Case Report

Pentalogy of Fallot with a Single Coronary Artery: A Rare Case Report

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Abstract

Anomalies of origin, course, and distribution of coronary arteries, including single coronary artery, are well known in patients with Tetralogy of Fallot. However, to the best of our knowledge, there is no published case report of Pentalogy of Fallot with a single coronary artery. Herein, we introduce a 22-year-old female patient diagnosed via echocardiography and cardiac catheterization preoperatively as Pentalogy of Fallot with a single coronary artery arising from the left coronary sinus.

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Introduction

Pentalogy of Fallot is a rare form of cyanotic congenital heart disease, characterized by an association of atrial septal defect (ASD) with tetralogy of Fallot (TOF).[1] Coronary artery anomalies are prevalent in patients with TOF[1, 2] and must be recognized preoperatively to plan an appropriate surgical approach.[3-5] We describe a rare case report of pentalogy of Fallot associated with a single coronary artery.

Case Report

A 22-year-old female patient presented to us with a history of cyanosis since 6 months of age as well as exertional breathlessness and palpitations since 5 years of age. On general examination, she had a thin build, central cyanosis, and grade IV clubbing. Her heart rate was 80 beats per minute, blood pressure was 120/70 mmHg, and resting arterial oxygen saturation was 72%. The mean jugular venous pressure (JVP) was not raised, but waveforms showed large v-waves and a steep y-descent. Precordial examination revealed a prominent right ventricular (RV) impulse, single loud second heart sound, and a grade 3/6 ejection systolic murmur at the upper left sternal edge. Electrocardiography showed normal sinus rhythm, right axis deviation, right atrial abnormality, and RV hypertrophy. Chest roentgenogram (Figure 1) revealed an RV type of apex, inconspicuous pulmonary bay, left-sided aortic arch, and oligemic lung fields. Echocardiography (Figures 2 and Figure 3) demonstrated large sub-aortic perimembranous ventricular septal defect (VSD) with 50% aortic override, large ostium-secundum atrial septal defect (ASD), thickened septal leaflet of the tricuspid valve with severe tricuspid

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regurgitation, dilated and hypertrophied RV, dilated right atrium (RA), and thick-dysplastic pulmonary valve. The pulmonary artery could not be visualized, and RV systolic pressure was 120 mmHg + right atrial pressure. Furthermore, the coronary arteries could not be visualized due to limited echocardiographic window.

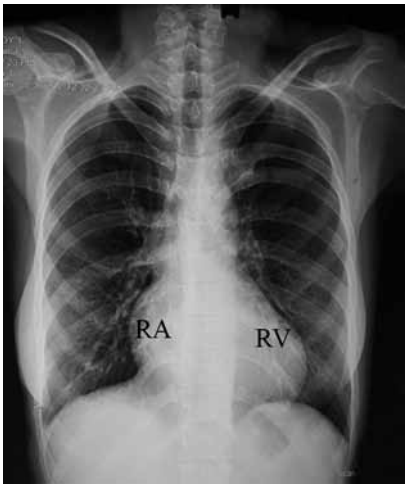

Figure 1. Chest X-ray (posteroanterior view), revealing oligemic lung fields and right ventricular type cardiomegaly
RA, Right atrium; RV, Right ventricle

The patient was taken up for cardiac catheterization. RV angiography was performed using a 6-French pigtail catheter coursing antegradely from the RA, through the tricuspid valve, into the RV. Also, left ventricular (LV) angiography was done antegradely with the catheter passing from the RA, through the ASD and mitral valve, and into the LV, as well as retrogradely with the catheter passing from the aorta, through the aortic valve, and into LV. The angiograms confirmed the echocardiographic findings of sub-aortic VSD, aortic override, ASD, and severe infundibular and valvular pulmonary stenosis. The pulmonary arteries were visualized and well developed. Aortic root injection revealed an absent right coronary sinus and a single coronary artery arising from the left coronary sinus, which was selectively catheterized using a 6-French Judkin's left catheter (size 4). Coronary angiograms (Figure 4) in standard and oblique views with caudal and cranial tilts confirmed the presence of a single coronary artery arising from the left coronary sinus giving rise to the right coronary artery (RCA) from its proximal segment and subsequently dividing into the left anterior descending (LAD) and left circumflex coronary arteries. The RCA followed a posterior course behind the aorta and pulmonary artery reaching the right atrioventricular groove and supplied the RV myocardium and the right atrium. The LAD and left circumflex coronary arteries followed their normal course and distribution.

Considering all these findings, a diagnosis of Pentalogy of Fallot with a single coronary artery was made, and the patient was scheduled for cardiac surgery for intracardiac repair.

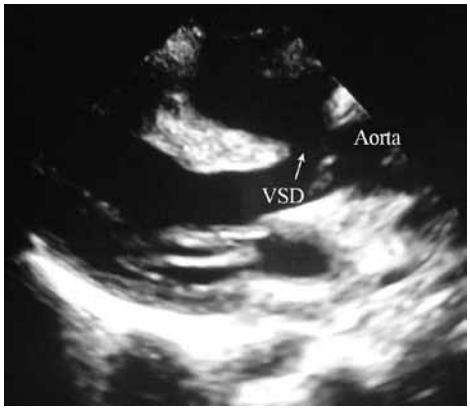

Figure 2. Echocardiographic parasternal long-axis view, revealing aortic override with a large ventricular septal defect (VSD)

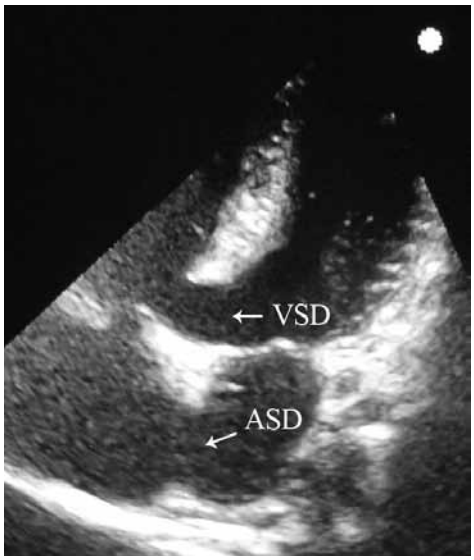

Figure 3. Echocardiographic apical four-chamber view, revealing atrial septal defect (ASD) and ventricular septal defect (VSD)

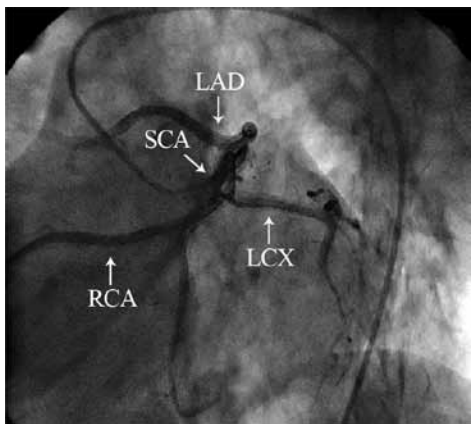

Figure 4. Coronary angiogram, revealing a single coronary artery (SCA) giving rise to the right coronary artery (RCA), left circumflex coronary artery (LCX), and left anterior descending coronary artery (LAD)

Discussion

Pentalogy of Fallot is a term used to describe an association of Tetralogy of Fallot (TOF) with an atrial septal defect.[1] TOF is the most common form of cyanotic congenital heart disease and accounts for 5% to 10% of all congenital heart diseases (CHDs), with an incidence of 3 in 10000 live births.[6] It is characterized by antero-cephalad deviation of the infundibular septum, resulting in a malalignment type of nonrestrictive perimembranous VSD, aorta overriding the VSD, RV hypertrophy, and RV outflow obstruction. An additional ASD is seen in 3% to 5% of cases, with ostium-secundum type being the most common, followed by ostium primum type and rarely sinus venosus type.[1] Coronary artery anomalies are well known in patients with TOF, with a prevalence of 5% to 12% in various angiographic, surgical, and autopsy series.[2] The most common variant is the LAD arising from the RCA, followed by single coronary artery arising from the left or right coronary sinus, RCA arising from the LAD or left circumflex coronary artery, and a large conus branch of the RCA.[7] A single coronary artery is a very rare coronary anomaly in which the entire coronary system originates from a single coronary ostium. Its incidence is 0.0240% to 0.066% in the general population undergoing coronary angiography,[8] but it is encountered more frequently with other congenital heart defects such as TOF, persistent truncus arteriosus, transposition of the great arteries, and pulmonary atresia.[9] It has been reported in 1.5% to 3.7% of patients with TOF.[1, 2] The reason for the high association of these anomalies is not completely known; however, dextroposition of the aorta or the failure of one of the coronary buds to develop has been postulated as the underlying mechanism. These anomalies of epicardial distribution are functionally insignificant and do not cause myocardial ischemia, but their main importance is related to surgical repair. Of greatest concern are those anomalies in which the aberrant artery courses anteriorly across the RV outflow tract, as inadvertent transsection of such a vessel while performing right ventriculotomy for TOF repair adds considerably to the surgical mortality and morbidity.[3] Hence, preoperative recognition of these anomalies is crucial in deciding the timing and technique of surgery. Alternative surgical techniques in such patients include the transatrial-transpulmonary approach,[4] modified right ventriculotomy,[3] and an extracardiac RV to pulmonary artery conduit.[5] Echocardiography has been used to evaluate the coronary anatomy in patients with TOF, but with limited sensitivity and specificity.[10] Although multidetector coronary computed tomography (CT) and magnetic resonance (MR) angiography are rapidly emerging as alternatives to catheter angiography for evaluating the coronary anatomy in patients with CHD,[11] many patients with TOF still undergo cardiac catheterization and angiography for such evaluation. The evaluation our patient led us to the diagnosis of Pentalogy of Fallot with a single coronary artery. We extensively reviewed the literature to look for any such reported association, but could not find one. Therefore, we report this extremely rare combination of congenital cardiac anomalies.

Conclusion

In this article, we described a very rare case of an adult patient with Pentalogy of Fallot and a single coronary artery arising from the left coronary sinus, which has not been published so far. Preoperative recognition of such an anomaly is important for selecting an appropriate technique for surgical repair.

References

1. Dabizzi RP, Teodori G, Barletta GA, Caprioli G, Baldrighi G, Baldrighi V. Associated coronary and cardiac anomalies in the tetralogy of Fallot. An angiographic study. Eur Heart J 1990;11:692-704.
2. Fellows KE, Freed MD, Keane JF, Praagh R, Bernhard WF, Castaneda AC. Results of routine preoperative coronary angiography in tetralogy of Fallot. Circulation 1975;51:561-566
3. Bonchek LI. A method of outflow tract reconstruction in tetralogy of Fallot with anomalous anterior descending coronary artery. Ann Thorac Surg 1976;21:451-453.
4. Hudspeth AS, Cordell AR, Johnston FR. Transatrial approach to total correction of tetralogy of Fallot. Circulation 1963;27:796-800.
5. Hurwitz RA, Smith W, King H, Girod DA, Caldwell RL. Tetralogy of Fallot with abnormal coronary artery: 1967 to 1977. J Thorac Cardiovasc Surg 1980;80:129-134.
6. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002;39:1890-1900.
7. Tchervenkov CI, Pelletier MP, Shum-Tim D, Béland MJ, Rohlicek C. Primary repair minimizing the use of conduits in neonates and infants with tetralogy or double-outlet right ventricle and anomalous coronary arteries. J Thorac Cardiovasc Surg 2000;119:314-323.
8. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. Radiology 1979;130:39-47.
9. Antonellis J, Rabaouni A, Kostopoulos K, Margaris N, Kranidis A, Salahas A, Ifantis G, Koroxenidis G. Single coronary artery from the right sinus of Valsalva, associated with absence of left anterior descending and an ostium-secundum-type atrial septal defect: a rare combination. A case report. Angiology 1996;47:621-625.
10. Jureidini SB, Appleton RS, Nouri S. Detection of coronary artery abnormalities in tetralogy of Fallot by two-dimensional echocardiography. J Am Coll Cardiol 1989;14:960-967.
11. Gilkeson RC, Ciancibello L, Zahka K. Pictorial essay. Multidetector CT evaluation of congenital heart disease in pediatric and adult patients. AJR Am J Roentgenol 2003;180:973-980.