Cardiology in the Young

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Single right coronary artery with right ventricular fistula and congenital absence of left coronary artery: an extremely rare combination

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Brief Report

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Abstract

We report an extremely rare case of a 14-month-old girl who was diagnosed with a single right coronary artery with coronary artery fistula communicating with the right ventricle and congenital absence of left coronary artery. Angiography showed a dilated and tortuous single right coronary artery draining into the right ventricle, absence of left coronary system, and left ventricular coronary circulation supplied via collateral vessels.

Introduction

A single coronary artery is a rare congenital anomaly of the coronary arteries, which is described as one coronary artery arising from the aortic trunk by a single coronary ostium and providing for perfusion of the entire myocardium. It is responsible for about 0.04% of congenital cardiac anomalies.[1] Coronary artery fistula is a rare congenital anomalous connection between the coronary arteries and a cardiac chamber or great vessel. The incidence of coronary artery fistula in the paediatric population is not exactly known. For the adult population, angiographic investigations show an overall incidence of 0.13–0.6%.[2] Single anatomic right coronary artery with the absent equivalents of the left coronary artery system is an extremely rare coronary artery anomaly with only a few adult cases reported hitherto.[3–5] The clinical significance of coronary anomalies depends on their potential to cause myocardial ischemia and sudden death, which in turn is dependent on their origin, course, and associated CHDs. Here we reported an extremely rare case who has a combination of single coronary artery, coronary artery fistula, and absence of equivalents of the left coronary artery system. According to our literature review, a combination of single coronary artery and coronary artery fistula is an exceedingly rare entity in paediatric population, and only a few cases have been reported.[1] However this case is the first paediatric case of single anatomic right coronary artery with coronary artery fistula and absent equivalents of the left coronary artery system.

Case report

A 14-month-old girl was referred to us for the evaluation of an incidentally detected cardiac murmur on routine clinical examination. General physical examination was unremarkable. Cardiovascular examination revealed a continuous murmur associated with thrill heard best over the right fourth intercostal space. There was no evidence of congestive cardiac failure. Routine electrocardiogram and chest X-ray were normal. The cardiac enzymes were normal. Two-dimensional echocardiography showed an aneurysmally dilated right coronary artery with the presence of coronary cameral fistula originating from right coronary artery and opening to right ventricular apex. The origin of the left main coronary artery could not be visualised. Left and right heart catheterisation was performed. On right heart catheterisation, the intra-cardiac and intravascular pressures were normal. Oxygen saturation showed a 3% step-up in the right ventricle, and the pulmonary/systemic flow ratio was 1.2:1. On left heart catheterisation ascending aortography showed a single aneurysmally dilated and tortuous right coronary artery arising from right coronary sinus and absence of left main coronary artery and equivalents of the left coronary artery system. Selective coronary angiography of right single coronary artery showed fistulous communication arising from single right coronary artery and draining into the right ventricle apex (Figure 1). Selective angiography of right single coronary artery showed multiple branches from superdominant posterior descending artery, posterolateral ventricular branch supplying the left coronary artery territory (Figure 2).

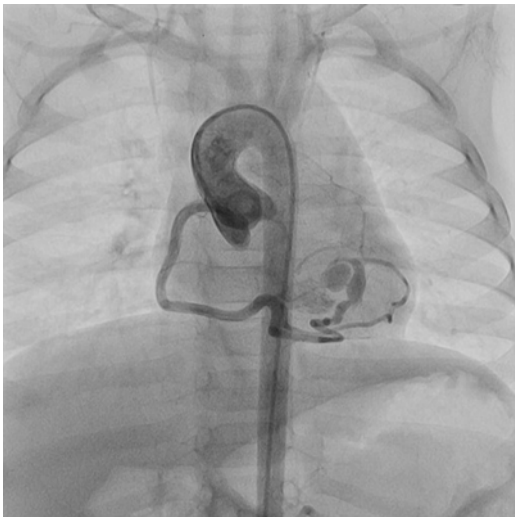

Figure 1. Aortic root injection (anteroposterior view). A dilated and tortuous vessel arises from right coronary sinus. The LMCA originating from the left coronary sinus and equivalents of the left coronary artery system is not detected. There is a fistula leading to the right ventricle apex.

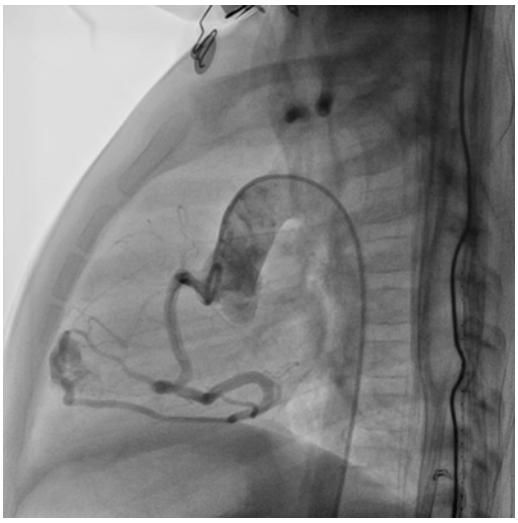

Figure 2. Aortic root injection (left lateral view). The LMCA originating from the left coronary sinus and equivalents of the left coronary artery system is not detected. Thin collateral vessels are supplying the left coronary artery territory.

Discussion

The coronary arterial circulation may rarely be supplied by a single coronary artery arising from the right, left, or posterior sinus of Valsalva. The single coronary artery combined with coronary artery fistula is a very rare entity, and there are a few children with this condition in the literature.[1] These cases showed that single left main coronary artery more common from single right coronary artery and coronary artery fistula draining into the right ventricle in the majority of patients.[1] There are two classifications of the single coronary artery (Smith and Lipton et al.) that are most commonly used.[6,7] But neither of them is insufficient to categorise cases like ours. Recently another new classification of the single coronary artery was made by Shirani and Roberts.[3] They classified the single coronary artery according to the site of origin, anatomical distribution of the branches, and the presence or absence of equivalent arteries of one of the absent main coronary arteries. They proposed a novel classification that included 20 possible types, encompassing all feasible anatomic combinations, and their manifestations.[3] According to this classification our case falls under class Type IIa which consists of a single right coronary artery with super-dominant branches supplying the left coronary artery territory in place of the left main, left anterior descending and left circumflex equivalents.

Coronary arterial fistulas are usually asymptomatic in the first two decades, especially when they are hemodynamically small.[2] The clinical presentation varies based on age, size, and anatomy of the coronary artery fistula.[1,2] The indications for the treatment of coronary arterial fistulas include the presence of a large or increasing left-to-right shunt, left ventricular volume overload, myocardial ischemia, left ventricular dysfunction, congestive cardiac failure and prevention of endocarditis/endarteritis.[8] If fistulas are detected in infancy and are asymptomatic, conservative management is appropriate.[8] However there are not enough data in the current literature about how to plan treatment for the single coronary artery with coronary artery fistula. Especially in our case the treatment plan is somewhat more complicated. Transcatheter or surgical closure of coronary artery fistula can cause impairment in coronary blood flow of left coronary artery territory. On the other hand, coronary artery fistula can cause a greater risk for coronary steal phenomena in left coronary artery territory. Our patient had no symptom, and the haemodynamic status of single right coronary artery showed no problem. So we encourage close follow-up, because the obstruction of coronary artery fistula could be fatal. Having a knowledge of this extremely rare anomaly will be helpful in the differential diagnosis of coronary artery abnormalities and assist the physician in treatment planning.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the Helsinki declaration of 1975, as revised in 2008, and has been approved by the Institutional Ethics Committee.

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