Case Report

Dysphagia due to an aberrant left subclavian artery in a right-sided aortic arch

E McKenna, B E Kelly, M Khan

Accepted 4 April 2001

In 1794, Bayford described the post-mortem findings of a woman with life-long dysphagia who eventually died of starvation, caused by oesophageal obstruction. At post-mortem a right subclavian artery was identified passing aberrantly from a left-sided aortic arch behind the oesophagus, causing the woman's dysphagia. Dr Bayford referred to this extraordinary disposition of the right subclavian artery as, lusus nature or sport of nature. Since then the term 'dysphagia lusoria' has been used to refer to all aortic root anomalies causing oesophageal dysphagia. We report a case of dysphagia lusoria caused by a right aortic arch with a large diverticulum (diverticulum of Kommerell) at the origin of an aberrant left subclavian artery.

CASE REPORT A 45 year old lady presented with a 2 month history of fatigue, shortness of breath, and a feeling of tightness at the lower end of her sternum associated with occasional episodes of dysphagia. Physical examination was unremarkable. Her ECG was normal. A chest radiograph demonstrated a right sided aortic arch. A subsequent barium meal demonstrated an area of constriction in the upper third of the thoracic oesophagus with an indentation arising posteriorly causing compression and contrast hold up. (Fig 1). The indentation corresponded to the level of the aortic arch and the radiological appearances suggested this was due to an aberrant vessel. Echocardiography was normal but subsequent

![Barium meal image showing compression of the oesophagus by an aberrant vessel.](image1)

Fig 1

![Axial T1-Weighted MR image through the thorax shows the right-sided aortic arch (open arrow) and the aberrant left subclavian artery with the wide mouthed diverticulum of Kommerell at its origin (closed arrow).](image2)

Fig 2

Department of Radiology, Royal Victoria Hospital, Grosvenor Road, Belfast BT12 6BA.
E McKenna, MB, BCh, BSc, MRCP, Specialist Registrar in Radiology.
B E Kelly, MD, FRCS, FRCR, FFRRCSI, Consultant Radiologist.
M Khan, MB, BS, FRCP, Consultant Cardiologist.
Correspondence to Dr Kelly.
During embryologic development the aortic arch begins as a duplicated system. As normal embryologic development continues, the right arch atrophies beyond the origin of the right common carotid and subclavian arteries. In 70% of the population the right subclavian and right common carotid merge to form the right innominate artery, the first branch of the normal left arch. In our patient the right arch persisted during embryologic development, while the left arch became vestigial between the origin of the left common carotid and left subclavian arteries. The diverticulum which develops from a vestigial remnant of the distal, embryologic left arch and gives off an aberrant left subclavian artery has come to be known as the diverticulum of Kommerell. A persistent right aortic arch with a diverticulum of Kommerell and aberrant left subclavian artery is an uncommon anomaly, estimated to occur in 1 in 1000 individuals. To actually see attributable symptoms associated with this anomaly is rare, as the ring that is formed, is generally loose. Importantly, 5% of patients with this aortic root anomaly will have an associated congenital heart defect and 2% of patients with tetralogy of F allot will have this aortic arch abnormality. Echocardiography in this patient revealed no such abnormality.

The diagnosis of dysphagia lusoria can be elusive. It is not uncommon for patients to undergo repeated investigations for dysphagia that are unrevealing. In cases where dysphagia lusoria is undiagnosed, patients have commonly been prescribed sedatives and tranquillisers for a presumed psychosomatic cause of their dysphagia.

The upper gastrointestinal barium study is an excellent method for the evaluation of dysphagia lusoria, but the diagnosis can easily be missed if the high thoracic oesophagus is not carefully examined, and if lateral or oblique projections of the oesophagus are not obtained. Endoscopy has the advantage of excluding other potential causes of dysphagia but it has been reported that it has a false negative rate of up to 50% in patients with dysphagia lusoria. Both computerised tomography (CT) and MR of the chest are helpful in evaluating the patient with suspected dysphagia lusoria. MR is particularly sensitive in the evaluation of the mediastinal vasculature and can graphically demonstrate aortic arch anomalies. With its inherent advantages of multiplanar imaging and

**Fig 3.** Coronal T1-Weighted MR image illustrating the diverticulum at the origin of the aberrant vessel.

Cardiac catheterisation confirmed the right sided aortic arch and also a diverticulum, at the origin of an aberrant left subclavian artery, indenting the oesophagus. To further define the anatomy a magnetic resonance scan (MR) was performed (Fig 2 and Fig 3) which demonstrates the aberrant vessel and the diverticulum.

Surgery was deemed inappropriate given the mild symptoms and a conservative approach was adopted.

**DISCUSSION**

Developmental anomalies of the aortic arch and its major branches are relatively common representing 3% in post mortem series, but they are usually asymptomatic. Aortic arch anomalies become symptomatic when they completely ‘ring’ the trachea and oesophagus or when coincident congenital heart defects occur.

Symptomatic aortic arch anomalies clinically present in a bimodal fashion. The trachea is compressible during infancy and these patients therefore typically present with respiratory symptoms: stridor, wheezing, cyanosis, or recurrent pneumonia when solid foods are introduced. In adults, as the trachea is rigid, the oesophagus is more likely to be compressed, resulting in dysphagia.
non ionising radiation, MR is rapidly becoming the imaging modality first employed. Angiography is still needed for pre-operative assessment.

Management of dysphagia lusoria is dependent upon the severity of the symptoms. In our patient, the symptoms were relatively mild and slight dietary modification and explanation of the cause of the symptoms has been adequate. She has undergone review at outpatients and a second MR scan performed five years after the initial study has shown no change in the size of the diverticulum. In patients with increasing dysphagia or weight loss, surgical cure is possible by ligation and resection of the aberrant vessel.

CONCLUSIONS

The diagnosis of dysphagia lusoria requires a high index of suspicion. These patients present with symptoms of intermittent dysphagia and a mediastinal abnormality seen on a chest radiograph. Non invasive imaging of the chest with either CT or MR are excellent methods for evaluating the mediastinum for solid tumours or vascular anomalies that can cause extrinsic oesophageal compression. Dysphagia lusoria caused by a persistent right embryologic aortic arch and diverticulum of Kommerell with an aberrant left subclavian artery may be managed by dietary modification and follow-up when the symptoms are mild.

REFERENCES

1. Asherson N. David Bayford: His syndrome and sign of dysphagia lusoria. Ann R Coll Surg Eng 1979; 61: 63-7.

2. McNally P R, Rak K M. Dysphagia Lusoria caused by persistent right aortic arch with aberrant left subclavian artery and diverticulum of Kommerell. Dig Dis Sci 1992; 37: 144-9.

3. Kommerell B. Verlagerung des Osophagus durch eine abnorm verlaufende Arteria Subclavia Dextra (Arteria Lusoria). Fortchr Geb Roentgenstr Nuklearmed 1936; 380: 303-15.

4. Shuford W H, Sybers R G. The Aortic Arch and its Malformations with Emphasis on the Angiographic Features. Springfield, Illinois, Charles C Thomas 1974; 18-92.

5. Payne D N R, Lincoln C, Bush A. Right sided aortic arch in children with persistent respiratory symptoms. BMJ 2000; 321: 687-8.

6. Shuford W H, Sybers R G, Edwards F K. The three types of aortic arch. Am J Roentgenol 1970; 109: 67-83.

7. Berenzweig H, Baue A K, Mc Callum R W. Dysphagia Lusoria: Report of a case and a review of the diagnostic and surgical approach. Dig Dis Sci 1980; 25: 630-6.

8. Janssen M et al. Dysphagia lusoria: clinical aspects, manometric findings, diagnosis, and therapy. Am J Gastroenterol 2000; 95: 1411-6.

9. Kersting-Sommerhoff B A et al. MR imaging of congenital anomalies of the aortic arch. AJR 1987; 149: 9-13.

10. Skinner D B, Belsey R H R. Management of esophageal disease. Philadelphia, W B Saunders 1988; 378-85.