Single Coronary Artery with Multiple Cardiac Anomalies

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Single coronary artery combined with other cardiac anomalies is uncommon. We present a 64-year-old man with unicuspid valvular and infundibular pulmonary stenosis, single coronary artery arising from right sinus of Valsalva, combined with coronary artery fistula and secundum atrial septum defect. This patient received operation of pulmonary valve replacement, infundibulectomy and ASD closure. He had complete resolution of symptoms after surgery.

Key words: Atrial septal defect, coronary artery fistula, Infundibular stenosis; Pulmonary stenosis; Single coronary artery

Congenital coronary artery anomaly occurs infrequently in general population. Single coronary artery is rare. Anomalous origin of the single coronary artery from right sinus of Valsalva is extremely rare, which is found in 0.6-1.2% of all coronary anomalies [1, 2]. Coronary artery fistula (CAF) communicating with a cardiac chamber, the coronary venous system or the pulmonary artery are also rare, a condition similar to valvular and infundibular pulmonary stenosis (PS) without ventricular septal defect (VSD) or tetralogy of Fallot.

We report a case with multiple cardiac anomalies including unicuspid valvular and infundibular PS, single coronary artery arising from the right sinus of Valsalva, coronary artery fistula and secundum atrial septal defect (ASD).

CASE REPORT

A 64-year-old male had previous medical history of old stroke with right hemiparesis for 10 years. Due to worsening dyspnea and chest tightness for 10 days, he visited our emergency department for help. Arterial desaturation with respiratory failure was noted in the emergency department. No orthopnea, paroxysmal nocturnal dyspnea, or low legs edema was noted except exercise intolerance before this episode. Physical examinations revealed a grade IV/VI systolic ejection murmur heard in the fourth intercostal space over the left sternal border. Chest radiography showed cardiomegaly with prominent left pulmonary artery. Transthoracic echocardiography demonstrated enlarged right atrium with paradoxical septal segmental wall motion and severe pulmonary hypertension (95mmHg). An inter-atrial septal defect with bilateral shunt was also demonstrated. Chest computed tomography showed ASD and pulmonary stenosis with marked post-stenotic dilatation of pulmonary artery (Figure 1). Right catheterization disclosed a peak systolic pressure gradient of 78
mmHg between right ventricle and main pulmonary artery. There was 8% oxygen step-up between mixed venous blood and the right atrium compatible with echocardiography finding of ASD. Right ventricular angiography revealed valvular and infundibular pulmonary stenosis (Figure 2). Coronary angiography showed anomalous origin of the coronary arteries all arose from the single ostium in right sinus of Valsalva and a CAF from left circumflex artery (LCX) (Figure 3).

Due to multiple cardiac anomalies with desaturation and refractory respiratory failure, he underwent unicuspid pulmonary valve replacement with infundibulectomy and ASD closure with prolene suture. This patient had complete resolution of the symptoms after surgery.

**DISCUSSION**

Overall, anomaly of the coronary artery is rather rare and the incidence of primary congenital

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**Figure 1.** a. Chest CT revealed the right ventricular outflow tract stenosis (white arrow) with marked post-stenotic dilatation of pulmonary artery. b. ASD (arrow head) and pulmonary stenosis (white arrow). c. Suspicious of the common ostium of three coronary arteries (black arrow). A: Aorta, LA: Left atrium, RA: Right atrium.

**Figure 2.** Right ventricular angiography revealed valvular (white arrow) and infundibular pulmonary stenosis (black arrow).
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coronary anomaly varies from 0.3% in necropsy series reported by Alexander and Griffith to 1.6% of patients undergoing cardiac catheterization in a series of more than 38,000 patients [3]. Although in some reports, these anomalies can cause ischemia or congestive heart failure, most anomalies are detected incidentally by cardiac catheterization and are asymptomatic.

Cardiac catheterization has traditionally been indicated to demonstrate the coronary vasculature in detail and has remained the reference standard imaging modality. Owing to the potentially complex three dimensional anatomy of the anomaly, conventional angiography, not infrequently, incompletely delineates the anatomical course of the coronary artery. Reliable, complete, and non-invasive assessment is therefore desirable and probably advantageous. It is difficult to determine the outflow drainage of CAF in our case due to combination of multiple anomalies. The anatomies of these anomalies were also challenging to delineate by means of cardiac catheterization. Other non-invasive tech-
niques such as nuclear medicine, computed tomography (CT) and cine-MR cardiac imaging may become increasingly important in this issue [10].

ECG gated multidetector row CT (MDCT) imaging and 3D MR imaging have increasingly been found to be a useful tool for evaluation of adult congenital heart diseases and coronary flow and function [8, 9]. Four slice CT coronary angiography was not sufficiently robust to produce consistent and reliable imaging for clinical use in terms of both reliability of image quality and accuracy of results due to motion artifact related to higher heart rate. If sixteen slice MDCT is not available, detailed and systemic reviewing to trace the whole course of coronary artery, or identification of associated anomalies may be an important clue in clinical radiologists’ interpretation.

Single coronary artery is very rare. It presents in 0.024% of general population. Hyrtl described the first case 160 years ago quoted by Neil et al [4]. The classification of single coronary artery by anatomic configuration was reported by Shirani et al. A single orifice in the left sinus of Valsalva is classified as type-I pattern, whereas type-II corresponds to a right-side solitary orifice. Type III is defined as the orifice in the non-coronary sinus of Valsalva. Our report case belonged to type-II according to the anatomic configuration. Single coronary artery from right sinus of Valsalva is extremely rare, found only in 0.6-1.2% of all coronary anomalies [1, 2].

Patients with single coronary artery present with various clinical and prognostic implications. Taylor et al reported an extremely high sudden death rate in patients with anomalous left coronary artery orifice from right sinus of Valsalva, especially in inter-arterial type [5]. Single coronary artery is usually combined with various cardiac anomalies such as truncus arteriosus, transposition of great vessels or tetralogy of Fallot. The present case had combined unicuspid pulmonary valvular stenosis, CAVF and ASD. This embryological time relation between the development of pulmonary valve and the coronary artery may explain the rare anomalies in the present case since the pulmonary valve develops between the 6th and 9th week of gestation, concomitant with the development of the truncus arteriosus and aortic sinus wall [7].

Unicuspid pulmonary valvular stenosis is a rare congenital anomaly and usually led to secondary right ventricular infundibular hypertrophy. Most patients with unicuspid pulmonary valvular stenosis were diagnosed and treated in the infancy due to symptoms. In this case, the patient had been asymptomatic except stroke diagnosed 10 years ago. Most serial studies of patients with pulmonary stenosis concentrate on progression of stenotic severity with age but rarely diagnosed till elderly [6]. The therapeutic options for relief of pulmonary stenosis include surgical correction or percutaneous balloon valvuloplasty. Balloon valvuloplasty has emerged as a preferred modality in recent years, but is contraindicated when the obstruction is complicated with infundibular hypertrophy [7]. Due to previous reasons combined with ASD, valve replacement with infundibulectomy may be the preferred treatment in the present case.

**REFERENCE**

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單一心臟冠狀動脈合併多重心臟先天異常：個案報告

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單一心臟冠狀動脈合併其他心臟先天異常是非常罕見，我們報告一個64歲男性病例有多重心臟先天異常，包括單瓣及漏斗部肺動脈狹窄、單一心臟冠狀動脈、冠狀動脈瘤管、及第二型心房中隔缺損。病人接受肺動脈瓣置換手術併漏斗部刮除，及心房中隔缺損修補手術。在手術後，病人症狀完全緩解。

關鍵詞：心房中隔缺損；冠狀動脈瘤管；漏斗部狹窄；肺動脈狹窄；單一心臟冠狀動脈