Cor triatriatum and coronary artery fistula in tetralogy of Fallot

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ABSTRACT

Coexistence of divided left atrium with tetralogy of Fallot is rare. Preoperative diagnosis of this rare association is difficult. We here report preoperative diagnosis of this rare combination. In addition, the patient also had coronary to left ventricle fistula.

Keywords: Cor triatriatum, coronary artery fistula, partitioned left atrium, tetralogy of Fallot

CASE REPORT

A 10-year-old cyanotic boy was referred to our center for evaluation. Clinical examination revealed central cyanosis and Grade 3 clubbing of digits. On precordial examination there was a Grade 3/6 Crescendo-decrescendo murmur along the upper left sternal border. A short mid diastolic rumble was heard at the apex with the patient in the left lateral position. Two-dimensional echocardiography revealed a malaligned ventricular septal defect (VSD) with aortic override. It also showed presence of divided left atrium (suggestive of Cor triatriatum) [Figure 1, Video 1] and aneurysmally dilated left and right coronary arteries giving horizontal figure of eight appearance [Figure 2]. There was no evidence of aortic regurgitation (AR) or hypertension. Cine angiogram showed malaligned type of ventricular septal defect with aortic override and a well-developed left ventricle (LV) [Video 2]. An aortic root angiogram revealed dilated coronary arteries and no evidence of AR. Selective coronary angiogram showed aneurysmally dilated left coronary artery with contrast entering the left ventricle almost immediately [Video 3]. This confirmed the presence of coronary artery fistulae draining from left coronary artery to left ventricle. Selective coronary angiogram of right coronary artery also showed a dilated right coronary artery.

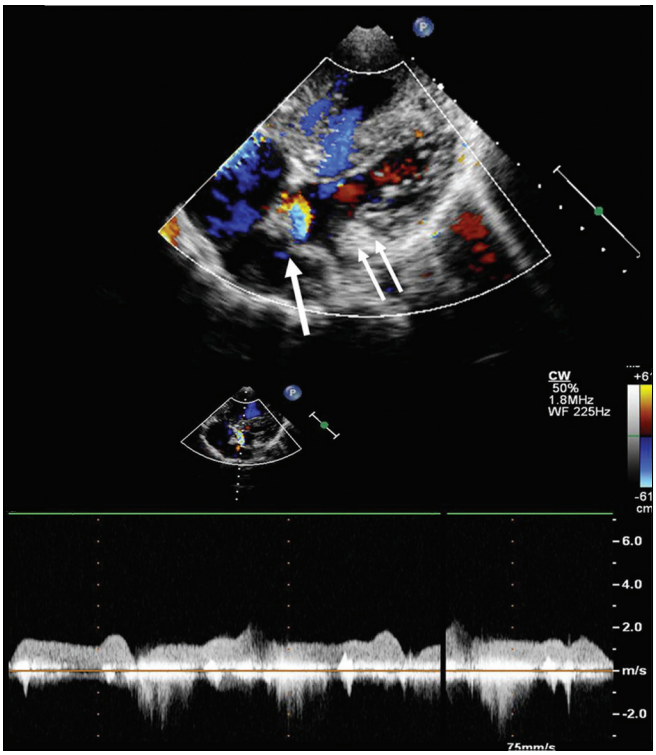

Figure 1: Apical four chamber view showing divided left atrium (arrow) and mitral valve leaflet (two arrows). Continuous wave Doppler across the membrane shows a diastolic gradient

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DISCUSSION

We here describe rare association of Cor triatriatum and coronary cameral fistulae with tetralogy of Fallot (TOF). All these defects were diagnosed preoperatively with help of echocardiography. Divided left atrium (LA) or Cor-triatriatum is a rare cardiac malformation in which LA is divided into two portions.[1] Its association with TOF is extremely rare with only one case diagnosed

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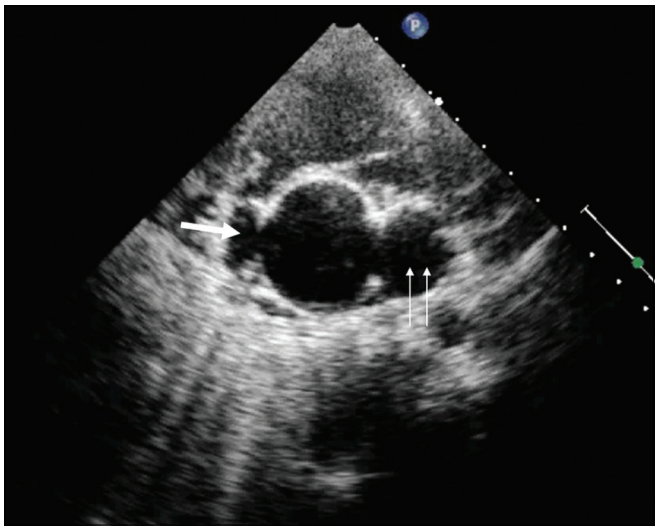

Figure 2: Parasternal short axis view at the level of aortic valve showing dilated left coronary artery (two arrows) and right coronary artery (single arrow)

preoperatively till date.[2] Coronary cameral fistulae in a case of TOF are also uncommon. Despite having inflow obstruction of left ventricle and presence of TOF his LV was relatively well developed. Identification of divided LA in such a case preoperatively is of immense important because successful repair of these defects can be carried out during repair. Failure to do so can result in death from pulmonary edema.[2] This is because surgical repair of tetralogy results in worsening of the previously unsuspected pulmonary venous obstruction associated with divided left atrium, since pulmonary blood flow is now unobstructed.[2] Congenital communications between coronary arteries and cardiac chambers are rare.[3] The receiving chamber is most commonly on the right side of the heart and fistulous drainage into the left side of the heart is extremely rare.[4] In the present case, coronary fistula was draining into the left ventricle. These fistulae represent persistent junctions of primordial epicardial vessels with intramyocardial sinusoidal circulation.[3] Rao *et al.* have described similar drainage of coronary arteries into the left ventricle through fistulous communications in absence of coronary sinus.[5] However, coronary sinus was present in this case. We call attention to this rare combination of anomalies and it's clinical importance.

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