Vascular ring presenting as dysphagia in an adult woman: a case report

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ABSTRACT

A 48-year-old woman was seen in a surgical outpatient clinic with a 2 year history of progressive dysphagia with occasional regurgitation, partially controlled with a proton pump inhibitor. Primary investigations of pH testing and gastroscopy were normal, although a barium swallow study revealed significant hold-up at the aortic arch impression and a posterior right-sided oesophageal impression suggestive of a right-sided aortic arch. A follow-up computed tomography angiogram discovered a vascular ring encircling the trachea and oesophagus, formed by a right-sided aortic arch with aberrant aortic branches, and a Kommerell's diverticulum. It was deemed that the patient's symptoms were related to this vascular ring. The patient underwent stage-one surgery – an extra-anatomic bypass of the double aortic arch and right subclavian artery – and 4 months later a stent graft insertion over the origin of the diverticulum with the aim of complete symptomatic relief. This case presents a common symptom familiar to any clinician (dysphagia), which has been caused by a rare pathology. It is even more unusual that this should present itself in adulthood.

KEYWORDS

Vascular ring – Dysphagia – Adult – Kommerell's diverticulum

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Introduction

A 48-year-old woman, previously fit and well, was seen in a surgical outpatient clinic after a general practitioner referral for dysphagia. Her swallowing difficulties had become progressively worse over the previous 2 years; dysphagia was mainly with solid foods (bread and crisps in particular) and resolved by drinking water. She had also noted some occasional regurgitation of undigested food, a 'heartburn' type chest pain and bloating. She did not have weight loss or bowel symptoms. Symptoms were partially controlled with a trial of a proton pump inhibitor. Examination revealed only a mildly tender epigastrium with no palpable masses. She was a non-smoker, non-drinker and of normal body habitus.

Preliminary investigations included a normal gastroscopy, showing no evidence of hiatus hernia or oesophagitis, and pH testing, revealing prolonged episodes of reflux and oesophageal pH of < 4. These were followed by a barium swallow to exclude dysmotility: the oesophageal component of swallow showed significant hold up of solid material at the aortic arch indentation (Fig 1) and there was an impression on the oesophagus to the right side and posteriorly, suggestive of a right-sided aortic arch (Fig 2). The conclusive diagnosis came from a computed tomography (CT) angiogram aorta, showing a vascular ring encircling the trachea and oesophagus. This vascular ring was formed by a dominant right-sided aortic arch with aberrant branches as follows: left brachiocephalic artery (branching into left common carotid and

left subclavian arteries), right common carotid artery and right brachiocephalic artery (branching into right vertebral and right subclavian arteries). A Kommerell's diverticulum was seen at the proximal descending thoracic aorta connected to the left subclavian artery via a non-patent vestigial remnant. At this point, the oesophagus was bound by the trachea, the diverticulum and the left subclavian artery (Fig 3). It is with some certainty that the patient's dysphagia was related to this vascular ring. Stage-one surgery was undertaken 5 months later – an extra-anatomic bypass of the aortic arch and right subclavian artery – and 4 months after that a stent graft was inserted over the origin of the diverticulum as definitive treatment for complete symptomatic relief.

Discussion

Vascular rings, where an abnormal aorta encircles the trachea and oesophagus, are formed when there is abnormal regression of the aortic arches during embryological development.¹ They can be associated with other cardiovascular abnormalities, such as ventricular septal defects and tetralogy of Fallot, in as many as 40% of cases.² The first description of a vascular ring was a double aortic arch reported by Hommel in 1737 but the term 'vascular ring' was not coined until 1945, when Gross published the case of his first successful division of an aortic arch.³ Since then, there have been multiple reports of successfully treating this condition and the various forms of aortic arch have been delineated.

Figure 1 Barium swallow showing significant hold up of solid material at the aortic arch indentation

The incidence of vascular rings is incredibly rare and, although exact figures are not widely reported, they represent less than 1% of all congenital cardiac abnormalities. The double aortic arch variant represents 46% of vascular rings, where bilateral arches form a ring encircling the oesophagus and trachea and combine to form the descending thoracic aorta. With the second most common variant, right aortic arch with left ligamentum arteriosum, there is a right-sided aortic arch with an aberrant left subclavian artery giving rise to the ligamentum arteriosum. It may involve a Kommerell's diverticulum, a bulbous origin of the left subclavian artery. The trachea and oesophagus are surrounded by the ascending and descending aortas, the right-sided aortic arch, and the left-sided ligamentum arteriosum.⁴ Figure 4 shows the anatomy of the patient in the above case for comparison.

Vascular rings present most commonly in infancy and are treated, so the finding of one in an adult is atypical, although case reports have shown that it is possible to reach late adulthood with this pathology.^{1,5} The mechanism by which vascular rings remain asymptomatic into adulthood is poorly understood, but is possibly due to the aging of the aorta (dilating at 0.1cm per decade), in addition to age-related changes of the thorax and mediastinal dimensions, and the

Figure 2 Barium swallow showing an impression on the oesophagus to the right side and posteriorly, suggestive of a right-sided aortic arch

contribution of hypertension and atherosclerosis to aortic compression on surrounding structures.⁵

The symptoms of vascular rings relate to either tracheal compression (the predominant symptoms in infancy) or oesophageal compression, which is the more common presentation in adults, owing to age-related tracheal development.¹ In children, the history alone is usually enough to arouse suspicion of a vascular ring and cases are diagnosed with magnetic resonance imaging (to minimise radiation associated with CT scans). In adults, it is often diagnosed incidentally through a chest x-ray (which may demonstrate widening of the mediastinum or an aortic knob) or, as in this case, barium swallow.³ This is then followed up by CT angiography, with three-dimensional reconstruction being

Figure 3 Computed tomography angiogram demonstrating the right-sided aorta and vascular ring formation by aberrant branches

Figure 4 Anatomy of the vascular ring of the case subject

particularly useful in planning surgery.¹ Echocardiography is indicated in younger patients to rule out other cardiac abnormalities.⁵

Surgery is indicated in all patients with symptomatic vascular rings. This usually involves division of the lesser aortic arch, with ligation of any compressive arterial branches via an ipsilateral thoracotomy or sternotomy.¹ Outcomes are excellent, with symptom resolution in around 80%. Only a small proportion of patients have remaining symptoms that require reoperation: residual tracheomalacia is a risk, especially if the patient had predominant respiratory symptoms.² One case report does tell of oesophageal stenting for symptom relief only and, while this method of treatment has a favourable cost-benefit ratio and is considered less risky, it does only provide temporary relief and surgery is currently the only permanent method of treatment.⁵

Conclusion

This case presents a common symptom familiar to any clinician (dysphagia), which has been caused by a rare pathology. It is even more unusual that this should present itself in adulthood. It highlights the need to consider rarer causes like this when faced with a common presenting complaint for which preliminary investigations bear no answer. Although the incidence of vascular rings is incredibly rare, they are commonly associated with other cardiac abnormalities. Surgery is currently the only permanent method of treatment and prognosis and chances of symptomatic relief are excellent.

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