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Case Series

Radiology Section

Right Sided Aortic Arch and its Rare Associations- A Case Series

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ABSTRACT

Right sided aortic arch is a rare incidental anomaly in adults often asymptomatic. Most common subtype is Right Aortic Arch (RAA) with aberrant left subclavian artery. We report three cases of right sided aortic arch with multiple vascular anomalies. Two cases were showing mirror image branching with situs inversus totalis and Tetrology of Fallot (TOF). Another one showing retroesophageal circumflex aorta with aberrant left subclavian artery ,supra-sinus origin of coronary arteries, bicuspid aortic valve, Persistant Left Superior Vena Cava (PLSVC) and Autosomal Dominant Polycystic Kidney Disease (ADPKD). Through cross sectional CT angiography of the aortic arch and surrounding structures, we defined anatomical relationships, which is useful for follow-up and treatment.

Keywords: Mirror image branching, Retroesophageal circumflex aorta, Situs inversus totalis, Tetrology of fallot,

Aortic arch abnormalities are common cardiovascular anomalies, accounting for 15% to 20% of all congenital cardiovascular diseases [1]. Most arch abnormalities result from errors in the embryologic development of branchial arches which consist of errors of involution, migration or abnormal persistence of vascular structures. They may be symptomatic due to airway or oesophageal compression but most are found incidentally owing to increased use of cross sectional imaging. RAA is a rare incidental entity. It results from dissolution of left dorsal aortic root instead of the usual right. The RAA with aberrant left subclavian artery is the most common subtype and usually exists as an isolated anomaly. RAA can cross midline posterior to oesophagus before descending onto the contralateral side which then is referred to as “circumflex retroesophageal aorta”. The presence of a mirror image RAA can be associated with congenital heart diseases such as pulmonary atresia with Ventricular Septal Defect (VSD) in 46.4%, TOF in 32%, and double outlet right ventricle with right atrial isomerism in 14% of cases. We describe three such cases here associated with RAA anomaly [Table/Fig-1]. These patients had presented for other complaints and RAA with other findings was incidental. CT-scan was performed after informed consent and in patients with normal renal function tests.

Case I	CASE II	CASE III
Situs Inversus Totalis	Tetrology of Fallot	ADPKD
RAA with mirror image branching	RAA with mirror image branching	Retroesophageal circumflex RAA with Aberrant left subclavian artery
	Dilated bronchial arteries	PLSVC, Suprasinus origin of coronary arteries, Bicuspid aortic valve

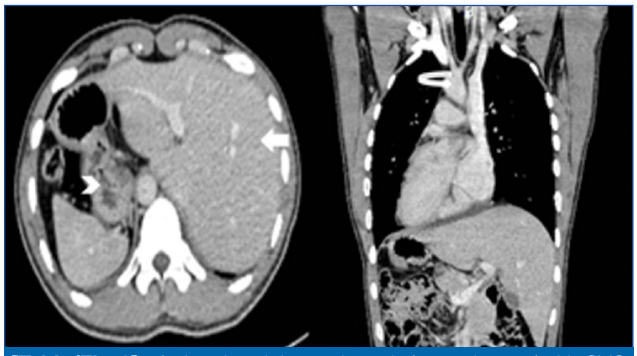
[Table/Fig-1]: Table showing the imaging findings in the above described cases.

CASE 1

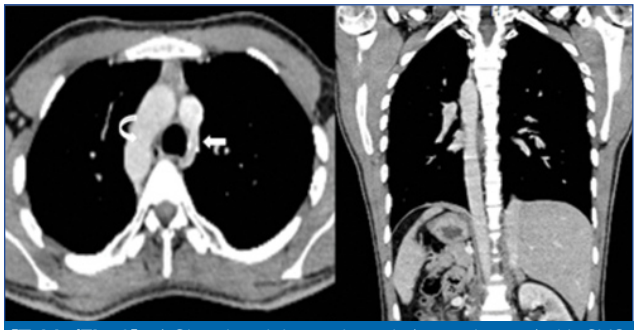
A 52-year-old male patient known for Hodgkin’s disease was referred for CT-scan for post chemotherapy evaluation of lymph nodes. He was found to have situs inversus totalis with cardia, spleen, tail of pancreas, two lobes of lung, aortic arch and descending thoracic aorta on right side, while liver, head of pancreas, three lung lobes and inferior vena cava in left side [Table/Fig-2a,b]. None of the lymph nodes were found to be abnormally enhancing or enlarged. Aorta was seen to arch onto the right side after its origin and descending on the right side of the vertebral bodies [Table/Fig-3a,b] with branches arising from the arch as- left brachiocephalic artery, right common carotid artery arising as a common origin and right subclavian artery as the last branch [Table/Fig-4a,b].

CASE 2

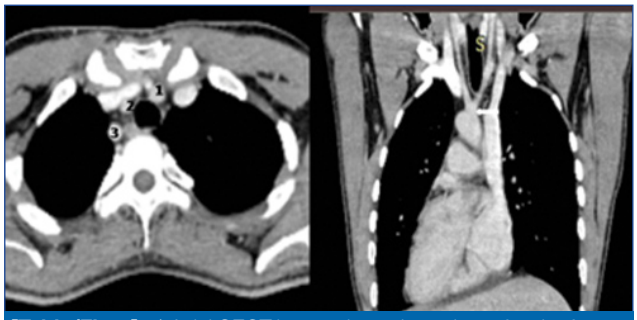
A 15-year-old female patient presented to hospital with complaints of respiratory difficulty on and off since birth.



[Table/Fig-2]: a) showing right aortic arch (curved arrow)with SVC and azygous vein on left (arrow); b) showing descending thoracic aorta on right side.

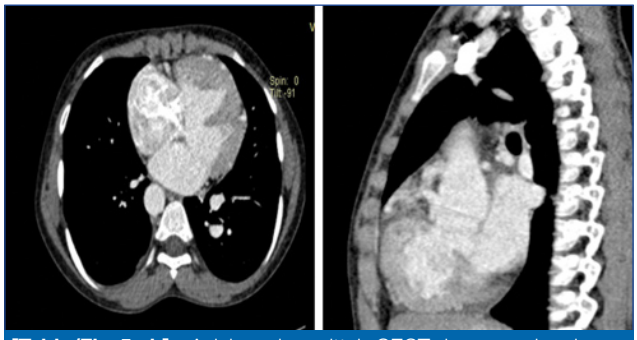


[Table/Fig-3]: a) Showing right aortic arch (curved arrow)with SVC and azygous vein on left (arrow); b) Showing descending thoracic aorta on right side.

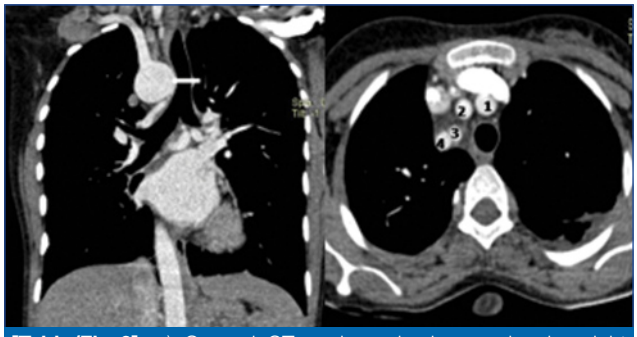


[Table/Fig-4]: a) Axial CECT image shows branches of arch of aorta as 1- Left brachiocephalic artery, 2-Common carotid artery and 3-Right subclavian artery. b) Coronal CECT image shows common origin of left brachiocephalic artery and right common carotid artery (arrow).

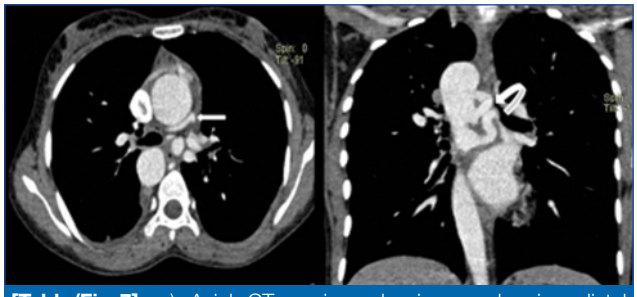
However, no obvious signs of cyanosis were present. 2D Echo revealed right ventricular hypertrophy, over riding of aorta, VSD and pulmonary atresia, findings consistent with TOF. CT thoracic angiography apart from confirming echo findings [Table/Fig-5a,b] showed right sided aortic arch with branching in the sequence as-left brachiocephalic artery, right common carotid artery, right vertebral and right subclavian artery [Table/Fig-6a,b] Proximal part of main pulmonary artery was not properly visualised. However, distal part and bifurcation was seen in normal location with severely narrowed caliber [Table/Fig-7a]. Tortuous vascular channels were seen arising from descending thoracic aorta at level of T5-6 and supplying the lungs consistent with bronchial arteries [Table/Fig-7b].



[Table/Fig-5a,b]: Axial and sagittal CECT images showing a subaortic VSD (arrow) with over-riding of aorta (arrow head).



[Table/Fig-6]: a) Coronal CT angiography image showing right aortic arch on right side of trachea. b)Axial CT angiography image shows branches of aortic arch in the order -1-Left brachiocephalic artery, 2-Right common carotid artery, 3-Right vertebral artery and 4- Right subclavian artery.

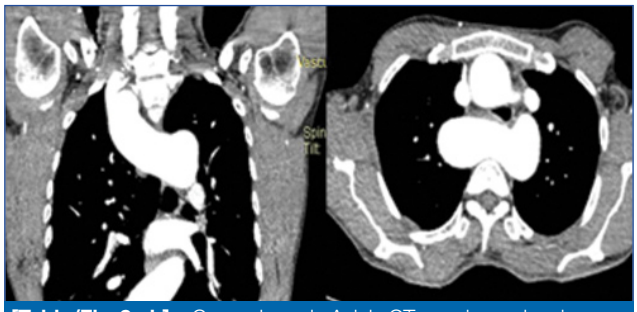


[Table/Fig-7]: a) Axial CT angiography image showing distal main pulmonary artery and its bifurcation (arrow). b) Coronal CT angiography image showing bronchial arteries (curved arrow).

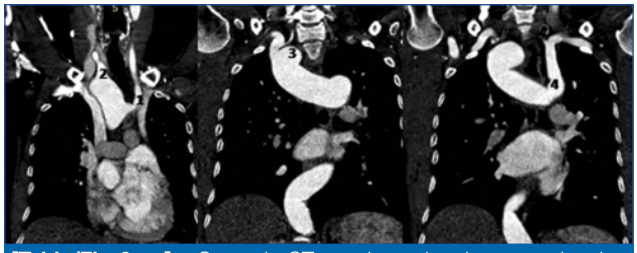
CASE 3

A 45-year-old male patient presented to hospital with complaints of difficulty in swallowing and breathing. Echocardiography showed bicuspid aortic valve and aortic regurgitation and was suspected to have some vascular anomalies for which he was referred to our department for CT angiography of thoracic aorta. CT angiography on a 128-slice multidetector scanner demonstrated right sided aortic arch crossing midline behind the oesophagus to descend on the left side of the

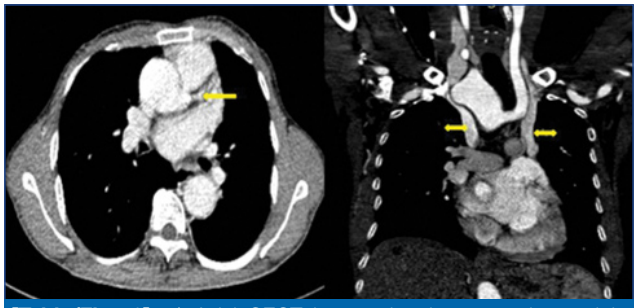
vertebral bodies [Table/Fig-8a,b]. From the aortic arch, vessels originated in the sequence as left common carotid artery, right common carotid artery, right subclavian artery [Table/Fig-9]. There was also suprasinus origin of coronary arteries [Table/Fig-10a]. There were two separate venous channels draining separating right and left upper halves of the body [Table/Fig-10b]. Right draining into right atrium while left into coronary sinus. This was suggestive of persistent left superior vena cava on left side. On abdominal screening, patient was found to have multiple non-enhancing fluid density cystic lesions in liver and bilateral kidneys suggestive of ADPKD [Table/Fig-11].



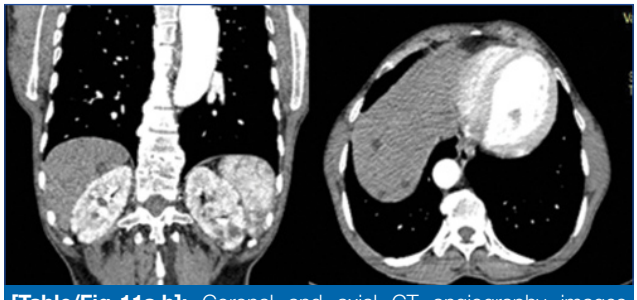
[Table/Fig-8a,b]: Coronal and Axial CT angiography images showing right aortic arch crossing midline posteriorly to descend on the left side (circumflex retroesophageal aorta).



[Table/Fig-9a-c]: Coronal CT angiography images showing branches of the right aortic arch as- 1-Left common carotid artery , 2-Right common carotid artery, 3-Right subclavian artery and 4-Left subclavian artery.



[Table/Fig-10]: a) Axial CECT image showing suprasinus origin of left coronary artery (arrow). b) Coronal CT angiography image showing bilateral SVCs (double headed arrows).



[Table/Fig-11a,b]: Coronal and axial CT angiography images showing multiple non enhancing cystic lesions in bilateral kidneys and liver.

DISCUSSION

Development of the aorta takes place during the third week of gestation [2]. It is a complex process associated with a variety of congenital variants. A right-sided aortic arch was first described by Fioratti and Aglietti [3]. It results from persistence of the right fourth aortic arch with suppression of the corresponding left vessels. The incidence of RAA was reported to be 0.04%-0.1% [4] according to a necropsy series, however actual incidence is probably much higher because of most of cases being asymptomatic. Many cases are detected now-a-days because of increased use of cross-sectional studies like MDCT and MRI. RAA has been classified into various types according to the arrangement of the arch vessels [5,6], relation with oesophagus and the presence of congenital heart anomalies. Of these, RAA with aberrant left subclavian artery is the most common.

On the basis of sequence of origin of its branches, RAA is divided as (Bedford and Parkinson) [7], the mirror-image Type (Type M of Adachi-Williams-Nakagawa) and the form with the left sub-clavian artery originating as the last branch (type N). The most common variant of a RAA is associated with aberrant left subclavian artery [8], in which the first branch arising from the aortic arch is the left common carotid artery, followed by the right carotid, right subclavian, and left subclavian arteries in that order. It is usually associated with an aberrant left subclavian artery arising either as the last branch of the right sided aortic arch or from an aortic diverticulum, known as a Kommerell's diverticulum, which is a remnant of the left dorsal aortic arch. It is more common and in only 5-10% associated with congenital heart anomalies. It may cause symptoms of tracheal or oesophageal compression because of its course, such as dysphagia, dyspnoea, stridor, wheezing, cough, choking spells, or chest pain [9].

Case 1

RAA with mirror image branching is the second most common form of a right sided arch, after right arch with aberrant left subclavian artery [9]. In this, left subclavian artery originates together (as innominate artery) or proximal to left common carotid artery. It results from break in the left fourth arch between left ductus and descending aorta with regression of the dorsal left arch between left ductus and descending aorta. It is usually not associated with any symptoms of compression of trachea or oesophagus, and can be found as an incidental finding.

Situs inversus totalis which is described as total transposition of abdominal and thoracic viscera (mirror image of internal organs) is associated with dextrocardia and only 3-5% incidence of congenital heart diseases. About 80% of these patients have associated right sided aortic arch as was seen in our case. As a general rule, in dextrocardia and levocardia associated with situs solitus or inversus, the atria follow the position of the viscera [10] and the aortic arch is usually opposite the side of the venous atrium. In our case, there was dextrocardia with morphological right atrium and liver on left side, whereas, aortic arch with descending thoracic aorta was on the right side.

Case 2

RAA with mirror image branching is associated with congenital heart anomalies in approximately 75% of these patients, including TOF, pulmonary stenosis with VSDs, tricuspid atresia, and truncus arteriosus. Fifty-five years after the description of right sided aortic arch, Corvisart (1818) [11] reported its occurrence in a case of tetralogy of fallot. The association of a RAA with TOF (Corvisart's disease) is well known since then. It varies from 13 to 34% in different series [12-14]. Hastreiter AR et al., reported the diagnostic importance of RAA accompanying a TOF complex in 34% of their 167 patients with this anomaly [4]. So a detailed evaluation of cardiac chambers is required. In our case, RAA with mirror image branching was associated with features of TOF and presented with respiratory difficulty at 15 years of age.

Case 3

Depending on the relation of right arch to the oesophagus, right arch can either remain on right of oesophagus and descend on the right side of the spine or infrequently it can cross midline behind the oesophagus to descend on the left side [15]. The latter is known as "circumflex retroesophageal RAA [4,16] "and is most commonly symptomatic due to oesophageal compression as was in our case. Shuford WH et al., presented radiologic features of three such cases [6]. They used barium swallow, aortography and CT to diagnose such cases. There was bilateral symmetric enlargement of the superior mediastinum, simulating a mass in all three cases. They also revealed the branching of the arch vessels which was typical of RAA with aberrant left subclavian artery. Since, most radiologists are unfamiliar with the appearance of a circumflex RAA, this anomaly may not be considered in the differential diagnosis of a widened mediastinal shadow. In another study conducted by Hastreiter AR et al., out of 116 patients with RAA, nine had a circumflex arch with left descending thoracic aorta [4].

A right sided aortic arch can present with anomalies of the coronary arteries [11]. Lee JKT et al., reported a case of right-sided aortic arch with supra-sinus origin of coronary arteries and coronary arteriovenous fistula [17]. In our case also, patient along with retroesophageal circumflex RAA showed suprasinus origin of coronary arteries.

Persistent left SVC (PLSVC) is the most common congenital thoracic venous anomaly with a prevalence of 0.3-0.5% in the general population resulting from failure of regression of the caudal portion of left superior cardinal vein. Such a vein empties into coronary sinus. PLSVC can occur with or without the presence of right SVCs, with presence of both right and left SVCs being more common. Majority of patients with PLSVC have associated cardiac anomalies [18] such as atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, and cor triatriatum. In our study, PLSVC with coexistence of right SVC was found on CT. Patient was found to have bicuspid aortic valve and aortic regurgitation on 2D ECHO.

ADPKD is the most common hereditary kidney disease with multiple cysts in kidneys and various organs leading to renal failure. They are known to be associated with aneurysms and dissections of various vessels including aorta, its branches and intracranial vessels as documented by Perrone RD et al., [19]. However, to the best of our knowledge no association with right sided aortic arch or retroesophageal circumflex aorta has been documented as was seen in our case. This could be a co-incidental finding as well.

The mirror image RAA associated with cyanotic congenital heart disease requires the repair of cardiac anomalies. The isolated mirror image RAA without associated cardiac anomalies does not require surgical treatment. In our first case of RAA with situs inversus, no treatment was required for the vascular anomalies. However, in second case of TOF with RAA, patient was referred to Cardiology Department for further management.

In retroesophageal circumflex aorta, with symptoms secondary to compression of the trachea or oesophagus, a meticulous analysis of vascular structures using 3D CT angiography is required for detailed evaluation of the vascular anomalies. Imaging of the aorta in such cases can improve accuracy of diagnosis of multiple vascular anomalies and facilitate endovascular or surgical planning. In our case, patient was referred to higher centre for further management.

CONCLUSION

Majority of the vascular anomalies of aorta are incidentally found on imaging studies. Knowledge of these anomalies may lead to search for other associated anomalies and has an impact on the surgical management of patients. CT angiography with 3D reconstructions could be the gold standard for detecting multiple vascular malformations and their relationships with adjacent structures in these patients.

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