Clinical presentation, radiological features and outcome of vascular rings and pulmonary slings in young patients

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Introduction

Congenital anomalies of the aorta are important to recognise as they can be associated with vascular ring or sling, and cause compression to the airway and esophagus. The purpose of this poster is to describe the embryology and anatomy of the aortic arch system, demonstrate the imaging findings in certain congenital aortic arch anomalies and discuss the clinical outcome and prognosis in patients with these anomalies.

Development of the aorta starts during the third week of gestation. A primitive aorta consists of a ventral segment and a dorsal segment. The ventral segments fuse to form the aortic sac, whereas the dorsal segments form the midline descending aorta. Six pairs of branchial arch arteries are formed between the ventral and dorsal segments. These primitive branchial arches appear and regress sequentially in a caudocranial order. A normal and mature aortic system is formed when some arches regress and others persist.

Materials and Methods

All patients with confirmed diagnosis of vascular ring or pulmonary artery sling by computer tomography (CT) or magnetic resonance imaging (MRI) referred from the Department of Paediatrics, Queen Elizabeth Hospital between July 2009 and July 2019 were retrospectively reviewed. Their clinical presentation, comorbidities, imaging findings, treatment and follow-ups were studied.

A total of 21 patients were included, with a median age of 3 months (range: 1 day to 22 years old) at the time of diagnosis, including 4 cases with double aortic arch, 12 with right aortic arch with aberrant left subclavian artery and 5 with pulmonary artery sling (1 case with type I, 4 cases with type II).

Results

Double Aortic Arch

Double aortic arch is the commonest cause of a symptomatic vascular ring. It results from persistent right and left arches, each side forming carotid and subclavian arteries ipsilaterally. One of the sixth branchial arches and left arches, each side forming carotid and subclavian arteries just above the arches, the "four vessel sign".

Right Aortic Arch with Atrioventricular Septal Defect

Right aortic arch with atrioventricular septal defect is the second most common anomaly of a vascular ring. It results from regression of the left fourth arch and persistence of left sixth arch which later forms the ligamentum arteriosum. Part of the left dorsal aorta typically persists as the retro-esophageal diverticulum of Kommerell and gives rise to the aberrant left subclavian artery. A patent left sided ductus arteriosus connects the diverticulum and the left pulmonary artery, therefore completing the vascular ring.

Similar to double aortic arch, symptoms may arise from compression of the trachea and esophagus. In the reviewed cases, all of the patients remain symptom-free.

Pulmonary Artery Sling

In this rarer vascular developmental abnormality, aberrant left pulmonary artery arises from the right pulmonary artery, takes its course between the trachea and esophagus to reach the left hilum. Therefore, a sling is formed around the distal trachea and proximal right main bronchus.

In the included cases, patients with type I pulmonary artery sling was diagnosed incidentally. All patients with type II pulmonary artery sling presented in early infancy with respiratory failure, required intensive care and early surgical correction.

Treatment and Outcome

All patients with double aortic arch and right aortic arch with aberrant left subclavian artery remained asymptomatic despite esophageal and tracheal compression. None of them required surgical treatment and remained symptom-free with a median follow-up of 27 months. All patients with type II pulmonary artery sling showed early respiratory symptoms and required surgical treatment.

Conclusions

Patients with double aortic arch and right aortic arch with aberrant left subclavian artery can remain asymptomatic, suggesting watchful waiting without intervention a viable approach. Patients with pulmonary artery sling have generally worse prognosis, particularly those with type II anomaly. Cross-sectional study is crucial in patients with clinical suspicion of pulmonary artery sling, in order to evaluate the type of pulmonary artery sling and severity of airway compromise which has prognostic implications.

References