



Right Aortic Arch with Aberrant Left Subclavian Artery : A Case Report

KEYWORDS

Right sided aortic arch, aberrant left subclavian artery, embryonic aortic arches, right fourth branchial arch, dysphagia lusoria.

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ABSTRACT Right sided aortic arch is a developmental anomaly resulting from persistence of right fourth branchial arch during embryonic development of the aorta. It is usually asymptomatic and is commonly an incidental finding, but may at times be associated with extrinsic esophageal compression and dysphagia or congenital heart disease. We present a female patient with restrictive lung disease and pulmonary hypertension with cardiac failure, who was incidentally found to have a right sided aortic arch with aberrant left subclavian artery.

INTRODUCTION

Right sided aortic arch is an aortic developmental anomaly occurring in about 0.05% of the population¹. It results from persistence of the right fourth branchial arch during embryonic development of the aorta^{1,2}. Right sided aortic arch is commonly an incidental finding, though it may sometimes be associated congenital heart disease^{1,3,4,5}. Right sided aortic arch anomaly may occasionally be associated with dysphagia due to extrinsic esophageal compression caused by an aberrant subclavian artery, known as dysphagia lusoria^{6,7,8}. We present a female patient admitted with lung parenchymal disease, pulmonary hypertension and cardiac failure, who was incidentally found to have a right sided aortic arch and an aberrant left subclavian artery.

CASE REPORT

A 65 year old female patient was admitted with a one month history of exertional breathlessness, intermittent palpitations and bilateral lower limb swelling. On examination, she had an irregular pulse (84/min); was tachypnoeic, pale, with significant pedal edema, raised JVP and a left sided thyroid swelling. On examination of the cardiovascular system, she had an ejection systolic murmur at the base; chest examination revealed bilateral basal crepitations; on abdominal examination she had mild hepatomegaly and presence of free fluid. The patient was administered diuretics, oxygen therapy and hematinics. Investigations of the patient were as follows: Hemoglobin - 9.4 gm%, peripheral smear - microcytic, hypochromic RBCs; ECG showed atrial fibrillation with a ventricular rate of 80/min. Chest x-ray was suggestive of cardiomegaly, prominent pulmonary trunk and a right sided aortic arch (Image 1). Neck ultrasound revealed enlarged left lobe of thyroid with multiloculated left sided lesions; thyroid function tests were suggestive of hyper-thyroidism. Pulmonary function tests were suggestive of moderate restriction; echocardiography showed dilated right atrium and right ventricle, moderate pulmonary hypertension and an ejection fraction of 55%.

Computed tomography of the chest was performed which showed a right sided aortic arch and an aberrant left subclavian artery coursing posterior to the esophagus, but without causing any esophageal compression (Images 2,3,4).

Treatment was continued in the form of diuretics and hematinics; a calcium channel blocker and phosphodiesterase inhibitor were added in view of significant pulmonary hypertension. The patient was also put on anti-thyroid therapy. Patient improved considerably and was discharged after a couple of weeks.

DISCUSSION

Right sided aortic arch is a developmental anomaly of the thoracic aorta resulting from persistence of the right fourth branchial arch during embryonic development of the aorta^{1,2}. Aortic development occurs during 3rd week of gestation. Each primitive aorta consists of a ventral and a dorsal segment that are continuous through the 1st aortic arch. The two ventral aortae fuse to form the aortic sac. The dorsal aortae fuse to form the midline descending aorta. Six paired aortic arches, the so-called branchial arch arteries develop between the ventral and dorsal aortae. Each of the pairs of arch arteries give rise to arteries as follows²:

1. Proximal segment of the 3rd pair: Common carotid arteries.
2. Left arch of the 4th pair: Segment of normal left aortic arch between the left common carotid and the left subclavian arteries.
3. Right 4th arch: Right subclavian artery.
4. Bilateral 6th arches: Pulmonary arteries. (Image 5).

The 1st, 2nd and 5th arch pairs eventually disappear.

Anomaly in the embryonic development of aorta with persistence of the right 4th branchial arch gives rise to a right sided aortic arch. The various arch anomalies that can be encountered as described below include^{2,9,10}:

1. Left aortic arch with aberrant right subclavian artery.
2. Right aortic arch with aberrant left subclavian artery.
3. Double aortic arch.
4. Cervical aortic arch.
5. Interrupted aortic arch.

Right sided aortic arch anomaly is usually asymptomatic and an incidental finding, but may occasionally be associated with abnormal course of an aberrant left subclavian artery^{6,7,8}. The aberrant left subclavian artery courses posterior to the esophagus and can cause extrinsic esophageal compression and dysphagia^{6,7,8}. This is known as dysphagia lusoria⁶.

Right aortic arch anomaly may also be associated with congenital cardiac anomalies. There are case reports of concurrent occurrence of right sided aortic arch with an atrial septal defect⁴ or anomalous origin of right pulmonary artery⁵. Our patient did not have dysphagia or any congenital cardiac anomaly.

CONCLUSION

Right sided aortic arch is a developmental anomaly resulting from persistence of the right fourth branchial arch during embryonic development of the aorta and its branches. It is most often symptomatic and an incidental finding, but may occasionally be associated with congenital cardiac anomalies or dysphagia due to esophageal compression by an aberrant left subclavian artery.

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Image 1: X-Ray Chest showing right aortic arch



Image 2: CT image showing right aortic arch



Image 3: CT image showing descending aorta seen in the right hemithorax.

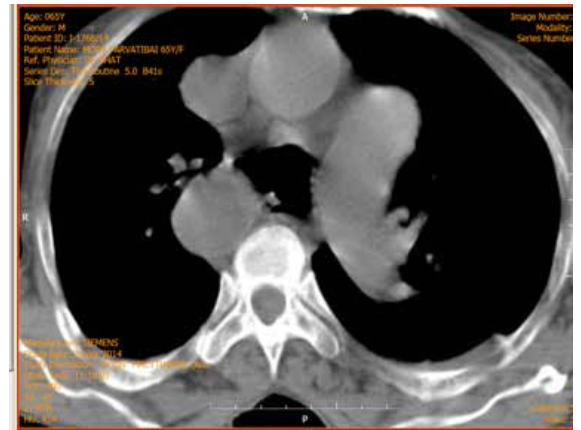


Image 4: CT image showing aberrant left subclavian artery

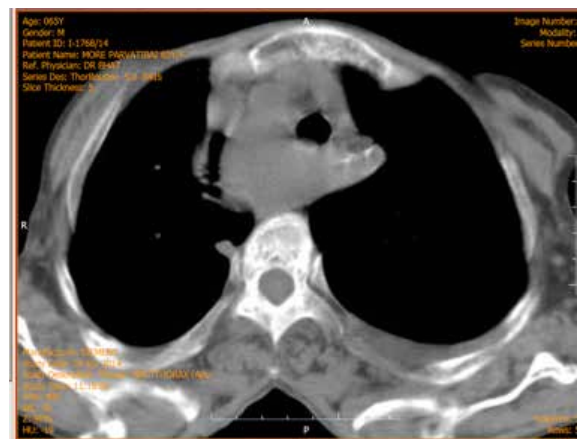
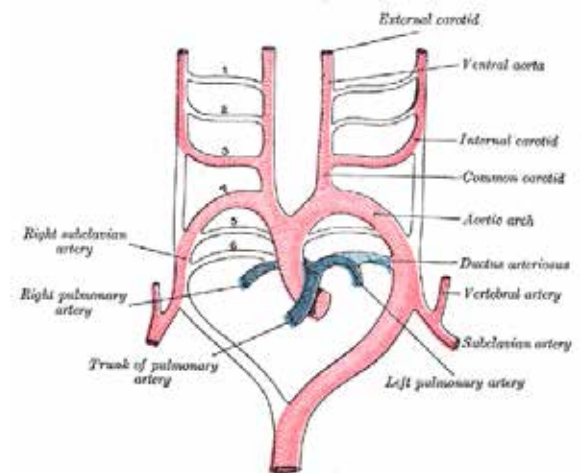


Image 5: Embryonic development of the aorta and its branches (Courtesy: Gray's Human Anatomy: Image in public domain)



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