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Research Article

ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERY IN A PATIENT WITH SITUS INVERSUS TOTALIS

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ABSTRACT

Anomalous Aortic Origin of Coronary Artery is rare congenital anomaly seen in 0.1 – 0.2 % of general population. Situs inversus with dextrocardia is known as situs inversus totalis. Authors report a case of anomalous aortic origin of right coronary artery from left sinus of Valsalva in a patient with situs inversus totalis , who also had other anomalies like ventricular septal defect and persistent superior vena cava.

INTRODUCTION

Anomalous Aortic Origin of Coronary Artery (AAOCA) is rare congenital anomaly. Anomalous origin of Right Coronary Artery (RCA) from left sinus of Valsalva is more commoner and has benign natural history when compared to the left coronary artery originating from right sinus, which has got more malignant natural history. Situs inversus with dextrocardia is known as Situs Inversus Totalis (SIT) or Situs transversus. AAOCA is very rarely seen in SIT[1,2,3]. We report a case of anomalous origin of RCA from left sinus of Valsalva in a patient with SIT who also had other anomalies, perimembranous ventricular septal defect and additional Superior Vena Cava (SVC) present on right side draining into coronary sinus.

Case Report

A 26 years old male was diagnosed to have SIT with ventricular septal defect, was referred to our institution for intracardiac repair. He presented with history of dyspnea on exertion (NYHA-Class-II) since 9 months. On examination jugular venous pressure was normal, hyperdynamic right precordium, holosystolic murmur over right parasternal area with clear lung fields.

Electrocardiogram findings were consistent with dextrocardia. ***Chest X-Ray:*** Dextrocardia with pulmonary plethora, liver shadow on left side and fundal gas on right side.

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Transthoracic echocardiogram: Dextrocardia, large preimembranous ventricular septal defect with septal aneurysm, mild tricuspid regurgitation, persistent SVC present on the right side draining into coronary sinus, left main coronary artery and RCA originating from left Sinus of Valsalva.

Trans-esophageal echocardiogram confirmed trans thoracic echocardiogram findings (Figure1).

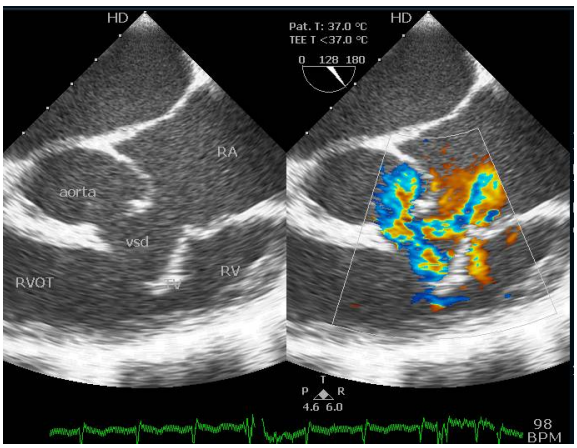

Figure 1 Trans esophageal echocardiogram: Perimembranous ventricular septal defect with septal aneurysm.

Computed Tomography Thorax and Abdomen: Reversal of abdominal viscera with no evidence of bronchiectasis (Figure 2)

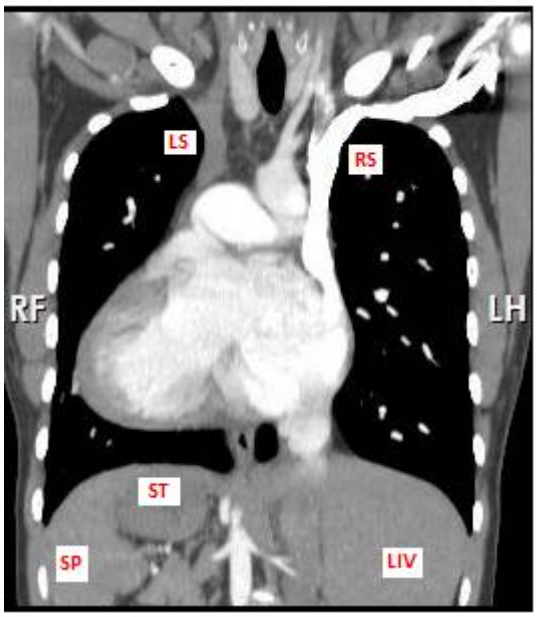

Figure 2 Computed Tomography of thorax and abdomen (Coronal view): LS - Left SVC, RS - Right SVC, ST - Stomach, Liv - Liver, SP - Spleen.

Computed Tomography coronary angiogram: Left main coronary artery arising from left sinus of valsalva deviding into left anterior descending and left circumflex artery. RCA was also arising from the left sinus very close to the origin of left main ostia. There was no intramural segment and it had normal ostia. After its origin there was no angulation or kinking, it was coursing between aorta and main pulmonary artery before entering right atrio-ventricular groove (Figure3).

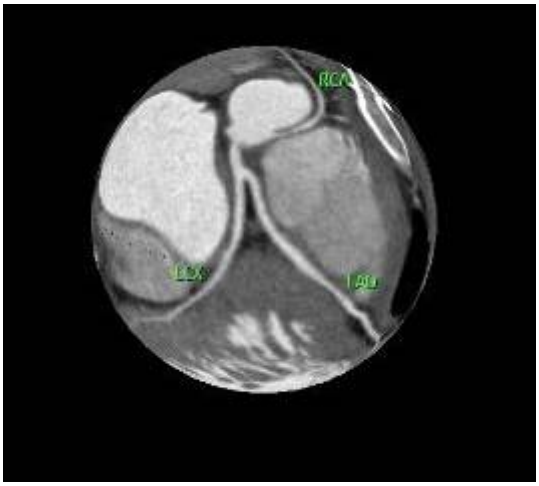

Figure 3 Computed Tomography Coronary Angiogram: Left main coronary artery and RCA arising from left sinus of valsalva. Right coronary ostia very close to the left main ostia. RCA coursing between aorta and main pulmonary artery, before entering right atrioventicular groove. RCA - Right Coronary Artery; LCX - Left Circumflex Artery; LAD - Left Anterior Descending Artery.

This patient underwent intra cardiac repair (only Ventricular Septal Defect was addressed). Since anomalous RCA had normal ostia with no intramural segment, and there was no angulation or kink, only option was to reimplant RCA. Anomalous RCA was not addressed as it was not suitable for reimplantation, since RCA ostia was very close to the left main ostia(insufficient tissue to create a button around the ostia). Post operative period was uneventful and he was discharged on sixth post operative day. He was asymptomatic during subsequent follow ups.

Comment

Situs inversus with dextrocardia is known as SIT. It is a rare congenital anomaly with an estimated incidence of 1:10000. Here all the major visceral organs are mirror imaged from their normal position. Incidence of cardiac anomalies is rare and seen in 3-5 % of the cases. Commonly seen cardiac anomalies are ventricular septal defect, pulmonary stenosis, atrial septal defect. Commonly associated extra-cardiac anomalies are asplenia, multiple spleens, duodenal atresia, horse shoe kidney, ectopic kidney.

Cardiac Situs is determined by the position of atria. In situs inversus morphologic right atrium is on the left and morphologic left atrium on the right. Merklin and Varano studied 111 cases and classified situs inversus into: (a) complete situs inversus; (b) dextrocardia with situs solitus; (c) partial situs inversus; (d) dextro-position of the heart; and (e) levocardia[4].

20-25% of individuals with SIT have features of primary ciliary dyskinesia and is known as Kartgener Syndrome[5]. Kartgener syndrome is a autosomal recessive disorder characterized by triad of situs inversus, chronic sinusitis, bronchiectasis.

In this case patient had multiple cardiac anomalies, anomalous aortic origin of RCA, peri-membranous ventricular septal defect and additional superior vena-cava (present on the right side draining into coronary sinus).

AAOCA is a rare congenital anomaly seen in 0.1-0.2% of general population. Anomalous origin of RCA from left sinus of Valsalva is six times more prevalent than anomalous origin of left from right sinus. Anomalous left coronary artery is responsible for upto 85% of sudden cardiac deaths related to AAOCA, hence it is more lethal than anomalous RCA.

AAOCA may remain asymptomatic or present with symptoms of myocardial ischemia or sudden cardiac death. Most of the symptoms or sudden cardiac death follow strenuous exercise. Possible mechanisms of myocardial ischemia are, (A) Inter-arterial compression between aorta and main pulmonary artery or right ventricular outflow tract. (B) Compression of intramural portion, due to aortic expansion during excercise. (C)Kinking or angulation of anomalous artery as it leaves sinus of Valsava. (D) Slit like ostium or flap over the ostium.

Management options are conservative and surgical. Conservative treatment is indicated in older patients with anomalous RCA from left sinus, with no angulation, no intramural segment also when the anatomy is not favourable for surgical correction. It includes use of beta-blockers and limitation of physical activity. Surgical options depends on the anatomy. It includes reimplantation, un-roofing, creation of neo-ostium, pulmonary artery translocation, coronary artery bypass.

Reimplantation is indicated when there is no intramural course and separate orifices which are well separated. It is unsuitable if both the orifices are very close to each other, because of insufficient tissue to create a button around the orifice. Unroofing is indicated when there is a intramural course, slit like orifice, kinking or narrowing as it exits from

aorta. Unroofing eliminates the flap and slit like orifice, widens the narrowed intramural artery thus preventing compression and kinking during systole. Unroofing sometimes may require detachment and resuspension of the commissure when intramural segment is distal to the commissure, which may result in aortic regurgitation. To avoid this complication other option is to create a neoostium between the commissure and distal portion of intramural artery and then obliterate the portion of the artery between the commissure and the original ostium.

Pulmonary artery translocation (Anterior or lateral) is indicated when there is a single ostium or single coronary artery with no intramural course.

Coronary artery bypass should be reserved for those with concomitant obstructive atherosclerotic coronary artery disease. In absence of coronary artery disease bypass results in early graft failure due to competitive flow phenomena.

Till now only five cases of AAOCA in SIT have been reported. Three found during angioplasty had single coronary artery [1], one detected during evaluation of aortic regurgitation [2], and one discovered at autopsy [3].

In SIT, SVC is positioned on the left side and drains into morphologic right atrium. If additional SVC is present on the right side it is considered as combined anomaly. It usually drains into right atrium through coronary sinus, rarely into left atrium through unroofed coronary sinus. In our case additional SVC was present and it was draining into morphologic right atrium through coronary sinus.

To the best of our knowledge till now only five cases of AAOCA in SIT have been reported and also very few cases of SIT with multiple cardiac anomalies have been reported.

Conflict of interest: None declared.

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