Single Coronary Artery Arising from the Right Sinus of Valsalva (Case Report)

By

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Summary: A single coronary artery is a rare congenital anomaly of the coronary arteries where only one coronary artery arises from the aortic trunk by a single coronary ostium, supplying the entire heart. We report a case of a 70 years-old man with mitral valvular insufficiency and atherosclerotic right and left circumflex coronary arteries, in whom coronary angiography showed a single coronary artery arising from a single ostium in the right sinus of Valsalva (R-II-B subtype) and transverse trunk coursed between aorta and pulmonary artery. The clinical significance and subtype of the single coronary artery are discussed.

Single coronary artery is rare congenital anomaly of the coronary arteries in which only one coronary artery arises from the aortic trunk with a single coronary ostium.

Thebesius (1716; as cited by Desmet *et al.*, 1992)¹ reported the first case of single coronary artery and Halperin *et al.* (1967; as cited by Desmet *et al.*, 1992)¹ was made the first antemortem diagnosis by means of coronary angiography. The prognosis of individuals with single coronary artery is unclear. Some authors studied sudden death associated with isolated congenital coronary artery anomalies on autopsy population and observed that sudden death occurred in patients with single coronary artery^{2–4}. Yamanaka and Hobbs reported in their paper that a retired professional basketball player (Pete Maravich) with an unsuspected single coronary artery died suddenly during exercise⁵. But some authors didn't find evidence that these patients with single coronary artery in the absence of coronary atherosclerosis have angina, myocardial ischemia or sudden death^{1,6}.

Case Report

The patient is a 70 year-old male with one year history of exertional chest pain and palpitation. In present physical examination, heart rate was 70/min with a regular rhythm, and the blood pressure was 100/60 mmHg. The routine laboratory findings were normal. The electrocardiograph of the patient showed sinus rhythm. An echocardiogram showed mild concentric left ventricular hypertrophy and the first grade of mitral valvular insufficiency. Coronary angiography was then performed using Judking technique in standard projections. In coronary angiography, it was observed that stenosis of the left circumflex, (70% stenosis) and right coronary arteries (minor stenosis), and congenital coronary artery anomaly. The congenital coronary artery anomaly is single coronary artery arising right sinus of Valsalva by a single coronary ostium. The main trunk was short and it was then bifurcated as right and left. In right side, it coursed in the atrioventricular groove in a manner

similar to the normal right coronary artery. In left side, transverse trunk (anomalous left main trunk) coursed between the aorta and pulmonary artery and then gave off the circumflex and anterior descending branches in the usual fashion (Figs. 1, 2, 3). When the sinus node artery examined where it arose, it was observed the sinus node artery arose from the left circumflex coronary artery.

In present case, the single coronary artery was classified as R-II-B subtype single coronary artery (in which the anomalous left main trunk run between the great vessels) according to previous study⁵⁻⁷).

It was thought that his complaints depended on the sclerosis of left circumflex and right coronary arteries and mitral valvular insufficiency and then the patient was maintained on oral medication.

Discussion

A single coronary artery ostium, in the absence of other cardiac disease, is a rare coronary artery

anomaly. As an isolated single coronary artery occurs in approximately 0.024%⁶⁾ to 0.066%¹⁾ of the population. In association with certain other congenital anomalies, however, it is found considerably more frequently; Butto *et al.* reported that the incidence in a series of 54 cases of persistent truncus arteriosus was 18.5%⁸⁾, and in another paper it was reported to be 18%⁹⁾, Shrivastava *et al.* examined coronary angiograms of 296 patients with tetralogy of Fallot and they found a single coronary artery in seven cases¹⁰⁾ and Calder *et al.* found that the incidence of the single coronary artery in pulmonary atresia was around 17%¹¹⁾.

According to the site of origin and anatomical distribution of the branches, isolated single coronary arteries were classified in three groups by Lipton *et al.*,⁶⁾ (Fig. 4) and further modified in 1990 year by Yamanaka and Hobbs⁵⁾. The anomalous coronary artery is first designed with "R" or "L" depending upon whether the ostium is located in the right or left sinus of Valsalva. It is then designated as group I, II, III. Group I have an anatomical course of either a right or left coronary ar-

Fig. 1. The single coronary artery originates from the right sinus of Valsalva. RAO view.
RCA: Right coronary artery. LAD: Left anterior descending coronary artery.

Fig. 2. The single coronary artery originates from the right sinus of Valsalva. Lateral view.
RCA: Right coronary artery. LCx: Left circumflex coronary artery. LAD: Left anterior descending coronary artery.

tery. For example; when the artery originates from the right sinus of Valsalva, it follows the course of a normal right coronary artery, giving off the posterior descending artery in the normal fashion near the crux and then continues in the left atrioventricular groove to give off posterolateral left ventricular branches. Group II anomalies arise from the proximal part of the normal right or left coronary artery, and cross the base of the heart before assuming the normal position of the inherent coronary artery. The subgroups designation of Group II refers to the relation ship between the anomalous coronary artery and the aorta and pulmonary artery. The letters "A", "B", and "P" refer to "anterior", "between", and "posterior" patterns. Yamanaka and Hobbs modified the Lipton's classification by adding "septal" and "combined" types, designated as "S" and "C" in order to describe the anatomical variations more precisely⁵⁾. The present case was classified R-II-B subtype single coronary artery (in which the anomalous left main trunk run between the great vessels) according to previous

study⁵⁻⁷⁾. Angiographically, in the RAO projection, the left main forms a cranial-posterior loop in its initial course between the aortic root and the right ventricular infundibulum at the pulmonic valve level⁷⁾. Group III describes the anomaly where the left anterior descending coronary artery and left circumflex coronary artery arise separately from the proximal part of the normal right coronary artery.

R-II-B subtype single coronary artery is very rare anomaly. This subtype single coronary artery incidence was reported from 0.04%⁶⁾ to 0.004%¹⁾. A review of the literature, it was suggested an association between single coronary artery of type R-II-B or anomalous left main coronary artery right sinus of Valsalva (in which the anomalous left main trunk run between the great vessels) and sudden death during or after exercise²⁻⁴⁾. Cheitlin *et al.* observed that young patients with the left main coronary artery arising from right sinus of Valsalva or right coronary artery and passing between the pulmonary artery and the aorta before bifurcating

Fig. 3. The single coronary artery originates from the right sinus of Valsalva. LAO view.
RCA: Right coronary artery. LCx: Left circumflex coronary artery. LAD: Left anterior descending coronary artery.

died suddenly during or after exercise². Taylor *et al.* also studied sudden cardiac death associated with isolated congenital coronary artery anomalies in 242 patients. They found that sudden death occurred in 6 (14%) patients with single coronary artery of 44 patients with congenital coronary artery anomalies. Sudden death was more common when the single coronary artery originated in the right coronary sinus (18%) versus the left coronary sinus (9%). When they examined an association sudden death and the anomalous left main trunk according to its course, they observed that sudden death was particularly common when the left coronary artery courses between the aorta and pulmonary artery. Twenty-three (82%) of the 28 patients with this anomaly died suddenly³. Frescura *et al.* studied anomalous origin of coronary arteries and risk of sudden death on an autopsy population. They observed the left main coronary artery arising from right sinus of Valsalva in four specimens. All of them died suddenly during exercise⁴.

The exact etiology and mechanisms of sudden death in anomalous coronary arteries are still not

unanimously agreed upon. Four hypotheses have been suggested: 1-Acute angle of take-off of the anomalous coronary artery from the opposite aortic sinus. This becomes exaggerated with the expansion of the ascending aorta associated with exercise, converting the round orifice of the coronary artery into a slit-like opening, which is then actually occluded by a flap-like closure of the orifice². 2-Extrinsic compression of the anomalous coronary artery as it courses between the aorta and pulmonary artery during exercise when the aortic root and pulmonary trunk dilate⁴. 3-Spasm, torsion, or kinking of the anomalous coronary artery³. This mechanism was reported the angiographic demonstration of an anomalous right coronary artery arising from above the left sinus of Valsalva showing both kinking at its origin consistent with the pathologically described slit-like orifice and systolic compression by the aorta and pulmonary artery¹². 4-Superimposed coronary atherosclerosis. Click *et al.* reported that a greater degree of atherosclerotic involvement in the left circumflex coronary arteries¹³. In present case, the left circumflex

Fig. 4. Classification of angiographic types of isolated single coronary artery patterns. Key for abbreviations: R: right, L: left, RCA: Right coronary artery, LCA: left coronary artery, LAD: left anterior descending coronary artery, LCx: left circumflex coronary artery, Ao: aorta, S: septal branches, PA: pulmonary artery, A: anterior, B: between, and P: posterior to the great vessels. Shading of the transverse trunk indicates this artery courses posterior to the aorta⁶⁾.

coronary artery was also more stenosis.

It can be thought that R-II-B subtype single coronary artery is also potentially serious congenital coronary anomaly as the anomalous left main

coronary artery from the right sinus of Valsalva with anomalous course between the great vessels although in the present case, R-II-B subtype single coronary artery was not fatal. Therefore, greater effort for early detection and surgical repair of this anomaly is warranted. The angiographic recognition of this vessel may be useful for physicians dealing with diagnosis and treatment of the R-II-B subtype single coronary artery.

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