Aberrant subclavian artery is the most frequent aortic arch anomaly and occurs in 0.5% of normal individuals. The majority of people with this anomaly are asymptomatic and nothing needs to be done. However, cervicomediastinal surgery for unrelated conditions predisposes patients to potential morbidity and even fatal outcomes. This paper describes the management of two symptomatic patients, with a literature review.

Case reports

Case 1
A 49-year-old man presented with left lower limb rest pain due to chronic arterial insufficiency. He had moderate weight loss, nocturnal cough with copious yellowish-green sputum in the mornings and dysphagia. He had clubbing of the fingers, right-sided chest crepitations and bronchial breathing. His blood pressure, taken in both upper limbs, was 140/80 mmHg and his pulse rate 80/min. The left femoral and distal pulses were absent, with no ulceration. A chest radiograph showed right middle-lobe bronchiectasis, and routine blood test results were normal. Chest physiotherapy and oral antibiotics were commenced. A barium swallow showed external compression of the upper third of the thoracic oesophagus (Fig. 1).

Angiography revealed an aberrant right subclavian artery (Fig. 2) and an abrupt left common iliac artery occlusion. The patient underwent a right subclavian-carotid transposition through a right cervicotomy without an intraluminal shunt (stump pressure 98 mmHg) and concomitant femoro-femoral bypass.

His postoperative course required intensive chest physiotherapy to regain right pulmonary function. His postoperative swallowing is normal, his barium swallow shows no obstruction and his rest pain is cured.

Case 2
A 76-year-old man presented with dysphagia and weight loss. He was found to be emaciated and dehydrated and had distended neck veins. A diagnosis of cancer of the oesophagus was made, but the barium swallow showed external compression. A computed tomography (CT) scan showed an aneurysmal lusorium vessel (Fig. 3).
Dysphagia is thought to occur in the elderly population, as are atherosclerotic aneurysms as a result of non-involution of the right dorsal aorta, known as Kommerell’s diverticulum. Atherosclerotic aneurysm formation is a sign of progression of atherosclerosis to the right subclavian artery (from Sadler[1]).

Fig. 3. CT scan showing a thrombus-lined aneurysm compressing the oesophagus (arrow).

Endoscopy revealed multiple linear erosions in the oesophagus. A two-stage operative approach was planned. As a first step, a subclavian-common-carotid transposition was carried out through a right supraclavicular approach without intraluminal shunting (the shunt pressure was 68 mmHg).

The vessel was ligated to the left of the oesophagus. A left thoracotomy and ligation of the aneurysmal aberrant subclavian artery was planned for a later date. The patient was awake when taken to the ICU for controlled extubation and haemodialysis. Unfortunately he developed pulmonary oedema and died 4 days later.

Discussion

The aberrant right subclavian artery is the commonest embryological variant of the aortic arch branches and occurs in 1 in 200 of the population.1-4 The majority of these individuals are asymptomatic; however, cervico-mediastinal surgery for unrelated conditions predisposes patients to potential morbidity and even fatal outcomes.5-10

The condition was first described by Hunauld in 1735 and was recognised as a cause of dysphagia by Bayford in 1794 who termed it ‘dysphagia lusus naturae’, meaning dysphagia caused by a freak of nature.1,2,4,11 Over the years this condition became known simply as dysphagia lusoria.11 The first successful operative procedure was simple ligation performed by Gross in 1946.12 Subclavian-carotid transposition was first performed by Hallman and Cooley.1 In 1965 Bailey and associates13 were the first to perform a repair including reconstruction of flow to the right upper limb using the ascending aorta to prevent ischaemia to the upper limb and subclavian steal syndrome.

The embryological explanation of the anomaly is thought to be obliteration of the right 4th aortic arch and proximal portion of the right dorsal aorta with persistence of the distal portion of the right dorsal aorta and the 7th intersegmental artery (Fig. 4, A and B).14

The course of this errant vessel through the mediastinum is 80% retro-oesophageal, 15% anterior to the oesophagus and 5% ante-tracheal. It may be aneurysmal at its origin as a result of non-involution of the right dorsal aorta, known as Kommerell’s diverticulum. Atherosclerotic aneurysm formation is thought to occur in the elderly population, as are atherosclerotic stenotic lesions.2,3,15

The diagnosis is aided by a plain chest radiograph, which shows a postero-superior mediastinal mass. Barium swallow demonstrates a bayonet configuration with a rounded or oblique external indentation on the upper third of the thoracic oesophagus at the level of the 4th thoracic vertebra.19 Oesophagoscopy done cautiously may reveal a pulsatile swelling in the upper oesophagus. The right radial arterial pulse is said to disappear on compression of this mass. However, this practice should be avoided because perforation or rupture of the vessel may ensue with catastrophic results.1,2 CT or magnetic resonance imaging has been found to be useful, as in our second patient.6-10

The management of the condition has evolved over the years since it was first described.17 Kieffer et al. recommend classification into four simple groups based on clinical presentation and the anatomy of the lesion on angiography:1

Group 1: aberrant right subclavian artery causing compression of the oesophagus and trachea
Group 2: occlusive disease in the aberrant subclavian artery causing ischaemia
Group 3: aneurysmal group with distal thrombo-embolism with or without aortic arch disease
Group 4: aberrant subclavian artery arising from a diseased aortic arch, either aneurysmal or the result of occlusive disease.

The diagnosis is confirmed and surgery planned for symptomatic patients and those with aneurysm on the basis of angiographic appearance; contrast angiography remains the investigation of choice.3 Angiography may require left axillary access to avoid the aneurysm. Recommended surgical correction16 is via a right-sided cervicotommy as first described by Orvald et al.21 in 1972 and modified by Valentine et al.12 in 1987, ligating the erratic vessel to the left of the oesophagus and as close as possible to its aortic arch origin. A right subclavian-carotid direct transposition is performed or an interposition prosthesis is utilised.1,4 This suffices for most group 1 and group 2 lesions. For groups 3 and 4, a contralateral thoracotomy or sternotomy is necessary to excise the origin of the errant vessel and cardiopulmonary bypass is frequently necessary.4 Aneurysm rupture and death have been reported in unoperated patients.1,12 Revascularisation using the ascending aorta or aortic replacement prosthesis may be utilised,17 and axillo-axillary bypass and a left thoracotomy to excise the right aberrant subclavian artery at its origin have been performed.1

Our first patient presented with bronchiectasis of the right
middle lobe from chronic nocturnal aspiration. This complication has not been reported in adults. The cause of the iliac occlusion could have been embolic, although no cardiac or aortic lesion was found during investigation. The second patient presented with superior mediastinal syndrome due to the aneurysm and had transposition only because of his poor general condition, as recommended by Kieffer et al.\(^1\)

**Conclusion**

Symptomatic patients with a lusorian vessel require surgical correction. Asymptomatic patients with an aneurysm of this vessel should be considered for intervention. A high index of suspicion should be adopted in cervicomediastinal elective and emergency surgery including trauma. Contrast angiography seems to be the investigation of choice.

**REFERENCES**


**Erratum**

In the article on the Department of Surgery, University of the Witwatersrand, which appeared in the May 2006 issue of *SAJS*, the photo on the right on p. 49 was incorrectly captioned. It is not Percy Fox, but his son Edwin.