, with no history of abdominal, thoracic or neck surgery. She took regular

antihypertensive medications only. Clinical impression on her initial assessment at this clinic was of progression of her diverticulum. Fluoroscopic barium oesophagography was therefore arranged as the initial investigation to assess size and position.

INVESTIGATIONS

Pre-operative fluoroscopic barium oesophagography with the patient in a seated position demonstrated highly abnormal appearances of the oesophagus. There was a predominantly right-sided ZD at the level of C5–6 vertebrae. This filled with contrast, which then partially emptied into a grossly dilated oesophagus with multiple fluid levels demonstrated as the contrast solution cascaded downwards over folds of oesophageal mucosa (figure 1A–C, video 1). Radiologically this was described as a 'cup-and-spill' appearance, similar to that seen in the rare normal variant of gastric anatomy where contrast first fills the gastric fundus before spilling over into the gastric body and antrum. There was no hold-up of contrast in the oesophagus. However, the lower oesophagus showed a smoothly tapered segment. On the basis of these results the patient was listed for oesophagocopy and endoscopic stapled diverticulotomy.

Endoscopic examination under general anaesthesia showed a right-sided posterolateral diverticulum, a thick cricopharyngeal bar, profuse gastric regurgitation and a grossly dilated oesophagus with mucosal appearances suggestive of oesophagitis. Stapled division of the cricopharyngeal bar was successfully performed and mucosal biopsies were taken from the lower oesophagus (figure 1D–F). A histological analysis of these specimens showed chronic inflammatory changes and fungal hyphae with appearances suggestive of candida oesophagitis. There was no evidence of dysplasia or malignancy.

Postoperatively the patient recovered well and was discharged home the following day. On review in the outpatient clinic 2 weeks later she reported significant improvement in her symptoms. To assess the underlying abnormality in the oesophagus she was referred for CT as she was reluctant to undergo flexible oesophagogastrroduodenography. Cross-sectional imaging confirmed successful endoscopic stapling of the diverticulum, and showed a massively dilated oesophagus filled with food residue (with the patient supine) and a normal stomach in the abdomen (figure 1G). There was no evidence of malignancy.

On high-resolution oesophageal manometry (postoperatively) the upper oesophageal sphincter showed normal basal pressure, and normal

CrossMark

To cite: Sayles M, Harrison L, McGlashan JA, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-200702

Figure 1 (A–C) Fluoroscopic barium oesophagography; oblique images. A right-sided diverticulum measuring approximately 5×4 cm (p) is demonstrated at C5/6 vertebral level and fills with contrast, with overflow into a grossly dilated oesophagus with multiple fluid levels overflowing distally ('cup-and-spill' pattern). There is pronounced smooth tapering at the gastro-oesophageal junction (arrow). (D–F) Photographs at endoscopic stapling of Zenker's diverticulum showing dilated oesophagus with mucosal folds (D), and endoscopic stapled division of the cricopharyngeal bar (E and F). (G) Coronal average IP reconstruction of CT of the thorax following endoscopic intervention. The oesophagus remains grossly dilated and filled with food residue with the patient lying supine in the scanner.

relaxation and opening. The oesophagus showed aperistalsis and pan-oesophageal pressurisation with water swallows with obstruction to flow with multiple swallows (figure 2). The gastro-oesophageal junction could not be crossed due to gross anatomical abnormality. These findings are consistent with type II oesophageal achalasia according to the Chicago classification (table 1).^{1 2}

DIFFERENTIAL DIAGNOSIS

Given the presenting symptom of dysphagia to both solids and liquids, the differential diagnosis includes all causes of an oesophageal motility disorder—the most likely being either achalasia or diffuse oesophageal spasm. Other causes of oesophageal dysmotility to consider include systemic disorders such as scleroderma, neurological disorders such as multiple sclerosis and motor neuron disease, and Chagas disease (a specific form of achalasia due to infection with the protozoan *Trypanosoma cruzi*, endemic in areas of central and south America). On detailed history and examination the patient exhibited no other features suggestive of these diagnoses. The added feature of regurgitation of swallowed food and the prior demonstration of ZD at her previous diagnostic work-up suggested the possibility of either concomitant disease, or an atypical presentation of ZD.

TREATMENT

The patient has been maintained on medical treatment with a proton-pump inhibitor and sodium alginate suspension for symptom control. She underwent endoscopic stapled division of the cricopharyngeal bar as definitive management of the ZD. In view of her dilated oesophagus, the patient was pre-operatively warned of the significant risk of worsening her reflux symptoms after division of cricopharyngeus muscle.

OUTCOME AND FOLLOW-UP

The patient was initially followed up 2 weeks after endoscopic treatment. She reported relief from her dysphagia and food regurgitation symptoms. However, she remained troubled by significant gastro-oesophageal reflux but did not perceive this to have subjectively worsened. Since then she has been seen at three monthly intervals, and referred to colleagues in gastroenterology for further management of achalasia. She is currently under further investigation by gastroenterology. Following a planned oesophagogastroduodenoscopy she will be offered definitive treatment.

DISCUSSION

ZD is a rare cause of high dysphagia typically seen in elderly patients with an annual incidence of 2/100 000 in the UK.³ Anatomically, ZD is a mucosal pouch which develops in the dorsal wall of the most caudal part of the hypopharynx in an area of relative weakness ('Killian's dehiscence') between the oblique fibres of the thyropharyngeus muscle above and the horizontal fibres of the cricopharyngeus muscle below.⁴ The precise aetiology is not known, although several theories exist. ZD may be the result of age-related reduced tissue elasticity, or secondary to increased tone in the upper oesophageal sphincter.⁵ Cook *et al*^{6 7} suggested these diverticulae result from reduced cricopharyngeal compliance. Manometry showed normal relaxation but inadequate opening of the cricopharyngeus. This theory holds that the resultant high intrabolus swallowing pressures within the pharynx leads to pulsion diverticulum formation. Surgery for ZD is now commonly via the endoscopic route, either with stapled division or carbon dioxide laser division of the cricopharyngeal bar. Compared with traditional open approaches to diverticulectomy and cricopharyngeal myotomy, endoscopic techniques are safer, quicker and have a shorter recovery time.⁵

Oesophageal achalasia is a rare disorder of oesophageal motility with an uncertain aetiology.⁸ Annual incidence rates are typically 0.5–1.0/100 000 in northern Europe.⁹ Diagnostic testing

Video 1 Fluoroscopic barium oesophagography; movie showing the multiple "cup" and "spill" levels in the thoracic oesophagus. The contrast initially fills the Zenker's diverticulum and then cascades into the dilated lower oesophagus over multiple folds of mucosa. There is no holdup of contrast at the gastro-oesophageal junction, with passage into the stomach via a narrow tapered segment.

Table 1 Chicago achalasia classification based on high-resolution manometry findings^{1 2}

Achalasia classification	Pattern of oesophageal body contractility
Type I	No significant pressurisation
Type II	Rapidly propagated pressurisation, localised to distal oesophagus or across entire length
Type III	Pressurisation attributable to spastic contraction

often involves endoscopy to rule out mechanical obstruction, and fluoroscopic barium oesophagography. However, manometry is considered the gold-standard for diagnosis.^{10 11} Important diagnostic features are absent or incomplete lower oesophageal sphincter relaxation in response to swallowing on manometry, with aperistalsis in the oesophageal body, and characteristic appearances such as a 'bird's beak' lower oesophageal tapering on barium oesophagography. Evidence suggests achalasia is the result of a loss of inhibitory nitrergic neurotransmission secondary to loss of myenteric nerve cells, which is often accompanied by a lymphocytic inflammatory infiltrate.⁸ The underlying aetio-pathogenesis however remains uncertain. Theories include those based on infectious, autoimmune and hereditary causes.^{8 12-14}

Management of achalasia is broadly classified as medical and surgical. Medication used to relax the lower oesophageal sphincter includes oral calcium channel blockers such as nifedipine, and nitrates such as sublingual isosorbide dinitrate.¹⁵ These are not without significant side effects such as pedal oedema, headaches and hypotension are therefore tend to be reserved for patients unable to undergo more definitive treatment. Botulinum toxin (BoTox) has been used with some success, although the response to a single injection into the lower oesophageal sphincter decreases over time.¹⁶ Surgical treatments include endoscopic, laparoscopic and open surgical approaches. Endoscopic treatment has focused on pneumatic dilatation,¹⁷ although recently peroral oesophageal myotomy has been developed as an alternative endoscopic technique.¹⁸ Overall, a laparoscopic Heller myotomy with fundoplication offers the best short-term and long-term functional results in achalasia, although it is comparable to pneumatic dilatation in the short term.^{11 17}

Figure 2 High-resolution oesophageal manometry results. There is pan-oesophageal pressurisation on repeated swallowing of liquid. Lower oesophageal sphincter was not crossed due to the gross anatomical abnormality of the thoracic oesophagus.

The pan-oesophageal pressurisation despite the concomitant dilatation seen on the manometry in our patient is a good prognostic indicator for a successful outcome following either medical, endoscopic or laparoscopic management of achalasia.^{1–19} Recent data from the European Achalasia Trial indicates superiority of endoscopic pneumatic dilatation over laparoscopic Heller myotomy in the management of type II achalasia. However, both strategies have excellent success rates in this setting.¹⁹

A search of the PubMed and EMBASE electronic databases using the keywords 'achalasia' and either 'Zenker's diverticulum' or 'pharyngeal pouch' revealed only one previous report of concomitant oesophageal achalasia and ZD.²⁰ Jones *et al*²⁰ previously described two patients with oesophageal achalasia in whom the ZD was identified on barium oesophagography. It is important to remain cognisant of the distinction between oesophageal achalasia and what is often termed cricopharyngeal achalasia, that is functional obstruction at the level of the upper oesophageal sphincter due to failure of relaxation of the cricopharyngeus. We are aware of only one previous report of concomitant oesophageal achalasia and Zenker's diverticulum,²⁰ although the association of ZD with cricopharyngeal achalasia is well known. While it remains possible that the association between oesophageal achalasia and ZD here is coincidental, there is some evidence to suggest a causative association. Changes in the structure and function of the upper oesophageal sphincter occurring secondary to incomplete lower oesophageal sphincter relaxation and increased pressure in the oesophageal body have been shown to lead to the development of a prominent cricopharyngeal bar.^{21–22} This or similar mechanisms may have predisposed the patient described in this report to develop symptomatic ZD. After endoscopic stapled division of the cricopharyngeal bar the patient noticed a significant improvement in symptoms.

Learning points

- This case illustrates the importance of preoperative barium oesophagography in the assessment of dysphagia. Careful radiological examination can reveal complex pathologies such as the combination of Zenker's diverticulum and oesophageal achalasia.
- High intrabolus pressures in oesophageal achalasia may predispose to the development of Zenker's diverticulum.
- Endoscopic stapled diverticulotomy for Zenker's diverticulum in the presence of oesophageal achalasia is a feasible management strategy which does not preclude further endoscopic or surgical intervention to treat the underlying oesophageal achalasia.

Acknowledgements The authors would like to thank Dr Timothy Taylor for his expert radiological opinion.

Contributors JAM and LH investigated and operated on the patient. MS and DGG wrote the first draft of the manuscript and produced the figures and video. All authors were involved in writing and editing the final version of the manuscript text. All authors approved the final edited version of the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Pandolfino JE, Kwiatek MA, Nealis T, *et al*. Achalasia: a new clinically relevant classification by high-resolution manometry. *Gastroenterology* 2008;135:1526–33.
- 2 Pandolfino JE, Ghosh SK, Rice J, *et al*. Classifying esophageal motility by pressure topography characteristics: a study of 400 patients and 75 controls. *Am J Gastroenterol* 2008;103:27–37.
- 3 Bradley PJ, Kochaar A, Quraishi MS. Pharyngeal pouch carcinoma: real or imaginary risks? *Ann Otol Rhinol Laryngol* 1999;108(11 Pt 1):1027–32.
- 4 Van Overbeek JJM. Meditation on the pathogenesis of hypopharyngeal (Zenkers) diverticulum and a report of endoscopic treatment in 545 patients. *Ann Otol Rhinol Laryngol* 1994;103:178–85.
- 5 Verhaegen VJ, Feuth T, van den Hoogen FJ, *et al*. Endoscopic carbon dioxide laser diverticulostomy versus endoscopic staple-assisted diverticulostomy to treat Zenker's diverticulum. *Head Neck* 2011;33:154–9.
- 6 Cook IJ, Blumbers P, Cash K, *et al*. Structural abnormalities of the cricopharyngeus muscle in patients with pharyngeal (Zenker's) diverticulum. *J Gastroenterol Hepatol* 1992;7:556–62.
- 7 Cook IJ, Gabb M, Panagopoulos V, *et al*. Pharyngeal (Zenker's) diverticulum is a disorder of upper esophageal sphincter opening. *Gastroenterology* 1992;103:1229–35.
- 8 Kraichely RE, Farrugia G. Achalasia: physiology and etiopathogenesis. *Dis Esophagus* 2006;19:213–23.
- 9 Birgisson S, Richter JE. Achalasia in Iceland, 1952–2002: an epidemiologic study. *Dig Dis Sci* 2007;52:1855–60.
- 10 Patti MG, Herbella FA. Achalasia and other esophageal motility disorders. *J Gastrointest Surg* 2011;15:703–7.
- 11 Dobrowolsky A, Fisichella PM. The management of esophageal achalasia: from diagnosis to surgical treatment. *Updates Surg* 2013;2. Published Online First: Epub Date. doi: 10.1007/s13304-013-0224-1.
- 12 Kimber J, McLean BN, Prevett M, *et al*. Algrove or 4 'A' syndrome: an autosomal recessive syndrome causing multisystem neurological disease. *J Neurol Neurosurg Psychiatry* 2003;74:654–7.
- 13 Robertson CS, Martin BA, Atkinson M. Varicella-zoster virus DNA in the oesophageal myenteric plexus in achalasia. *Gut* 1993;34:299–302.
- 14 Wong RK, Maydonovitch CL, Metz SJ, *et al*. Significant DQW1 association in achalasia. *Dig Dis Sci* 1989;34:349–52.
- 15 Vaezi MF, Richter JE. Current therapies for achalasia: comparison and efficacy. *J Clin Gastroenterol* 1998;27:21–35.
- 16 Bassotti G, Annese V. Review article: pharmacological options in achalasia. *Aliment Pharmacol Ther* 1999;13:1391–6.
- 17 Weber CE, Davis CS, Kramer HJ, *et al*. Medium and long-term outcomes after pneumatic dilation or laparoscopic Heller myotomy for achalasia: a meta-analysis. *Surg Laparosc Endosc Percutan Tech* 2012;22:289–96.
- 18 Inoue H, Minami H, Kobayashi Y, *et al*. Peroral endoscopic myotomy (POEM) for esophageal achalasia. *Endoscopy* 2010;42:265–71.
- 19 Rohof WO, Salvador R, Annese V, *et al*. Outcomes of treatment for achalasia depend on manometric subtype. *Gastroenterology* 2013;144:718–25.
- 20 Jones B, Donner MW, Rubesin SE, *et al*. Pharyngeal findings in 21 patients with achalasia of the esophagus. *Dysphagia* 1987;2:87–92.
- 21 Zhang ZG, Diamant NE. Repetitive contractions of the upper esophageal body and sphincter in achalasia. *Dysphagia* 1994;9:12–19.
- 22 Jones B. Pharyngo-oesophageal interrelationships and reflexes involved in airway protection. In: Jones B, ed. *Normal and abnormal swallowing: imaging in diagnosis and therapy*. Springer, 2002:91–102.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow