The Journal of Laryngology & Otology
October 2000, Vol. **114,** pp. 805–807

Pharyngeal pouch and polymyositis: association and implications for aetiology of Zenker's diverticulum

CHRISTOS GEORGALAS, M.D., SIMON T. BAER, F.R.C.S.(ORL)*

Abstract
A case of pharyngeal (Zenker's) pouch in a patient suffering from polymyositis is presented. Although dysphagia is a recognized manifestation of polymyositis, in this unique case it was caused by a pharyngeal pouch. The aetiology of Zenker's diverticulum is discussed in the light of this unexpected association.

Key words: Zenker's Diverticulum; Polymyositis; Deglutition Disorders

Case report

In 1983, a 63-year-old woman, recently diagnosed with polymyositis, was evaluated for mild dysphagia. A barium swallow performed indicated a narrowing of the oesophagus at the C6–C7 level, largely symmetrical and smooth, arising posteriorly. A direct pharyngo-oesophagoscopy arranged to rule out a postcricoid neoplasm revealed no obvious abnormality.

Over the next few years, she was followed up at irregular intervals for her dysphagia, that did not significantly affect her quality of life. Her main disease (polymyositis) had a wavering course, and necessitated the addition of methotrexate and the progressive tapering of prednisolone. Her levels of creatinine kinase returned to normal after the first year of therapy, and there was a progressive but definite improvement of her symptoms.

In 1997 she was referred again to the ENT clinic for increasing difficulty in swallowing. This time she described moderate to severe intermittent difficulty swallowing solids, especially large tablets and certain foods. She also described occasional episodes of choking but no delayed or significant regurgitation. Fibre-optic pharyngolaryngoscopy showed no evidence of pooling and both her pharynx and larynx were healthy. A barium swallow (Figure 1) confirmed that she had a medium sized pharyngeal pouch, but at this stage, weighing up the risks and benefits, surgery was deferred.

Over the next two years she was followed up at regular intervals at the ENT clinic for her swallowing problems, until in June 1999 her dysphagia reached a level where, in spite of her multiple concomitant problems, a decision was taken to undertake endoscopic stapling of the pouch, which took place in September 1999. Her immediate postoperative course was uneventful.

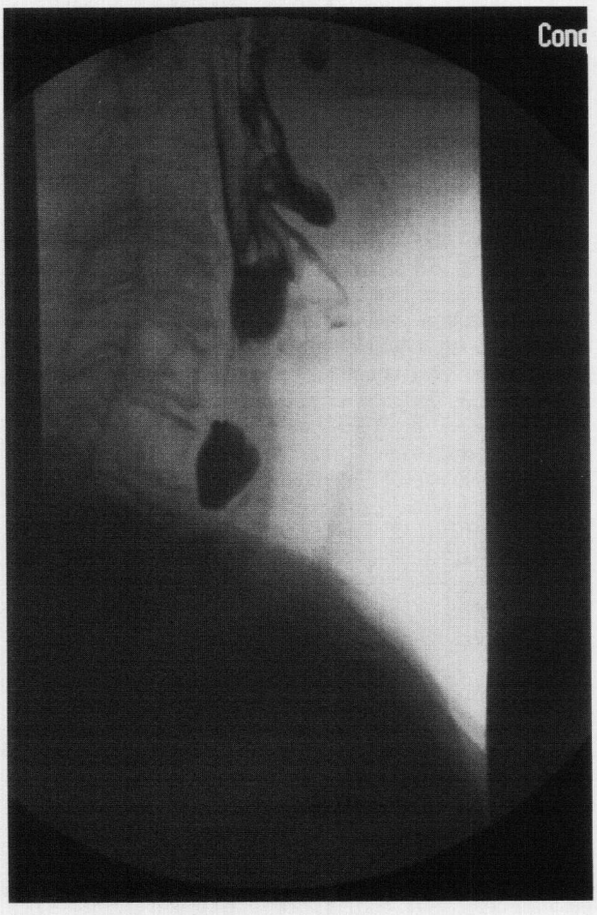

FIG. 1
Barium swallow of oesophagus, showing moderately sized Zenker's diverticulum.

Discussion

Since the first description of pharyngoesophageal diverticulum by an English surgeon[1] many theories have been proposed regarding its pathogenesis. In the first series of 27 patients the diverticulum was described as a protrusion of

From the Department of Otolaryngology, St Mary's Hospital, London and The *Conquest Hospital, East Sussex, UK.
Accepted for publication: 19 May 2000.

mucosa and submucosa through the fibres of the oesophageal muscularis propria.[2] Today we know that this diverticulum develops in an area of relative weakness called Killian's dehiscence or the triangle of Killian. This triangle is bordered superiorly by the oblique fibres of the inferior constrictor muscles of the hypopharynx and inferiorly by the transverse fibres of the cricopharyngeus, which constitutes the upper oesophageal sphincter (UES) at the oesophageal inlet.[3]

It has long been postulated that the underlying mechanism leading to the creation of the pharyngeal pouch is a functional disturbance of the hypopharynx, either contraction of the pharyngeal muscles against a hypertonic UES,or incoordination (premature relaxation and contraction of the UES during swallowing). However, when manometry was used to test these hypotheses, conflicting results were produced by different investigators, as summarized elegantly in a recent review.[4] Although patients with Zenker's diverticulum have been found to exhibit lower UES pressures in six controlled studies, UES pressure was no different from controls' in seven studies and, although incoordination between the pharyngeal contraction and the UES relaxation has been shown in eight studies, no such results have been demonstrated in 10. In a recent review of pharyngeal pouch,[5] it is acknowledged that a recent study probably provides us with the most plausible mechanism. This study[6] was the first controlled study to use simultaneous videoradiography and manometry, and to examine the correlation of disease status, bolus volume, pressure curves and co-ordination of pharyngeal contraction and sphincter relaxation in real time. What characterized the patients with Zenker's diverticulum was not neuromuscular incoordination or muscle weakness but a mechanical failure of the UES to open fully (although its relaxation is complete). This in turn caused a marked increase in hypopharyngeal bolus pressure during the phase of transsphincteric bolus flow. This abnormality of the cricopharyngeus muscle is founded upon abnormal histology, that includes degeneration of muscle fibres and their replacement by fat and connective tissue, as shown in biopsy segments of patients undergoing pouch excision and cricopharyngeal myotomy.[7]

Polymyositis is an idiopathic inflammatory myopathy, one of the group of inflammatory myopathies (dermatomyositis, polymyositis and inclusion-body myositis), characterized by chronic muscle inflammation leading to their major clinical features of weakness, muscle tenderness and ultimately atrophy and fibrosis of the muscles, especially, but not exclusively in striated muscles. All three forms are characterized by proximal and often symmetric muscle weakness that develops relatively slowly (weeks to months). Patients usually report difficulty with everyday tasks involving the use of proximal muscles, such as getting up from a chair, climbing steps, lifting objects or combing their hair. Characteristically the face muscles are never affected. While severe weakness is almost always associated with muscle wasting, the sensation remains normal.[8] Dermatomyositis is distinguished from polymyositis by the typical 'heliotrope' eruption, which consists of blue-purple discoloration and oedema of the upper eyelids. The diagnosis of these inflammatory myopathies is confirmed with measurement of serum muscle enzymes (creatinine kinase, aspartate aminotransferase, alanine aminotransferase and lactate dehydrogenate), myopathic potentials in needle electromyography, and most importantly, the muscle biopsy, that shows characteristic inflammatory infiltrates, necrosis and degeneration of muscle fibres. Although dysphagia is a well-documented feature of polymyositis, representing its most prominent extramuscular manifestation, its incidence ranges from 12 per cent to 50 per cent[10] of the patients studied.

Dysphagia is thought to be caused by various mechanisms. The pharyngeal constrictors and upper oesophageal muscle tone is reduced, resulting in diminished propulsion power.[11] Tongue weakness and calcific restrictions of the tongue from fibrosis are cited as contributing factors while the importance of the involvement of the smooth muscle of the lower oesophagus is also recognized.[12]

However, in a patient with weak upper oesophageal constrictors and cricopharyngeus, how can we explain the creation of a pharyngeal pouch that is traditionally considered to be a product of strong peristalsis against an hypertonic UES?

A study of three patients showed that the level of obstruction was at the cricopharyngeus, and it was constriction and fibrosis that was present rather than mere weakness.[13] Interestingly, in these patients, the obstruction was noted during a period of remission or inactivity of their disease, as was the case in our patient. Similar reports of cricopharyngeal obstruction in the absence of active disease have been published earlier.[14]

In this patient there was evidence, both radiographic and endoscopic, of involvement of the cricopharyngeus muscle, of at least 15 years duration. Over this period, the disease progressed, and it would be safe to assume that the histological changes of long-term polymyositis would be present in the cricopharyngeal muscle.

These changes would not be the characteristic inflammation and necrosis/degeneration of muscle fibres, but rather an accumulation of lipid in conjunction with muscle atrophy and replacement of muscle by connective tissue. These changes could result in loss of compliance of the UES and indeed, they are identical to the changes observed in the cricopharyngeus by Cook in the cricopharyngeal muscle specimens. The associated loss of compliance and elevation of the intrabolus pressure could contribute to the creation of a pharyngeal pouch in our patient.

Conclusion

We think it is important to distinguish between a pharyngeal pouch as a cause of dysphagia as opposed to muscle incoordination or weakness in a patient with polymyositis for two reasons; firstly, because a pouch is a cause of dysphagia relatively easily amenable to surgical treatment, especially since the introduction of endoscopic stapling for pharyngeal pouches, which is a very effective as well as safe procedure. Secondly, and perhaps even more importantly, because, dysphagia in such a case does not constitute an index of disease activity and is not associated as one would think, with muscle weakness, but with decreased compliance and fibrosis of cricopharyngeus. Thus, a decision to raise the dose of medication, or interpret this symptom as non-response to therapy would be erroneous and expose the patient to unnecessary and potentially harmful treatment.

References

1 Ludlow A. A case of obstructed deglutition from a preternatural dilatation of and bag formed in the pharynx. *Society of Physicians (London)* 1769;**3**:85
2 Zenker FA, Von Ziemssen H. Krankheiten des oesophagus. In: Von Ziemssen H, ed. *Handbuch der spegiellen pathologie und therapie*. Leipzig: C Vogel, 1877
3 Watemberg S, Landau O, Avrahami R. Zenker's diverticulum: a reappraisal. *Am J Gastroenterol* 1996;**91**:1491–8
4 Fulp SR, Castell DO. Manometric aspects of Zenker's diverticulum *Hepato-Gastroenterology* 1992;**39**:123–6

5 Achkar E. Zenker's diverticulum. *Dig Dis Sci* 1998;**16**:144–51
6 Cook IJ, Gabb M, Panagopoulos V, Jamieson GG, Dodds WJ, Dent J, *et al.* Pharyngeal (Zenker's) diverticulum is a disorder of upper esophageal sphincter opening. *Gastroenterology* 1992;**103**:1229–35
7 Cook IJ, Blumbergs P, Cash K, Jamieson GG, Shearman DJ. Structural abnormalities of the cricopharyngeus muscle in patients with pharyngeal (Zenker's) diverticulum. *J Gastroenterol Hepatol* 1992;**7**:556–62
8 Dalakas MC. Polymyositis, dermatomyositis and inclusion – body myositis. *New Engl J Med* 1991;**325**:1487–98
9 Bohan A, Peter JB, Bowman RL, Pearson CM. Computer-assisted analysis of 153 patients with polymyositis and dermatomyositis. *Medicine (Baltimore)* 1977;**56**:255–86
10 Isenhower D, Schleuning AJ. Otolaryngologic manifestations of systemic disease. In: Paparella MM, Schmirck DA, Gluckmann JL, Meyerhoff WL, eds. *Otolaryngology*. Philadelphia, Pa, WB Saunders, 1991:810
11 Cunningham JD Jr, Lowry LD. Head and neck manifestations of dermatomyositis and polymyositis. *Otolaryngol Head Neck Surg* 1985;**93**:673–7
12 Horowitz M, McNeil JD, Maddern GJ, Collins PJ, Shearman DJ. Abnormalities of gastric and esophageal emptying in polymyositis and dermatomyositis. *Gastroenterology* 1986;**90**:434–9
13 Kagen LJ, Hochman RB, Strong EW. Cricopharyngeal obstruction in inflammatory myopathy: Report of three cases and review of the literature. *Arthritis Rheum* 1985;**28**:630–6
14 Dietz F, Logeman JA, Sahgal V, Schmid FR. Cricopharyngeal muscle dysfunction in the differential diagnosis of dysphagia in polymyositis. *Arthritis Rheum* 1980;**23**:491–5

Address for correspondence:
Christos Georgalas, MD.,
Department of Otolaryngology,
St Mary's Hospital,
Praed Street,
London W2 1NY, UK.

Fax: +44 (0)20 7886 1847

Dr C. Georgalas takes responsibility for the integrity of the content of the paper.
Competing interests: None declared
