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Rare Presentation of Congenitally Corrected Transposition of Great Arteries

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ABSTRACT

Congenitally corrected transposition of great arteries is a rare congenital heart defect with varied presentation. It can be fatal in infancy, to heart failure or complete heart block in adults. Clinical presentation depends on associated defects. Coronary artery anomalies are more frequent in CCTGA. We present a case of 43-year-old man with dextrocardia CCTGA asymptomatic all life present to us with non-ST segment elevation MI. He had no additional defect except systemic ventricular defect with moderate regurgitation of tricuspid valve. He was managed with early invasive therapy with coronary angiogram followed by percutaneous coronary intervention.

Learning Objective: Diagnosis of acute coronary syndrome in adult congenital heart disease needs expertise in both coronary and congenital heart disease fields. CCTGA is very rare congenital heart disease with varied presentation depending upon associated defects. Coronary intervention in CCTGA with dextrocardia requires technical and fluoroscopic anatomical orientation of heart. Radial access with coronary diagnostic and interventional procedures is feasible and drug eluting coronary stents can be used to manage occlusive atherosclerotic lesions.

Keywords: CCTGA, ACS, Congenital heart disease, Dextrocardia, Coronary artery disease, Radial intervention

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INTRODUCTION

CCTGA is a rare congenital heart disease.(1) Asymptomatic adult patient can present rarely with acute coronary syndrome. Previously there are rare case reports of patients with situs solitus CCTGA with ACS due to obstructive or non-obstructive CAD or due to anomalous coronary artery.(7,8) We present a case of isolated dextrocardia with CCTGA presenting as non-ST segment elevation myocardial infarction due to atherosclerotic obstructive CAD and was managed percutaneous intervention through radial access.

CASE PRESENTATION

43-year-old man presented with one day history of chest pain. Electrocardiogram showed left ventricular hypertrophy reverse progression of R wave in chest leads. ECG was taken with chest leads reversal which showed normal progression of R waves in chest leads. There were absent septal q waves [Figure 1(a)]. Echo showed situs solitus dextrocardia with atrioventricular and ventriculo-arterial discordance [Figure 1(b)]. Malposed great arteries with no stenosis for systemic ventricular out flow track. There was no ventricular septal defect. Systemic ventricle that is right ventricular hypertrophy was there with moderate regurgitation of tricuspid valve. He had dyslipidemia with Total cholesterol of 218mg/dl, LDL cholesterol of 172mg/dl. Hemoglobin was 15.1mg/dl and normal serum Right radial access was taken, and we engaged one coronary artery arising from right sided aortic sinus. Angiogram showed it to be left coronary artery (LCA) bifurcating into left anterior descending (LAD) running over interventricular grove between ventricles and left circumflex (LCX) (Figure 2 a,b). Both LAD and LCX were normal. Right coronary artery (RCA) was hooked from left coronary sinus which showed 95% tubular stenosis of acute marginal branch to morphological right ventricle (Figure 2 c,d). After informed consent and explaining the prognosis of the cardiac condition, patient underwent PTCA to acute marginal branch(Figure 2 e,f). RCA was initially tried to hook with Judkins right catheter but was not easily hooking so Judkins left guiding catheter was used. Lesion was crossed with 0.014" floppy wire and stented with 3 *18mm Xience prime. Cardiac CT was done which showed situs solitus dextrocardia CCTGA with posterior left sided coronary sinus was giving rise to RCA which was running in left AV groove with RV branches and stent in situ in one of major RV branch. The right anterior sinus was giving rise to left main coronary artery which bifurcated into left anterior descending and circumflex branches (Figure 3a-f). Patient was discharged in hemodynamically stable condition. On two months follow up patient is asymptomatic.

DISCUSSION

CCTGA is rare congenital anomaly. Its incidence is around 0.03 per 1000 live births and about 0.05% among congenital heart diseases. (1) It was first described by Von Rokitansky in 1875. Embryologically it is due to L-looping of ventricles rather than D-looping.(2) This results in ventricular inversion with right atrium draining the systemic venous blood connects to left ventricle (LV) which in turn connected to pulmonary artery. Left atrium draining pulmonary veins is connected to right ventricle (RV)through tricuspid valve,

which in turn connected to aorta. As result RV becomes systemic ventricle in place LV. If no additional defect present, patients will be asymptomatic in childhood and adolescence. But since RV is not morphologically designed to handle systemic pressures gradually fails which is exacerbated by regurgitation from abnormal tricuspid valve, ultimately leading to heart failure. 30% of patients develop complete heart block due to conduction disturbances. CCTGA can be situs solitus or situs inversus, the later being accounting for 34% of all cases. (3) Isolated dextrocardia with ccTGA is very rare. Embryologically L-looping of dextra-posed primitive heart tube should occur to maintain normal relationship. If D-looping occurs, then CCTGA configuration results. Complication rates of CCTGA with situs solitus is higher compared to situs inverses (In our case dextrocardia) due to abnormal septal alignment and hemodynamics.(4) Coronary arteries are usually follow ventricles.(5) Most common anomalies being single coronary artery.(6) Atherosclerotic coronary artery disease is very rare since survival to adult hood is less and usual cause of death being heart failure. Previous reported two cases have smoking and hypertension being risk factors in our case patient had dyslipidaemia. (7,8) Challenges in intervention being choosing the guiding catheter and angiographic views to delineate the lesions. Since RCA was supplying systemic ventricle and it was dysfunctional globally also, we decided for revascularization of RV branch to restore adequate blood supply. Fluoroscopic horizontal axis mirror image inversion techniques have been described in cases of dextrocardia. We used modified LAO-Cranial and RAO views for intervention. Radial access is feasible in selected patients for coronary intervention. Tiger catheter and Judkins left catheters are useful in engaging both left and right coronary ostia. DES is preferable option unless planned for additional cardiac surgery.

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CONFLICT OF INTEREST

The Authors declare that there is no conflict of interest.

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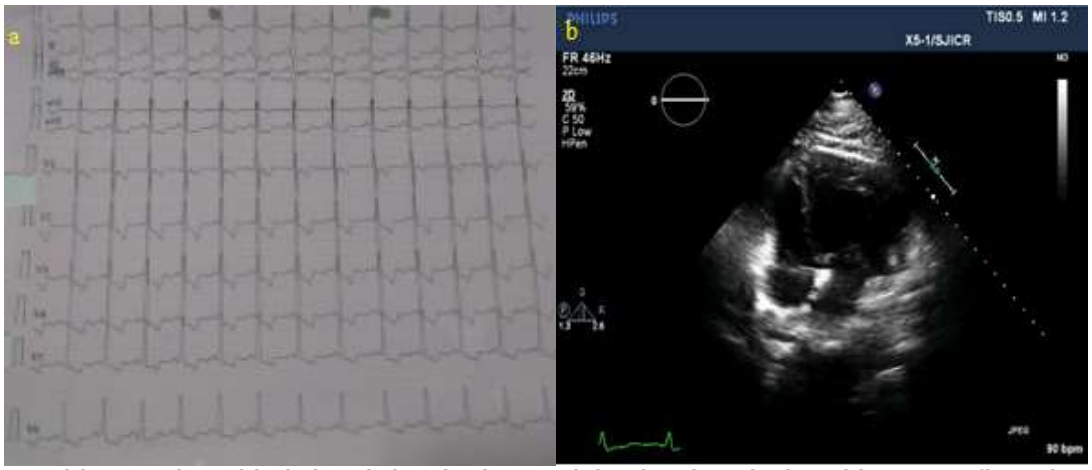

Figure 1: (a) ECG -Taken with Limb and Chest lead reversal showing sinus rhythm with LBBB configuration and absent septal q waves in all leads. (b) Transthoracic 2D-echocardiogram with probe on right side of chest apical four-chambered view RV on left side LV on right side.

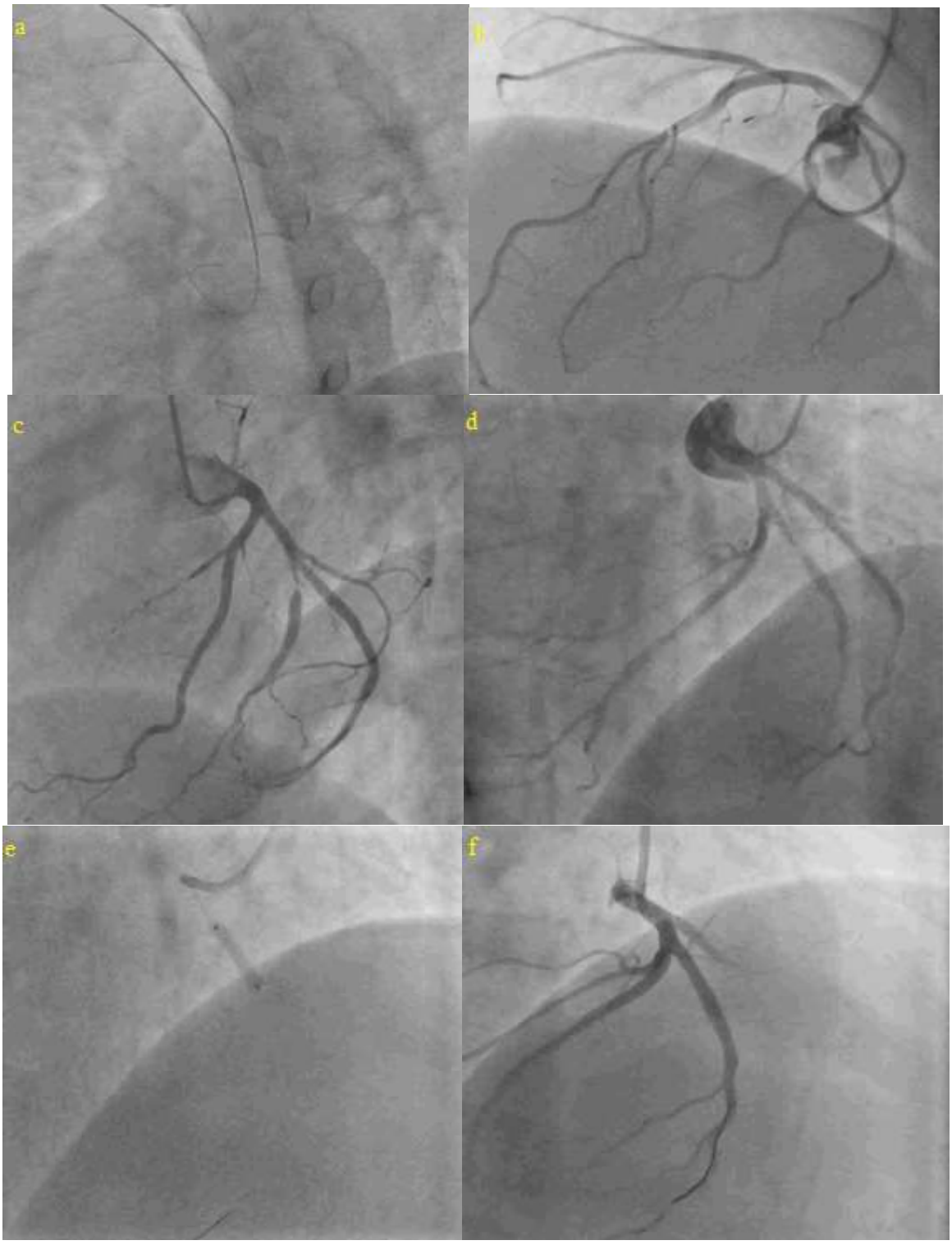

Figure 2: **Coronary angiogram and percutaneous intervention, (a) Right radial axis with tiger catheter over 0.35" Teflon** wire (b) LCA with left anterior descending and circumflex branches (c) RCA with branch showing tubular 95% disease in proximal branch. (d) Diagonally opposite view showing the tubular disease (e) Wired and stent deployed in the lesion after pre-dilatation. (f) Final result with fully expanded stent.

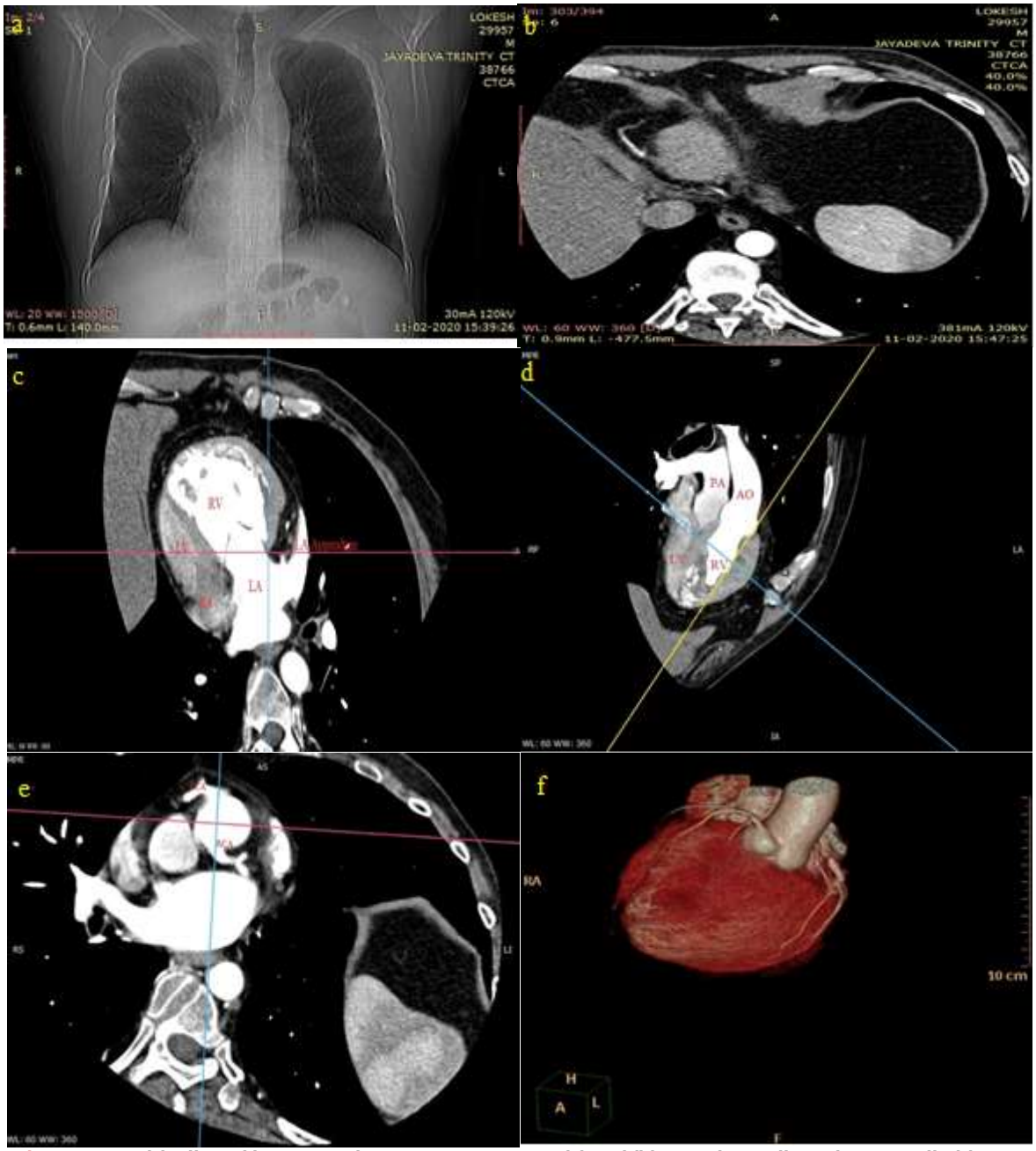

Figure 3: CT with aligned images to demonstrate anotomy (a) and (b) CT – Situs solitus, dextrocardia (c) Four chambered cut showing atrio-ventricular connections.(d) Out flow cut showing great artery relationship.(e) Origin of coronary arteries LCA from right anterior sinus and RCA from right posterior sinus. (f) 3D Reconstruction showing origin of coronary arteries and stent RCA branch.

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