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Abnormal single coronary artery from pulmonary artery (ASCAPA) associated with ventricular septal defect and pulmonary valve stenosis in young infant presented with heart failure: Case report

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ABSTRACT

Abnormal single coronary artery from pulmonary artery (ASCAPA) is a very rare anomaly; it is rarer than anomalous left coronary artery from pulmonary artery (ALCAPA). The incidence of ASCAPA is unknown because of little number of reported cases; it may be isolated or associated with other congenital heart diseases and is usually associated with significant myocardial ischemia and dysfunction timed with the physiological decrease of pulmonary vascular resistance (PVR) after birth. We are reporting a case of ASCAPA associated with perimembranous ventricular septal (VSD) defect and valvular pulmonary stenosis in an infant that presented with heart failure.

1. Case description

This case was 28 days old, full-term male baby, a product of a normal vaginal delivery. The patient was asymptomatic till the age of 2 weeks of life when his parents observed that their baby had shortness of breath and sweating that increased with feeding. There was no history of fever. Parents sought medical advice and were referred to the pediatric cardiology clinic. On examination the body weight was 3 kg (birth weight was also 3 kg), oxygen saturation was 97% on room air, respiratory rate was 65/min with intercostal and subcostal recessions, heart rate was 160/min, and there was also hepatomegaly (2 cm below costal margin). Cardiac auscultation revealed grade II ejection systolic murmur at the pulmonary area. 12 leads ECG revealed sinus tachycardia, northwest axis deviation, QS pattern in lead III, aVF, small q waves in a VR, V1, V3. There was no ST segment changes (Fig. 1). Chest X-ray revealed cardiomegaly with pulmonary venous congestion. The echocardiography showed: significant left and right ventricular dysfunction with echogenic papillary muscles, moderate size perimembranous ventricular septal defect (VSD) measuring 5 × 5 mm with mainly left to right shunting and with no significant pressure gradient (PG) across this VSD, thickening & doming of pulmonary valve with measured PG across the pulmonary valve 33 mm Hg. The presence of myocardial dysfunction with echogenic papillary muscles raised the suspicion of ALCAPA (Fig. 2A), so a detailed evaluation of coronary arteries was done using modified parasternal short axis that revealed antegrade flow in left coronary artery (LCA). Moreover, LCA seemed to arise from aorta and this was not consistent with ALCAPA (Fig. 2B), while high parasternal long axis view showed that the LCA seemed to arise from pulmonary artery which is consistent with ALCAPA but with antegrade flow (Fig. 2C). The right coronary artery couldn't be seen. Patient was then transferred to pediatric intensive care unit (PICU) for further evaluation and management. Because the echocardiography did not prove or exclude abnormal coronary anatomy multidetector computed tomography (MDCT) was done. MDCT showed an abnormal coronary artery from the pulmonary artery and from the sinus facing aorta (Fig. 2F). The CT couldn't determine the right coronary artery so we arranged for the diagnostic cardiac catheterization. In catheterization laboratory, the aortic angiography showed no coronary arteries arising from aorta (Fig. 2D) while pulmonary artery angiography showed doming of pulmonary valve, post stenotic dilatation of main pulmonary artery with abnormal single coronary artery from pulmonary artery (Fig. 2E). Hemodynamics showed markedly elevated left ventricular and right ventricular end diastolic pressures (26, 27 mm Hg respectively), and mild elevation of pulmonary artery pressure 46/31

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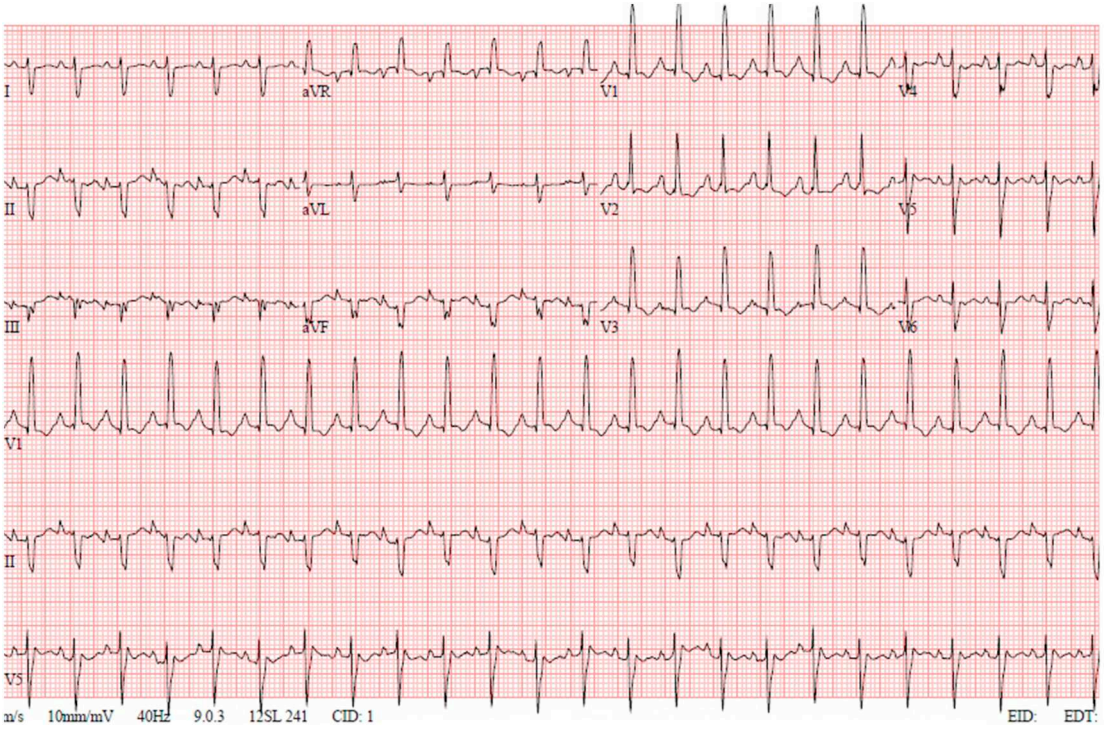

Fig. 1. ECG of the patient showing north west axis deviation, QS pattern in lead III, aVF, small q waves in a VR, V1, V3, there is no ST segment changes.

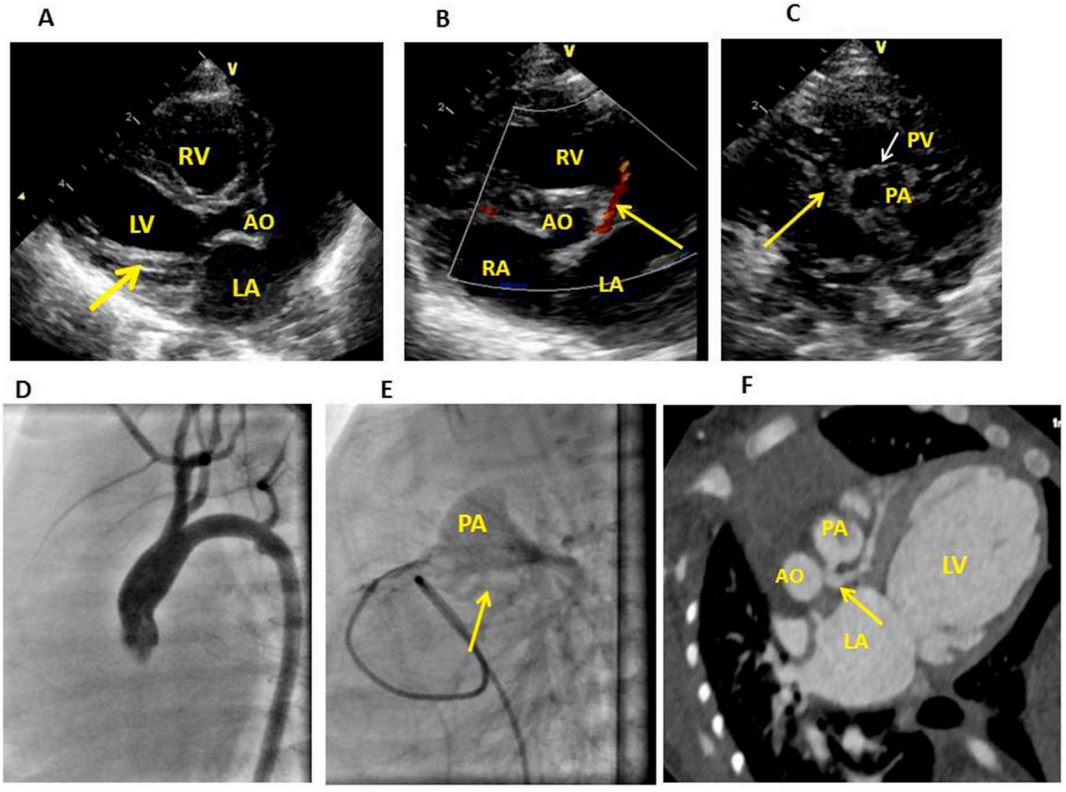

Fig. 2. A: Parasternal long axis view showing echogenic papillary muscles indicating myocardial ischemia (yellow arrow). B: Parasternal short axis view showing antegrade flow (red color) in the coronary artery (yellow arrow), in this view the coronary artery seems that it has normal origin from aorta this mainly is due to the poor lateral resolution of ultrasound, moreover the coronary artery arises from the pulmonary sinus facing the aorta then course rightward towards the left wall of ascending aorta as shown in (Fig. 2F). C: Parasternal long axis view with probe tilted upwards to show right ventricular outlet and pulmonary artery, in this view the pulmonary valve is seen thickened and domed (white arrow), and the coronary artery (yellow arrow) seems to originate from pulmonary artery. D: Aortic angiography with no coronary arteries are seen from the aortic sinuses. E: Pulmonary angiography showing post stenotic dilatation of pulmonary artery and the single coronary artery from the pulmonary sinus (yellow arrow). F: MDCT showing the origin of coronary artery from pulmonary artery then courses rightwards towards the aorta (yellow arrow). LV: left ventricle, LA: left atrium, Ao: aorta, RV right ventricle, PV pulmonary valve, PA; pulmonary artery, MDCT: multidetector computed tomography. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

(38) mm Hg. RV systolic pressure was 81 mm Hg; PG across pulmonary valve was 35 mm Hg. After clear delineation of coronary anatomy, patient transferred to operating room and after induction of anesthesia the patient developed cardiac arrest. Cardiopulmonary resuscitation was done for 8 min prior to cardiopulmonary bypass. After exploration, the myocardium looked dusky and ischemic with extensive endocardial scarring, re implantation of the single coronary artery, closure of VSD and pulmonary valvotomy were then done and the patient was discharged from operating room on extracorporeal membrane oxygenator (ECMO) and high inotropic support. Post-operative echocardiography revealed very poor contractility that didn't show any improvement during the successive days postoperatively. Full ECMO support continued for 5 days and couldn't be weaned and the patient developed ischemic color changes in his limbs. Echocardiography was done on the 5th day of full ECMO support revealed near absence of cardiac contractility. Then decision was taken to stop the ECMO support after talking with family then patient died.

2. Discussion

Abnormal single coronary artery from pulmonary artery (ASCAPA) is a very rare cardiac anomaly; it is rarer than anomalous left coronary artery from pulmonary artery (ALCAPA). The incidence of ASCAPA is unknown because of little number of reported cases; it may be isolated or associated with other congenital heart diseases [1–3]. ASCAPA usually associated with significant myocardial ischemia and dysfunction timed with the physiological decrease of pulmonary vascular resistance (PVR) after birth. In cases of ALCAPA the left coronary blood flow is dependent on the PVR and pulmonary artery pressure, so immediately after birth when PVR and pulmonary artery pressure are high, there is no decrease in coronary blood flow and no myocardial ischemia will happen. Later on, with physiological decrease of PVR, the blood flow in left coronary artery (LCA) will be retrograde from right coronary artery (RCA) to collaterals to LCA to pulmonary artery. At this time ischemia will happen and the echocardiography can show retrograde flow in LCA to the pulmonary artery, dilated right coronary artery, prominent septal perforators [4]. In cases of ASCAPA like our current case, there is only a single coronary artery that arises from pulmonary artery and no other coronaries arise from aorta and not associated with any collaterals from aorta supplying this single coronary artery resulting in profound ischemia and absence of retrograde flow in pulmonary artery that make echocardiographic diagnosis of such cases very difficult.

Normally the coronary perfusion depends on 1- The coronary perfusion pressure which is the difference between the aortic diastolic pressure (pulmonary diastolic pressure in our case) and the ventricular end-diastolic pressure (EDP), the more the pressure difference the more the coronary perfusion, the less pressure difference the less coronary perfusion. 2- The heart rate: as the coronary perfusion occurs only during diastole, higher heart rates are associated with shortening of diastolic time and hence decrease in coronary perfusion and this may explain the ventricular dysfunction associated with chronic tachyarrhythmia. In our case, the coronary perfusion pressure which is the difference between pulmonary diastolic pressure and ventricular EDPs was extremely low (coronary perfusion pressure was 5 mm Hg and 4 mm Hg for left and right ventricles respectively). More over the tachycardia associated with heart failure may explain the profound LV and RV dysfunction. Treatment of these cases is surgical either through reimplantation of coronary artery to aorta or through Takeuchi procedure (intrapulmonary baffle). In literature, mortality after surgical repair was higher (about 50%) especially in patient with severe LV dysfunction after surgery [1].

Recommendations

We learned from this case that any neonate with heart failure and myocardial dysfunction, evaluation of coronary anatomy is of paramount importance. MDCT angiography and/or cardiac catheterization should be done for these cases for better delineation of coronary anatomy.

Conflict of interest

The authors declare that there is no conflict of interest.

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Informed consent

Informed consent was obtained from the patient's parents.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ppedcard.2019.04.002.

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