Journal of Cardiology Cases 6 (2012) e39–e41

Contents lists available at www.sciencedirect.com

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

# A rare combination: Single coronary artery originating from the left sinus of valsalva coexisting with perimembranous ventricular septal defect

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ARTICLE INFO

*Article history:*
Received 7 December 2011
Received in revised form 21 March 2012
Accepted 6 April 2012

*Keywords:*
Single coronary artery
Ventricular septal defect
Coronary angiography

SUMMARY

Single coronary artery (SCA) is a rare congenital anomaly in which the entire coronary system arises from a solitary ostium. Although an uncommon congenital anomaly, SCA is encountered more frequently in the presence of other congenital cardiac malformations such as persistent truncus arteriosus, tetralogy of Fallot, transposition of the great arteries, or pulmonary atresia.

In this report, we present a case of SCA arising from the left sinus of valsalva coexisting with perimembranous ventricular septal defect.



## Introduction

Single coronary artery (SCA) is a rare congenital anomaly in which the entire coronary system arises from a solitary ostium. As an isolated finding, the incidence ranges between 0.024% and 0.066% in the general population undergoing coronary angiography [1–4]. Although an uncommon congenital anomaly, SCA is encountered more frequently in the presence of other congenital cardiac malformations such as persistent truncus arteriosus, tetralogy of Fallot, transposition of the great arteries, or pulmonary atresia. In this report, we present a case of SCA arising from the left sinus of valsalva coexisting with perimembranous ventricular septal defect.

## Case report

A 61-year-old hypertensive male patient without a family history of cardiac disease, was admitted to our outpatient clinic with the complaints of dyspnea and palpitation during daily activities for one year. His blood pressure and heart rate were measured as 140/90 mmHg and 80 beats/min, respectively at physical examination. On auscultation, grade 3/6 early systolic murmur over the left lower sternal border and grade 2/4 distolic rumble at the apical region was heard. Chest examination revealed no pathologic findings. The electrocardiogram showed normal sinus rhythm, left atrial hypertrophy, and right axis deviation. Chest radiography demonstrated normal cardiac silhouette and marked pulmonary vasculature, particularly at basal levels. Blood chemistry was normal except for mild hypertriglyceridemia and impaired fasting glucose (277 mg/dl and 111 mg/dl, respectively). A perimembranous ventricular septal defect with a diameter of 13 mm and left to right shunt and moderate tricuspid regurgitation was detected on color flow Doppler imaging by transthoracic echocardiography (Fig. 1a and b). Calculated Qp/Qs ratio was 1.8 and estimated pulmonary arterial systolic pressure was about 55 mmHg. Due to the advanced age of the patient, coronary angiography was performed via right femoral artery by the Judkins technique, which revealed SCA arising from the left sinus of Valsalva and mild non-obstructive atherosclerotic involvement in the entire coronary tree (Figs. 2 and 3). Left ventriculography at cranial left anterior-oblique projection demonstrated a perimembranous ventricular septal defect (VSD) (Figs. 4 and 5). The patient underwent cardiac surgery for VSD repair.

## Discussion

Angiographic classification of single coronary arteries is as follows [1,2]: 'R' and 'L' denote the origin of the SCA from the right and left sinus of Valsalva, respectively. Then, the anatomical course of

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1878-5409/$ – see front matter 
http://dx.doi.org/10.1016/j.jccase.2012.04.004

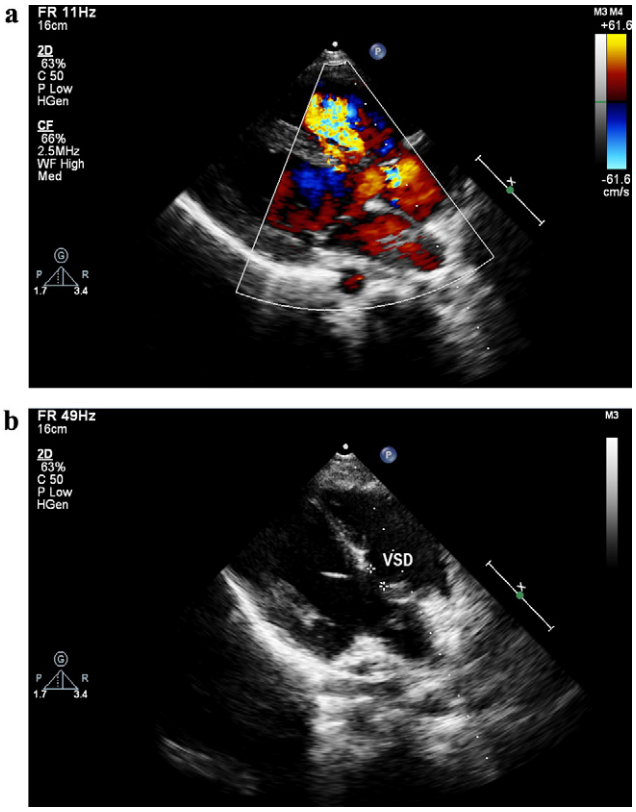


**Figure 1.** (a) and (b) A perimembranous ventricular septal defect (VSD) with left to right shunt and moderate tricuspid regurgitation on color flow Doppler imaging by transthoracic echocardiography.

the anomalous coronary artery is designated: Type I denotes an anatomical course of either a right or left coronary artery; Type II denotes one coronary artery arising from the proximal part of a normally located coronary artery; and Type III denotes the condition where the left anterior descending and left circumflex arteries arise separately from the proximal part of the normal right coronary artery; thus, this type can only be true for the 'R' type of SCA. Finally, the course of the anomalous artery in relation to the great vessels is designated with the letters 'A' (anterior), 'P' (posterior), 'B'

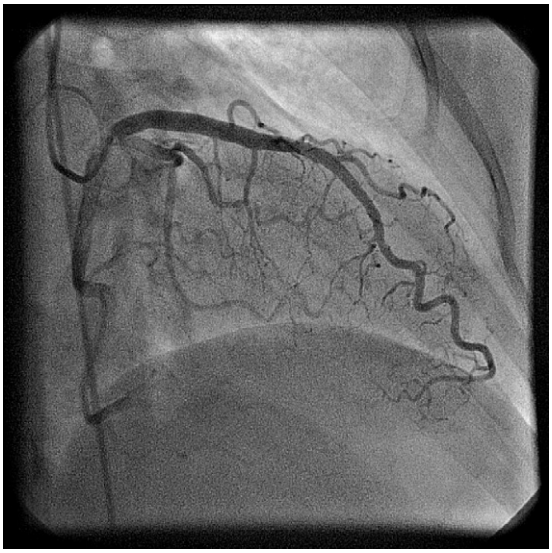

**Figure 2.** Left coronary angiogram in right anterior oblique view.

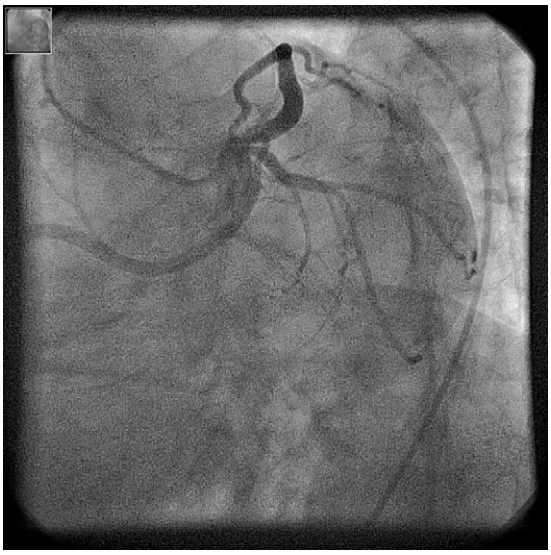

**Figure 3.** Left coronary angiogram in left anterior oblique caudal view.

(interarterial course between the aorta and the pulmonary artery), 'S' (part of the route is through the interventricular septum), and 'C' (a combination of diverse routes). In our case, the SCA was type L-IIA according to this classification.

Identification of the interarterial course of an arterial segment is important, since this type represents the highest risk for sudden cardiac death. Mechanical compression of the vessel between the aorta and the pulmonary artery or kinking are the potential causes of ischemia and sudden cardiac death [5,6]. Although the majority of patients are asymptomatic, patients with SCA can present with recurrent ischemia, acute myocardial infarction, heart failure, syncope, and nonfatal ventricular fibrillation [7]. In 15% of patients with ischemia, no obstructive atherosclerotic lesion is found [8]. Impaired coronary flow reserve, acute aortocoronary angulation, slit-like ostium, ostial tissue flaps, initial course of the coronary artery within the aortic wall, or spasm are the other mechanisms that cause ischemia in the absence of atherosclerosis. An anomalous SCA does not appear to be associated with an increased risk for the development of coronary atherosclerosis [9].

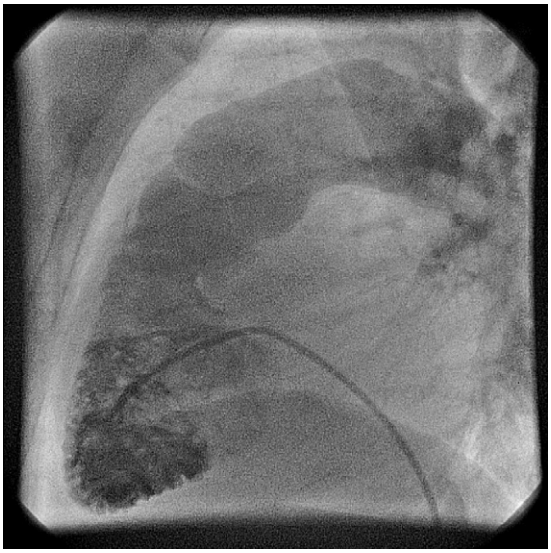

**Figure 4.** Left ventriculography at left anterior-oblique view. The image was obtained from the left ventricle by passing through the ventricular septal defect during right heart catheterization.

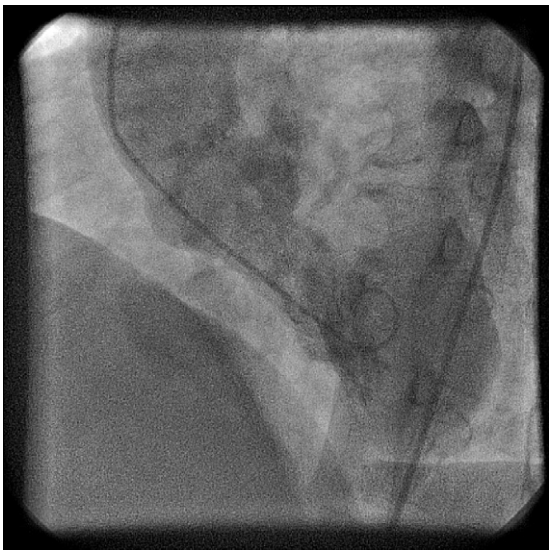

**Figure 5.** Left ventriculography at cranial angle. The image, obtained during left heart catheterization, which demonstrates right ventricular opacification and ventricular septal defect in the perimembranous region (under the aortic valve).

Association of SCA with membranous VSD in the adult population is very rare. We have found only 4 case reports in the literature to date, all of which were diagnosed by cardiac catheterization [10–13].

Embryologically, the development of the cardiac outflow tract and great vessels is a complex process that involves coordinated regulation of multiple progenitor cell populations. The absence of epicardium-derived cells leads to embryonic death, while delayed outgrowth may result in pathologies ranging from the absence of the main coronary arteries to pinpoint orifice formation. As coronary artery anomalies are more frequently expected to accompany congenital anomalies like persistent truncus arteriosus, tetralogy of Fallot, transposition of great arteries, and pulmonary atresia, our case represents a rare coexistence of VSD and a SCA.

## Disclosure

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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