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CASE REPORT

Dysphagia Lusoria in Children

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Introduction

The aberrant right subclavian artery (ARSA) is the most common of the aortic arch congenital vascular anomalies. It arises as the fourth branch of the aortic arch and occurs in 0.5% to 1.8% of the population.[1–3] The association of difficulty in swallowing and an aberrant retroesophageal right subclavian artery, is called dysphagia lusoria, and has been rarely reported in children.

This report presents two children with dysphagia lusoria who were successfully treated by surgery at our department. The clinical, diagnostic and therapeutic aspects of this rare condition are discussed.

Case Reports

Patient

A 2-year-old girl had a history of slow and difficult feeding since infancy. She refused to eat solid food, and failed to thrive. The results of a physical examination were normal, except for generalised hypotonia and mild developmental retardation. Electroencephalogram and computerised tomography were normal. A barium swallow showed an oblique indentation in the posterior esophagus (Fig. 1). Angiography showed an aberrant right subclavian artery, common origin of both carotid arteries, and a large anomalous vertebral artery originating from the aortic arch (Fig. 2). At operation, the right aberrant subclavian artery was ligated and divided via a left thoracotomy, without arterial reconstruction. A postoperative barium swallow was completely normal. At follow-up after 3 and 6 months, the child's refusal to eat solid food had completely resolved.

Patient 2

A 12-year-old girl had a history of poor feeding since the age of 7 months. She refused to eat solid food and only kept her weight steady thanks to large amounts of ice cream. Results of a physical examination were normal. Because of her continual refusal to eat over

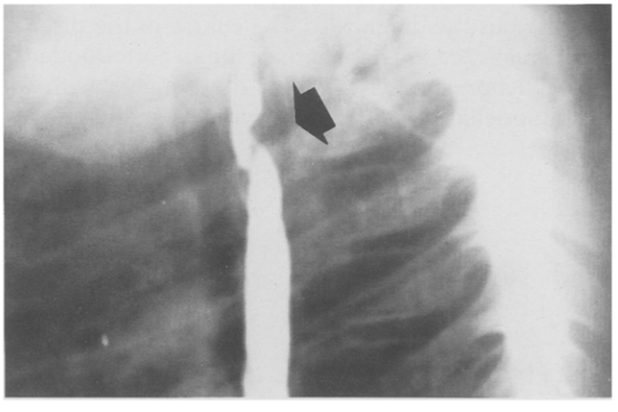

Fig. 1 Preoperative barium swallow of the first patient. Anteroposterior film showing oblique compression of the posterior oesophagus (arrow).

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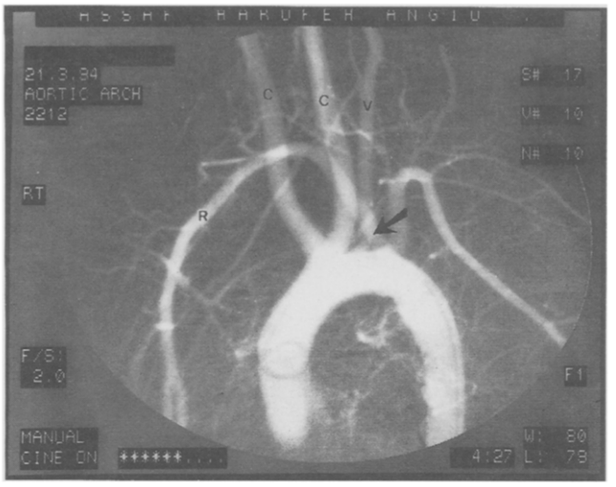

Fig. 2 Angiography of the first patient. The aberrant right subclavian artery (R) is seen to arise from the left side of the aortic arch (arrow). Note that the right and left common carotid arteries (C) have a common trunk, and that a large anomalous vertebral artery (V) arises directly from the aortic arch.

the years, a child psychiatrist was consulted and ruled out psychological reasons as the cause of her problem. An extensive medical examination was then conducted. A barium swallow showed an oblique indentation in the posterior esophagus. Esophageal manometry was normal, except for a pulsatile high-pressure zone distal to the upper esophageal sphincter. Angiography demonstrated an aberrant subclavian artery and a common origin of the carotid arteries (Fig. 3). At operation, a proximal transection of the right aberrant subclavian artery and end-to-side anastomosis to the common carotid was performed, via a right supraclavicular, extrathoracic approach. The postoperative course was completely uneventful. A transient right ptosis subsided within days. A postoperative barium swallow was completely normal. She began eating solid foods gradually and at follow-up 12 months later she is symptom free.

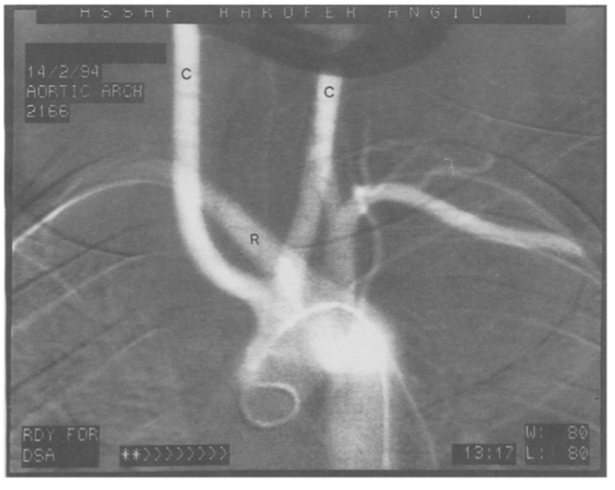

Fig. 3 Angiography of the second patient. The aberrant right subclavian artery (R) arises from the posterior aspect of the aortic arch. A common trunk is seen to give rise to two common carotid arteries (C).

Discussion

Bayford first reported the syndrome of symptomatic compression of the esophagus by the aberrant right subclavian artery in 1794.[4] He named it "dysphagia lusoria" after lusus naturae (jest of nature). A retro-oesophageal right subclavian artery is the most common congenital aortic arch abnormality, usually asymptomatic, and rarely associated with dysphagia. When posterior compression of the esophagus is present, digestive or respiratory symptoms may follow. Progressive dysphagia and severe loss of weight have been reported.[5–8]

The subclavian arteries are embryological products of the seventh intersegmental arteries. When the right subclavian artery is aberrant, there is a need for caution during surgery because of the right recurrent laryngeal nerve's new course. Persistence of the distal right dorsal aorta, described as an aortic diverticulum, is called "Kommerell's diverticulum"[9] and is more likely to produce symptoms from esophageal compression. The pathophysiology of dysphagia lusoria is based on Klinkhamer's observations.[10] He reviewed 292 patients with ARSA and came to the conclusion that symptoms occurred only when a common carotid trunk or two closely arising carotid arteries prevented the trachea and oesophagus from bending forward over the retro-oesophageal vessel, resulting in compression of these organs between the aberrant vessel posteriorly and two carotid arteries anteriorly. His observations were confirmed in our two patients.

The timing of the onset of symptoms depends upon the specific anatomy of the aberrant artery.[10] If the vessel significantly impinges on the esophagus at birth, difficulty in feeding can be observed when solid food is started. If adequate energy is obtained from liquids and soft foods, then weight can be normal, as was the case with our second patient, who ate ice cream round the clock.

Plain chest films, although nonspecific, may reveal findings of a superior mediastinal mass. Barium studies of the upper gastrointestinal tract will show an oblique indentation, a triangular posterior defect, in the posterior esophagus, excluding other oesophageal

abnormalities. Endoscopy will be consistent with an extraluminal mass, with the mucosa intact. Oesophageal manometric studies will show a pulsatile high pressure zone in the region of the aberrant artery. Computerised tomography and magnetic resonance can establish the diagnosis and also give further information regarding aneurysmatic dilatations. Angiography of the aortic arch and its branches, however, remains the "gold standard" for establishing the final diagnosis.

The only effective answer to dysphagia lusoria is surgical correction. Gross was the first to successfully perform simple ligation and division of the ARSA posterior to the esophagus via a left thoracotomy, with minimal morbidity and complete relief of dysphagia.[11] This simple and safe approach was used in the treatment of our first patient, with good results, and is still the method of choice for children under the age of 2 years. It is very important to divide the artery close to its origin, otherwise a stump can be left behind the oesophagus. Thrombosis of this stump may lead to persistent dysphagia, as reported by Piffare *et al.*[12] Subclavian steal syndrome[13] and ischaemia of the right arm, subsequent to interruption of the subclavian flow, have been rarely observed in infants and was not observed in our patient. Shumacker *et al.* were the first to divide and reanastomose the artery, by carrying out an end-to-side anastomosis with the right carotid artery, through a median sternotomy.[14] Orvald *et al.* were the first to demonstrate that the ARSA can be successfully divided and transposed into the right carotid artery, through a simple cervical incision.[15] This approach carries less risk than sternotomy or thoracotomy and re-establishes flow without compromising the carotid distribution, and eliminates the need for a prosthetic graft. It was our choice for the treatment of our second patient, with good results. Many surgical options are available for the treatment of severe dysphagia lusoria, all of which can provide good results with reasonable risk. The treatment options should be individualised to the patient and the presenting symptomatology.

In summary, aberrant right subclavian arteries are usually asymptomatic. When symptoms do occur, they can be successfully relieved by surgery. In early age (up to 2–3 years) simple ligation of the artery at its origin via a left thoracotomy will suffice. In the older patients, the procedure of choice should be transection of the aberrant vessel and end-to-side anastomosis to the common carotid, through an extrathoracic right subclavicular incision. If a Kommerel diverticulum or aneurysm of the ARSA is present, a combined two-stage surgical approach may be necessary.

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