Pulmonary atresia and ventricular septal defect with MAPCAs associated with right sided endocarditis and paradoxical embolic event

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Abstract  Pulmonary atresia and ventricular septal defect (PA-VSD) with major aortopulmonary collaterals (MAPCAs) is a complex and extremely heterogeneous anomaly. PA-VSD with both pulmonary arteries originating from systemic arterial circulation without MAPCAs and patent ductus arteriosus (PDA) is a very rare disease and according to our knowledge a case without cyanosis and symptoms of congestive heart failure after the first decade of life has not been reported. The majority of untreated patients die in their first decade of life as a result of intractable congestive heart failure or respiratory distress. This report informs about a 21-year-old PA-VSD patient who presented without cyanosis with both pulmonary arteries arising from aorta associated with right sided endocarditis and a paradoxical embolic event. © 2006 The European Society of Cardiology. Published by Elsevier Ltd. All rights reserved.

Introduction

Pulmonary atresia and ventricular septal defect (PA-VSD) with major aortopulmonary collaterals (MAPCAs) is a complex and extremely heterogeneous anomaly. A case of PA-VSD with both pulmonary arteries originating from systemic arterial circulation along with paradoxical embolia and tricuspid valve endocarditis has not been reported. This case seems to be the first to comprise all of these pathologies.

Case report

A 21-year-old boy was admitted with symptoms of fever, weakness and palpitation. He had symptoms of weakness and dyspnea previously, it was his first admission to a cardiology clinic. His physical examination revealed a remarkable systolic murmur in precordium and a continuous murmur in back. There were no clubbing and no cyanosis. Laboratory examination demonstrated leucocytosis, increased acute phase reactants, positive blood cultures of gram (+), oxidase (+)/C0 catalase (+) coccobacillus.

Chest X-ray showed increased right bronchovascularity, normal left bronchovascularity, right arcus aorta and increased CTI. Transthoracic 2D and Doppler echocardiography revealed wide VSD, 70% dextraposition of aorta, 0.8 × 1.3 cm sized, mobile vegetation on anterior cuspis of tricuspid valve, atresia of pulmonary valve and main pulmonary truncus. Suprasternal echocardiography revealed a large artery arising from left brachiocephalic truncus. Fig. 1A, B, and C shows suprasternal echocardiographic view of LSA-RPA (left subclavian artery-right pulmonary artery) collateral.

Pulmonary MRI angiography was performed to demonstrate the pulmonary anatomy. It revealed
right arcus aorta, right descending aorta, pulmonary atresia, left brachiocephalic truncus, Right Pulmonary Artery (RPA) arising from left brachiocephalic truncus, and Left Pulmonary Artery (LPA) arising from descending aorta. The pulmonary arteries were arising from aorta as well developed large branches. There were no extra collaterals. LPA and RPA were supplying all pulmonary segments. Pulmonary MRI angiographic images are shown in Fig. 2A and B.

Right and left heart catheterization showed pulmonary atresia and the anomalous origin of pulmonary arteries. Fig. 2C and D is the catheterization image. There was a stenosis in distal portion of left pulmonary artery and the systolic pressure of LPA (55 mmHg) was lower than the systolic pressure of RPA (100 mmHg). Systemic arterial and pulmonary arterial blood O₂ saturation was 96%. Fig. 2E demonstrates the whole abnormal anatomy of pulmonary arteries.

After evaluation of symptoms, echocardiography and blood culture analysis, patient was diagnosed as infective endocarditis. Vancomycin and gentamycin combination therapy was applied for 6 weeks. The clinical symptoms of endocarditis and echocardiographic sign of vegetation diminished during the therapy. On the 24th day of therapy, the patient underwent femoral embolectomy due to the right femoral embolia. Pathological examination of embolectomy material was consistent with healed vegetation. Macroscopy of the vegetation is shown in Fig. 3.

After the medical therapy of infective endocarditis, the patient recovered completely. Surgery for the complex congenital anomaly to reduce the pulmonary blood flow was planned. However, the patient and his family refused the operation due to its high risk. The patient is now followed up medically.

Discussion

PA-VSD and MAPCAs is a complex and rare lesion in which considerable morphologic variability exists regarding the sources of pulmonary blood flow leading to a wide spectrum of clinic presentation and difficulty in surgical therapy.⁴ PA-VSD represents as the most severe form of tetralogy of Fallot.

The Baltimore Washington Infant study reported an incidence of 0.07 per 1000 live births for PA-VSD. It accounts for 1.5% of all forms of congenital heart disease and 20% of all forms of TOF.

The source of pulmonary blood flow in PA-VSD is the systemic arterial circulation. PA-VSD is classified into three types according to the source of pulmonary blood flow.⁵ Surgical options of each type are based on the presence or absence of NPA (Native Pulmonary Arteries) and MAPCAs. In the most frequent type of PA-VSD, type A,
pulmonary circulation is through the PDA into the confluent pulmonary arteries supplying all of the bronchopulmonary segments. PA-VSD and MAPCAs comprises only about 25% of PA-VSD. In type B, NPA (native confluent pulmonary arteries) are present with MAPCAs. In type C, there are only MAPCAs, without NPA.

The nonconfluent pulmonary arteries exist in 20–30% of the PA-VSD patients. MAPCAs are large and distinct arteries, highly variable in number that usually arise from the descending aorta, but uncommonly may originate from the aortic arch, the subclavian, the carotid or even the coronary arteries. There is at least one MAPCA in the whole cases with nonconfluent pulmonary arteries.

Aotsuka et al. reported a case of anomalous origin of confluent pulmonary arteries arising from ascending aorta without an intracardiac defect. In previously defined PA-VSD congenital anomalies, nonconfluent well developed native pulmonary arteries without MAPCAs or PDA were not reported. In our case, the pulmonary arteries arise from the subclavian artery and the descending aorta like MAPCAs but they are well developed and branch to supply all of the pulmonary segments.

A complex congenital anomaly with right sided endocarditis that is complicated with peripheral embolia has not been reported previously. Infective endocarditis (IE) occurs less commonly in children than in adults (1/1280 per year). Because of the increased survival rate of children with congenital heart disease (CHD) and the overall decrease in rheumatic valvular heart disease in developed countries, CHD now constitutes the predominant underlying condition for IE in children. CHD with left to right shunt is a risk factor.

Figure 2  (A) Pulmonary MRI image demonstrating the large branch arising from the junction of left brachiocephalic truncus and the subclavian artery that forms the right pulmonary artery. (B) Pulmonary MRI image shows the large branch arising from the descending aorta that forms the left pulmonary artery. (C) Catheterization image. The right pulmonary artery arises from the left subclavian artery as a large branch. (D) The left pulmonary artery arises from descending aorta as a large branch. (E) Schematic explanation of anatomy of pulmonary arteries. LSA-RPA colat; left subclavian artery-right pulmonary artery collateral; Desc.Ao-LPA colat; descending aorta-left pulmonary artery collateral. LScA, left subclavian artery; RPA, right pulmonary artery; Desc.Ao, DA: descending aorta; LPA, left pulmonary artery; and obs; obstruction-stenosis in the distal portion of the branch.
for right sided endocarditis. The management of infective endocarditis is similar to those in adult endocarditis. Antimicrobial prophylaxis is particularly important in high risk groups.

The option of surgery of PA-VSD and MAPCAs without central pulmonary artery is the unifocalization surgery. In our opinion, the stenosis of the distal portion of LPA has protected the left pulmonary segments from advanced pulmonary disease. But due to the large systemic pulmonary arteries, pulmonary vascular disease development is inevitable in following years. A stepwise surgical approach to reduce the pulmonary blood flow was planned for our case.

References