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

伊利沙伯醫院
QUEEN ELIZABETH HOSPITAL

LY Lam, TWY Chin, JCY Lee
Department of Radiology and Imaging, Queen Elizabeth Hospital, Hong Kong SAR

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 craniocaudal order. A normal and mature aortic system is formed when some arches regress and others persist.

Figure 1: Development of the embryonic aortic arches and branches. Areas that are shaded grey represent segments that involute in normal development.

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 persist as the ductus arteriosus.

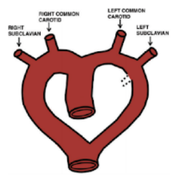

Figure 2: Double aortic arch resulting from the persistence of embryologic double arch system.

Clinical presentations include noisy breathing in young infants or dysphagia in some cases. However, for the reviewed cases, all of the patients were asymptomatic at the time of diagnosis despite esophageal and tracheal compression.

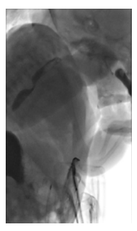

Figure 3: Radiograph in a barium swallow study depicting esophageal indentation by the double aortic arch in a young infant.

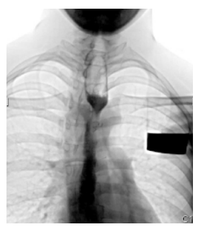

Figure 4: Frontal radiograph in VFSS study depicting esophageal narrowing and encircling by the double aortic arch in an adolescent.

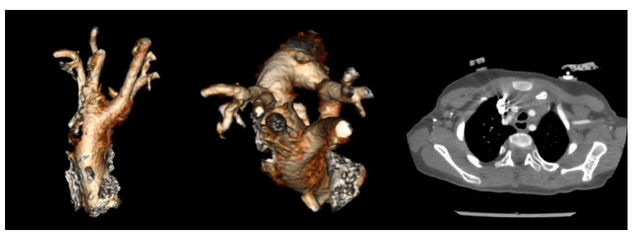

Figure 5: Volume-rendered CT image and corresponding axial CT image showing configuration of double aortic arch and symmetric appearance of bilateral common carotid arteries and subclavian arteries just above the arches, the "four vessel sign".

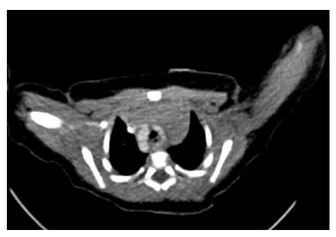

Figure 6: Axial CT image depicting a typical finding of a larger right arch.

Right Aortic Arch with Aberrant Left Subclavian Artery

Right aortic arch with aberrant left subclavian artery 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.

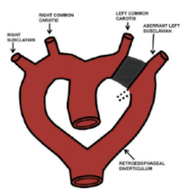

Figure 7: Right aortic arch with regression of the left arch. Ductus arteriosus or ligamentum arteriosum (dashed line) completes the vascular ring. Both the ligamentum arteriosum and aberrant left subclavian artery arise from the diverticulum of Kommerell.

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.

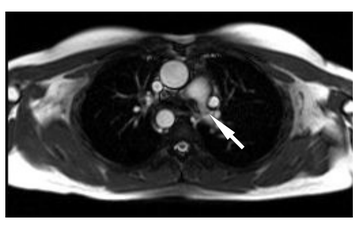

Figure 8: Axial MR image, showing a dimple on left pulmonary artery, suggestive of insertion of ligamentum arteriosum (white arrow).

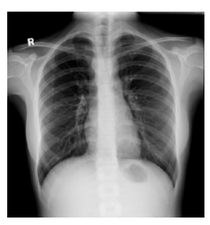

Figure 9: Frontal chest radiograph showing absence of normal left aortic notch. Presence of right arch shadow at right side of distal trachea.

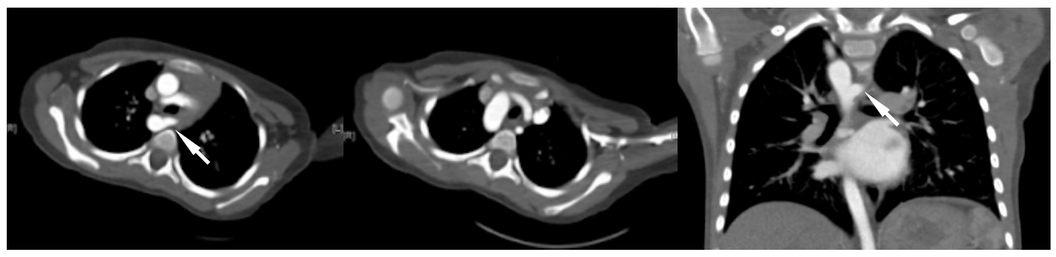

Figure 10: Axial and coronal CT images depicting diverticulum of Kommerell (white arrows), which gives rise to a retro-esoghgeal aberrant left subclavian artery.

Pulmonary Artery Sling

In this rarer vascular developmental anomaly, 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.

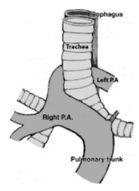

Figure 11: Diagram showing the anomalous origin of the left pulmonary artery from the right pulmonary artery, taking a retro-tracheal course to reach the left hilum.

Patients with pulmonary artery sling can be divided into two main types depending on the location of the carina: type I with a normal bronchial pattern and normally located carina at T4-5 levels; type II is characterized by a more inferiorly located left pulmonary artery sling and abnormal bronchial branching with a low T-shaped carina located at T6-T7 levels. In the latter type, associated long segment stenosis of the trachea is common, patients also suffer a higher mortality and morbidity during infancy. Type I patients may be asymptomatic.

In the included patients, one patient 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.

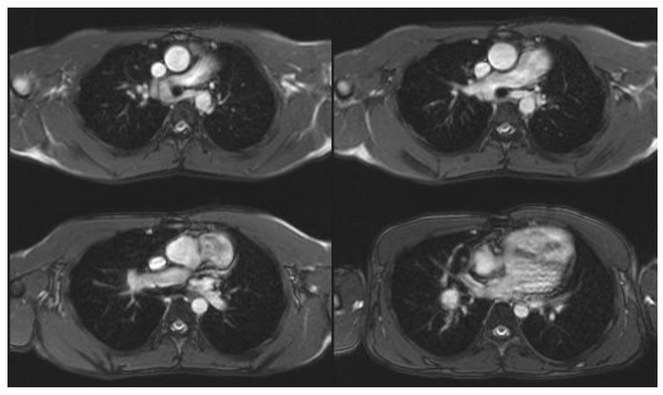

Figure 12: Case of type I pulmonary artery sling. Axial MR image showing the left pulmonary artery arises from the right pulmonary artery with aberrant retrotracheal course. The left pulmonary artery is smaller in calibre as compared to the right pulmonary artery.

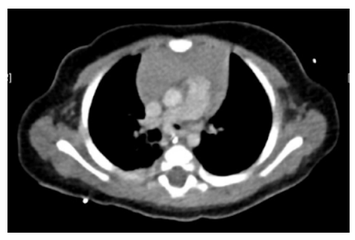

Figure 13: Case of type II pulmonary artery sling. Axial CT image depicts aberrant retrotracheal left pulmonary artery with compression to the trachea and esophagus (with feeding tube in-situ).

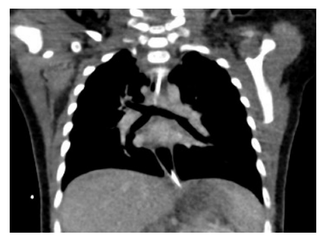

Figure 14: Case of type II pulmonary artery sling. Coronal reformatted CT image shows the carina located at a lower position at levels of T5-6. The left main bronchus shows long segment narrowing with a horizontal lie.

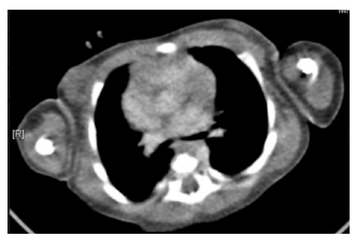

Figure 15: Case of type II pulmonary artery sling. Axial CT image depicts a low T-shaped carina.

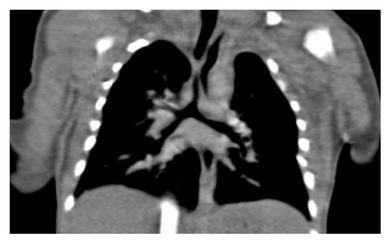

Figure 16: Case of type II pulmonary artery sling. Coronal reformatted CT image shows a narrowed lower trachea just above carina.

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

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