

Situs inversus totalis with Congenitally Corrected Transposition of the Great Arteries - Case Report & Review of Literature

Dr. Anagha Joshi¹, Dr. Sneha Deshpande Pai², Dr. Aakash Vaswani³

¹Professor & Head, Department of Radiology, LTMMC & GH – Sion, Mumbai, Maharashtra, India.

²Assistant Professor, Department of Radiology, LTMMC & GH – Sion, Mumbai, Maharashtra, India

³Resident, Department of Radiology, LTMMC and GH – Sion, Mumbai, Maharashtra, India

Abstract: Congenitally corrected transposition of great arteries with situs inversus totalis is a rare congenital anomaly, which can present in adulthood. This case report describes a 36 year old female with dyspnea on exertion and intermittent palpitations with a diagnosis of congenitally corrected transposition of great arteries with situs inversus totalis, which was established by 'one-stop-shop' CT coronary angiography with detailed evaluation of anatomy of cardiac chambers and coronary anatomy, essential for surgical correction.

Keywords: Situs inversus totalis, Cardiac CT, Congenitally corrected transposition of the great arteries, Congenital heart disease

1. Introduction

Congenitally corrected transposition of great arteries (ccTGA) is a rare anomaly representing approximately 0.5% of all congenital heart diseases. ccTGA is characterised by a combination of atrio-ventricular (AV) and ventriculo-arterial (VA) discordance, with or without associated anomalies [1]. Situs inversus totalis associated with congenitally corrected transposition of great arteries represents an extremely rare congenital anomaly, representing approximately 34% of ccTGA [2].

2. Case Description

A 36 year old female presented with dyspnoea on exertion (grade I) since 1 year and intermittent episodes of palpitation since 6 months. She had no history of chest pain, pedal edema or orthopnoea. No significant past surgical or medical history was mentioned.

Chest radiograph revealed dextrocardia with right sided aortic arch. No significant abnormality was noted in the lung fields. Fundic bubble was noted on right side. ECG was suggestive of atrioventricular re-entrant tachycardia, a type of supraventricular tachycardia. Echocardiography was suggestive of complex congenital heart disease with dextrocardia. An atrio-ventricular and ventriculo-arterial discordance was suspected. A small (4 mm) Ossecundum ASD was noted. No significant valvular stenosis or regurgitation was noted. Interventricular septum was normal.

Patient underwent cardiac CT in our department for further evaluation. Retrospective ECG-gated cardiac CT was performed with a 160-slice multi-detector CT system (Aquilion™PRIME 160 slice CT scanner, Toshiba, Japan).

A standardized protocol was followed with an ECG-dependent tube current modulation (10-90% RR-interval). 80 mL of non-ionic contrast (Omnipaque; 370mgI/mL) was injected at a rate of 5 mL/s through an 18 G intravenous antecubital catheter using a bi-phasic contrast delivery protocol.

CT revealed features of situs inversus totalis in the form of liver being positioned on the left side, while spleen & stomach on right side (Figure 1), bi-lobed right lung and tri-lobed left lung (Figure 2) with dextrocardia and right sided aortic arch and mirror image branching of arch vessels.



Figure 1:Visceral situs: Axial post contrast CT image showing liver positioned on the left while stomach and spleen on the right side.

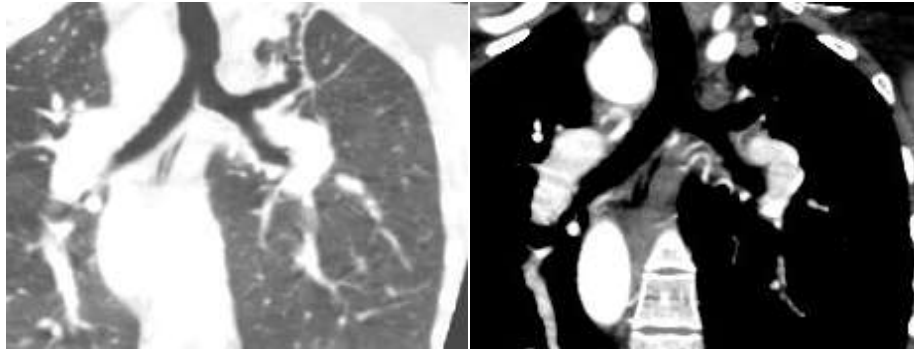


Figure 2: Bronchial situs: Early branching of the left main bronchus with eparterial bronchus on left and hyperarterial bronchus of the right

Mild cardiomegaly was noted. Superior vena cava (SVC) and Inferior vena cava (IVC) were seen draining into the morphological right atrium (which was anatomically positioned on left side) (**Figure 3**). Four pulmonary veins were noted draining into morphological left atrium (which was anatomically positioned on right side). Thus, veno-atrial concordance was established.



Figure 3: Coronal post contrast image showing SVC and IVC draining into atrium on left side (Morphological right atrium).

Moderator band, septal papillary muscle and prominent trabeculae were seen in the ventricle which was anatomically positioned on right side suggestive of morphological right ventricle (**Figure 4**).

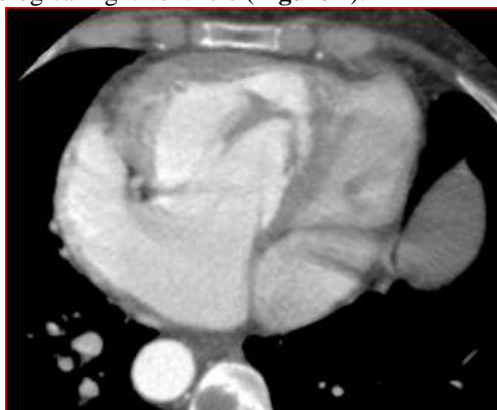


Figure 4: Moderator band with septal papillary muscle and prominent trabeculae in ventricle on right side of the septum suggestive of morphological right ventricle

The ventricle which was anatomically positioned on the left side of the septum showed fewer trabeculae and was labelled as morphological left ventricle. Right atrium was seen opening into morphological left ventricle and left atrium was opening into morphological right ventricle, suggestive of atrio-ventricular discordance. Morphological left ventricle was giving rise to pulmonary trunk and morphological right ventricle was giving rise to aorta, suggestive of ventriculo-arterial discordance (**Figure 5**). This led to a diagnosis of congenitally corrected transposition of great arteries (ccTGA). Other findings included right sided aortic arch with mirror image branching of the arch vessels and dilated pulmonary arteries (due to pulmonary hypertension). A small (4mm) sized Ossecundum ASD was also noted.

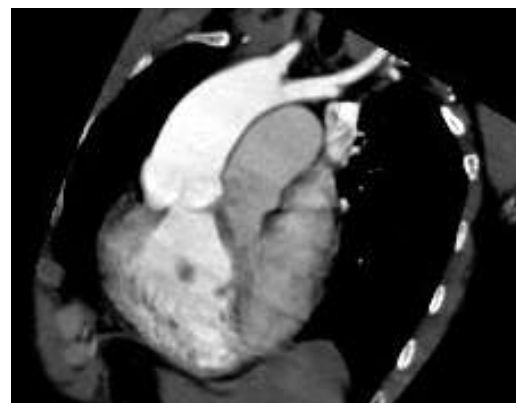


Figure 5: Oblique coronal image showing the aorta arising from morphological right ventricle & pulmonary trunk arising from morphological left ventricle.

Coronary artery anatomy was studied which showed left and right main coronary arteries arising separately from middle of the left and right posterior sinuses respectively and then curving to the right atrio-ventricular and anterior interventricular groove respectively (**Figure 6**); Type A by Yacoub and Radley-Smith classification system. Left main coronary artery was seen trifurcating into anterior descending artery, ramus intermedius and circumflex artery. Type I anterior descending artery was noted not reaching upto the apex. Circumflex artery was seen giving a single obtuse marginal branch (OM1). No malignant interarterial or intramural coronary course between Aorta and pulmonary artery was noted. Morphological right ventricle was supplied

by anterior descending artery. Morphological left ventricle was supplied by circumflex and OM1 branch.

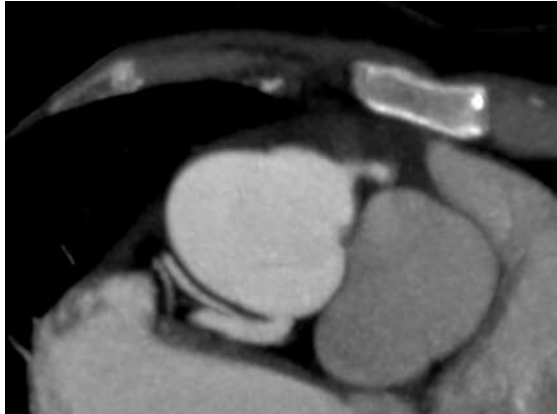


Figure 6: Aorta is positioned anterior and to the right of pulmonary artery. Left and right main coronary arteries are seen arising separately from middle of the left and right posterior sinuses respectively.

The patient then underwent cardiac catheterization, and the above findings were confirmed.

3. Discussion

Our patient was diagnosed as situs inversus totalis with congenitally corrected transposition of great arteries, a complex congenital heart anomaly with atrio-ventricular and ventricular-arterial discordance.

'Situs' refers to the anatomical positioning of normal asymmetric thoracic and abdominal structures. It is classified into three types: situs solitus (normal), situs inversus (mirror image of normal anatomical placement), and situs ambiguus.

In situs solitus, the right lung has three lobes with an eparterial bronchus which shows early origin of the upper lobe bronchus, whereas the left lung has two lobes with a hyparterial bronchus which shows a more distal origin of the upper lobe bronchus. Morphological right atrium is positioned to the right of morphological left atrium with levocardia. Morphological right atrium receives blood from the inferior vena cava and shows an appendage which is broad-based with coarser pectinate muscles. Morphological left atrium shows an appendage which is smaller and narrower. It receives blood from the pulmonary veins. Liver and gallbladder are positioned to the right, and stomach and spleen are on the left. The incidence of congenital heart disease in patients with situs solitus and levocardia is only 0.6%–0.8% [2].

Situs inversus which is seen in 0.01% of the population, refers to an anatomic arrangement that is the mirror image of situs solitus. With situs inversus, the left lung has three lobes with an eparterial bronchus and the right lung has two lobes with a hyparterial bronchus. Morphological right atrium is to the right of the morphological left atrium. Liver and gallbladder are positioned towards left while stomach and spleen are towards right. It is important to note that with situs solitus and situs inversus, the atrial situs always corresponds to the

visceral situs [3, 4]. 'Situs ambiguus' (termed "heterotaxy") refers to the third type of situs, which consists of various abnormal viscerocardiac configurations which do not entirely correspond to the complete or partial mirror image.

Situs inversus occurs more commonly with dextrocardia. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch [3, 5].

Congenitally corrected transposition of the great arteries (ccTGA or l-transposition), which is characterized by atrio-ventricular and ventriculo-arterial discordance accounts for less than 1% of all congenital cardiac anomalies. During embryological development, left-handed looping (l-loop) of the heart tube results in atrio-ventricular (AV) discordance, while failure of aorto-pulmonary septum to rotate through 180° results in ventriculo-arterial discordance. The atrio-ventricular valves follow their respective ventricles. The aortic valve is located more anterior and to the left of the pulmonary valve. Venous blood returns to the right atrium which passes through the mitral valve into morphological left ventricle. Blood then enters the pulmonary circulation via the pulmonary valve. Pulmonary venous blood returns to the left atrium which then flows through the tricuspid valve to the morphological right ventricle, exiting into the aorta via the aortic valve. Blood flows in an effective sequence, hence the name corrected; however, the right ventricle supports the systemic circulation in this disorder. [6]

Most patients with ccTGA have at least one additional cardiac abnormality which may consist of pulmonary stenosis (41%), ventricular septal defects (37%), atrial septal defects (19%), Ebstein's anomaly (7%) etc. [7]. The finding of ccTGA without significantly associated anomaly is much less frequent. Prognosis depends on AV conduction, arrhythmias, structural abnormalities and degree of hemodynamic disturbance. This congenital cardiac defect remains often asymptomatic until adulthood, which later presents with heart failure and cardiac arrhythmias. This is caused by progressive failure of the morphological right systemic ventricle that is burdened with high systemic pressures. This may be related to the coronary perfusion mismatch in the systemic right ventricle supplied by a single coronary artery as well as from the differences in the right ventricular fibre orientation, geometry, and microscopic structural features when functioning as the systemic ventricle [8, 9].

Multiple significant coronary artery anomalies have been described in literature with ccTGA [10, 11, 12]. For success of surgical correction in ccTGA patients, the identification of coronary anatomy is extremely important as it is essential to transfer coronary ostia without undue tension, torsion or kinking of proximal parts of coronary arteries or their early branches [13,14].

Multiple previous classifications of coronary artery anatomy in transposition of great arteries have been published, but they are not sufficient in themselves as they are limited to pathological or surgical case series. One of the popular

coronary artery anatomy classification by Yacoub MH and Radley-Smith describes 5 anatomical types of coronary arteries based on origin, orientation of coronary ostia along with course and length of main coronary artery. Type A is classified as the right and left coronary ostia arising from the middle of the right and left posterior aortic sinuses and curving forwards to reach the right atrioventricular groove or anterior interventricular groove respectively. Both coronary arteries arising by a single ostium is classified as type B, while in type C the two coronary ostia are situated posteriorly, very close to each other, in a position similar to that in type B. The origin of the coronary arteries in type D is similar to that of type A. However, the right coronary artery gives origin to the circumflex coronary artery that curves round the pulmonary vessel to reach the atrioventricular groove. In type E the right coronary artery arises in common with the left anterior descending artery from the left posterior sinus, while the circumflex artery arises separately from the right posterior sinus. However, this classification is not comprehensive and limited to ccTGA patients only. [14]

A new classification of great vessel and coronary artery anatomy in transposition and other coronary anomalies has been described by Sithampanathan S et al. using cardiac CT which includes both descriptive classification and an alphanumeric classification. This simple, universal codex overcomes limitations of previous classifications as it allows complete delineation of the coronary anatomy, anatomical relationship of the aorta to the pulmonary artery, as well as application to all congenital heart disease patients. It also provides the prognostic information by description about interarterial malignant course and the number of coronary ostia in each aortic sinus. [15]

4. Conclusion

Situs inversus with congenitally corrected transposition of great arteries is a rare anomaly which usually presents in late adulthood, unlike other congenital heart diseases, with cardiac failure or arrhythmias. Accurate delineation of cardiac chambers and coronary arterial anatomy is a prerequisite for successful management. Cardiac CT acts as a 'one-stop-shop' for evaluation of this anomaly. The information that can be obtained is useful for timely introduction of adequate therapy and allowing optimal treatment with improved quality of life in patients with incidentally detected congenital heart defects.

References

- [1] Hornung TS, Calder L Congenitally corrected transposition of the great arteries *Heart* 2010;96: 1154-1161.
- [2] Kandice Mah and Mark K Friedberg Congenitally Corrected Transposition of the Great Arteries *Circulation: Cardiovascular Imaging*. 2014;7:849-851
- [3] Maldijian PD, Saric M. Approach to dextrocardia in adults: Review. *Am J Roentgenol*. Jun 2007; 188(6 suppl):539-49
- [4] Van Praagh R. The importance of segmental situs in the diagnosis of congenital heart disease. *Semin Roentgenol* 1985; 20:254-271
- [5] Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: Imaging of heterotaxy Syndrome. *Radiographics* 1999; 19 (4): 837- 52.
- [6] Gunjan Gulati, Banashree Das, Akanksha Mangla Congenitally corrected TGA- A case diagnosed incidentally *Asian Pac. J. Health Sci.*, 2015; 2(2): 56-58
- [7] C. A. Warnes, "Transposition of the great arteries," *Circulation*, 2006 vol. 114, no. 24, pp. 2699-2709, 2006.
- [8] Graham TP Jr, Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol* 2000;36:255-261.
- [9] Heon Lee, MD, Byoung-Won Park, MD A Case Report of Congenitally Corrected Transposition of Great Arteries: Morphologic and Functional Evaluation with Cardiac CT *J Korean Soc Radiol* 2013;69(3):197-200
- [10] Neufeld HN, Schneeweiss A. *Coronary Artery Disease in Infants and Children*. 1st ed. Philadelphia, PA: Lee and Febiger; 1983. p. 91.
- [11] Dabizzi RP, Barletta GA, Caprioli G, et al. Coronary artery anatomy in corrected transposition of the great arteries. *J Am Coll Cardiol*. 1988;12:486-91.
- [12] McKay R, Anderson RH, Smith A. The coronary arteries in hearts with discordant atrioventricular connections. *J Thorac Cardiovasc Surg*. 1996;111:988-97.
- [13] S.K. Pasquali, V. Hasselblad, J.S. Li, D.F. Kong, S.P. Sanders Coronary artery pattern and outcome of arterial switch operation for transposition of the great arteries: a meta-analysis *Circulation*, 106 (2002), pp. 2575-2580
- [14] Yacoub MH, Radley-Smith R. Anatomy of the coronary arteries in transposition of the great arteries and methods for their transfer in anatomical correction. *Thorax*. 1978;33(4):418-424.
- [15] Sithampanathan S, Padley SP, Rubens MB, Gatzoulis MA, Ho SY, Nicol ED. Great vessel and coronary artery anatomy in transposition and other coronary anomalies: a universal descriptive and alphanumeric sequential classification. *JACC Cardiovasc Imaging* 2013;6:624-630.