

실신을 동반한 우측 관상동맥굴에서 기인한 좌관상동맥의 기형의 1예

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A Case of Anomalous Left Coronary Artery Originating from the Right Sinus of the Valsalva Presenting with Syncope

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Congenital anomalies of the coronary artery are associated with various symptoms including syncope, myocardial ischemia, and sudden cardiac death. The abnormality depends on the adjacent structure and pathway of the coronary artery. Most patients with an anomalous left coronary artery that arises from a right coronary sinus of the valsalva have no symptoms and are usually diagnosed at autopsy. Therefore, their first symptom might present as sudden death, particularly when the left coronary arterial course is between the aorta and the pulmonary trunk. Symptomatic patients could be diagnosed early with an anomalous coronary artery, and the risk of fatal events could be decreased by surgical correction. Here, we report the case of 62-year-old male who experienced a first episode of syncope with an anomalous left coronary artery arising from the right sinus of the valsalva with a separate orifice from the right coronary artery. He is alive and in good health receiving medical treatment, and has had no medical events for over 2 years. (Korean J Med 2014;87:722-727)

Keywords: Coronary vessel anomalies; Sinus of valsalva; Syncope

INTRODUCTION

Coronary arteries normally arise from the “facing” sinuses of the valsalva on either side of the commissural contact. Generally,

the left coronary artery (LCA) arises from a mid-position on the left anterior sinus of valsalva and sinuses on either side of the point of aortic and pulmonary commissural contact, just above the level of the free margin of the aortic valve leaflet and below

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the sinotubular junction. Coronary arteries do not normally arise from the “nonfacing”, or most distant sinus. However, variations in the coronary anatomy are common. Variations that occur in < 1% of the general population are considered abnormal or anomalies [1,2].

There are two major congenital anomalies of the coronary artery from the opposite sinus: the LCA arising from the right sinus of the aorta, and the right coronary artery (RCA) arising from the left sinus of the aorta. The risk of sudden cardiac death is higher in young patients whose left main coronary artery arises from the right aortic sinus because of the resulting passage between the aorta and the right ventricular outflow tract [3,4].

Here, we report a rare case of an anomalous origin of the LCA from the right sinus of the aorta in a patient presenting with syncope.

CASE REPORT

A 62-year-old male presented to the emergency room with a brief episode of syncope. The patient suddenly felt faintness after substernal chest discomfort and palpitation when he was walking without conscious effort. He collapsed and lost consciousness for ~5 min. The patient recovered consciousness in the ambulance, but his chest pain and palpitation persisted. He had a medical history of hypertension, diabetes mellitus, and dyslipidemia. He also ceased smoking 6 years previously. An initial examination revealed a blood pressure of 160/100 mmHg and a heart rate of 72 beats/min.

An electrocardiogram revealed a normal sinus rhythm without ST segment depression and T-wave abnormalities, but a brain computed tomography (CT) showed no pathological findings. The patient was referred to the cardiology department with suspected cardiogenic syncope. The initial laboratory findings revealed 15.2 g/dL hemoglobin, 6,550/μL white blood cells, 133.2 mmol/L Na, 4.34 mmol/L K, 284 mg/dL glucose, 1.68 ng/mL creatine kinase-myocardial band (range, 0-5.0), and 0.003 ng/mL cardiac troponin-I (range, 0-0.06).

Echocardiography showed a normal left ventricular ejection

fraction (62%) without regional wall motion abnormalities. The maximum carotid intima-media thickness (CIMT) was 1.25 mm, without significant luminal narrowing or unstable plaques. He underwent an exercise treadmill test, 24 h of Holter monitoring, and a head-up tilt table test to assess the cause of the syncope, but all were negative.

Coronary angiography showed that the left main coronary artery originated from the right sinus of the aorta with a separate origin, and the LCA proximal portion exhibited systolic luminal narrowing such as myocardial bridging. There was no evidence of any significant luminal stenosis or ruptured plaque in either coronary artery (Fig. 1). CT angiography showed that the long left main coronary artery originated from the right coronary sinus of the aorta with separate orifices. These two separate orifices were in close proximity but originated independently from the right coronary sinus of the aorta. In addition, the course of left main coronary artery was between the aorta and the pulmonary trunk (interarterial pathway), and proximal segment exhibited periodic narrowing during ventricular systole. The RCA arose normally from the right coronary sinus of the aorta, and a mild calcified plaque was seen in the mid-RCA (Fig. 2).

Coronary artery surgery was recommended to reduce the risk of fatal cardiac events, which might be caused by external compression of the proximal portion of the LCA during exertion. However, the patient did not want surgery, and he had no symptoms during ordinary exercise. We finally made a decision to treat medically using a β -adrenergic blocker and a calcium channel blocker. The patient lives in good health with regular follow-up at the outpatient department. He had no further medical events in the subsequent 2 years.

DISCUSSION

Congenital anomalies of the coronary artery have various symptoms, including syncope, myocardial ischemia, and sudden cardiac death, that depend on the surrounding structural location around the origin and termination of the coronary artery [5-7]. Therefore, the origin and pathway of the anomalous coronary ar-

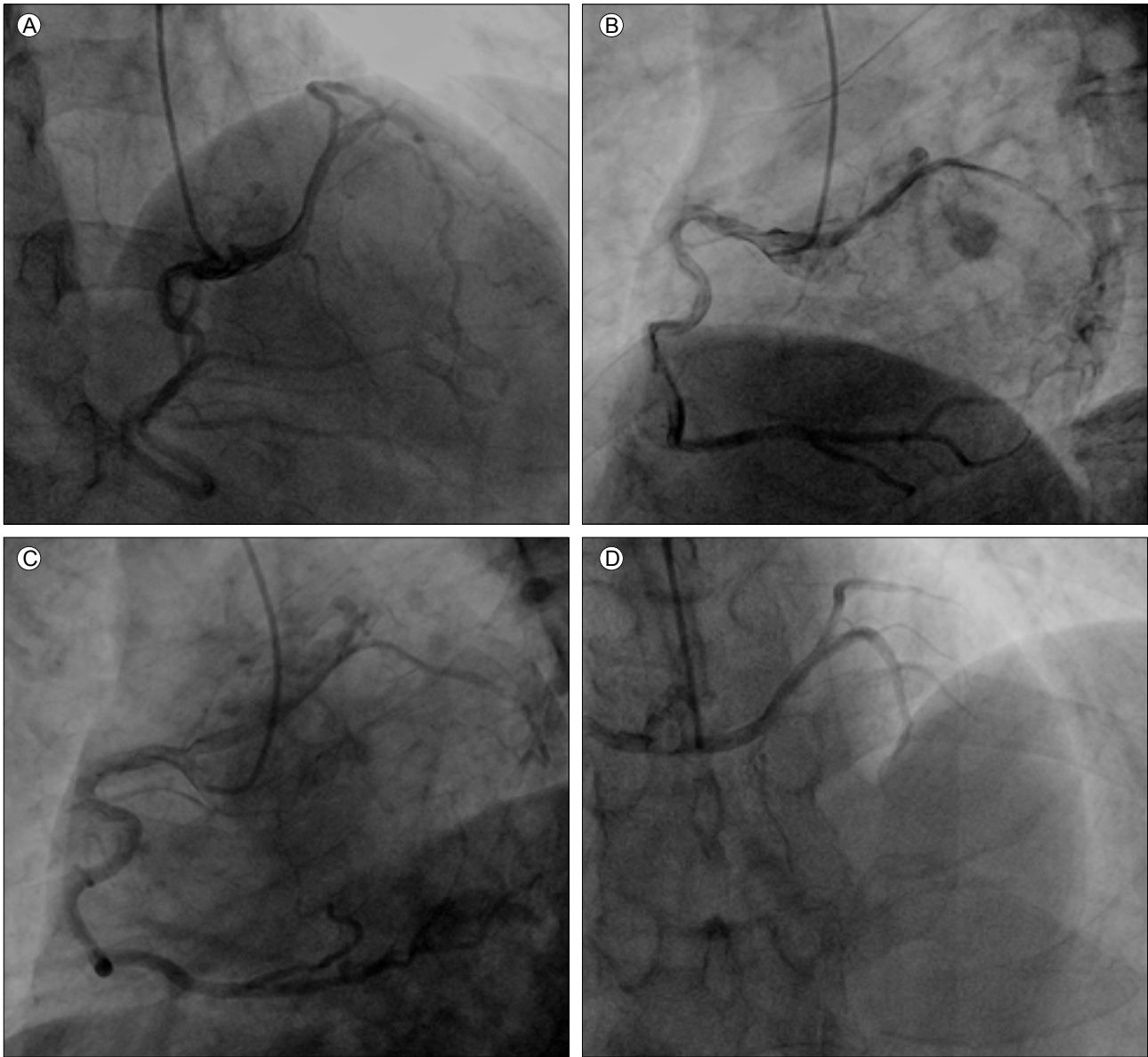


Figure 1. Coronary angiography. Left main coronary artery originating from the right sinus of the valsalva with a separate origin. There is no significant luminal narrowing or ruptured plaques. The right coronary artery also originated from the right coronary sinus of the valsalva without significant luminal narrowing. (A) Right coronary artery (RCA): right anterior oblique view. (B) RCA: left anterior oblique view (LAO). (C) RCA: LAO view. (D) RCA: LAO view.

teries and surrounding structures must be identified and classified.

There are four common pathways of LCA anomalies that arise from the right coronary sinus of the valsalva: interarterial, anterior, posterior, and septal (Fig. 3). All of these predispose otherwise young, healthy individuals to myocardial ischemia or sudden cardiac death [8]. An interarterial pathway with an anom-

alous LCA arising from the right coronary sinus of the valsalva has an acute angle of origin and resulting pathway. This pathway might cause stenosis during extreme exercise because of the increased cardiac output and circulation, which expands the aorta roots and pulmonary trunk. These anatomical pathways and dynamic structural changes compress the anomalous coronary artery externally between the expanded great arteries. Thus, my-

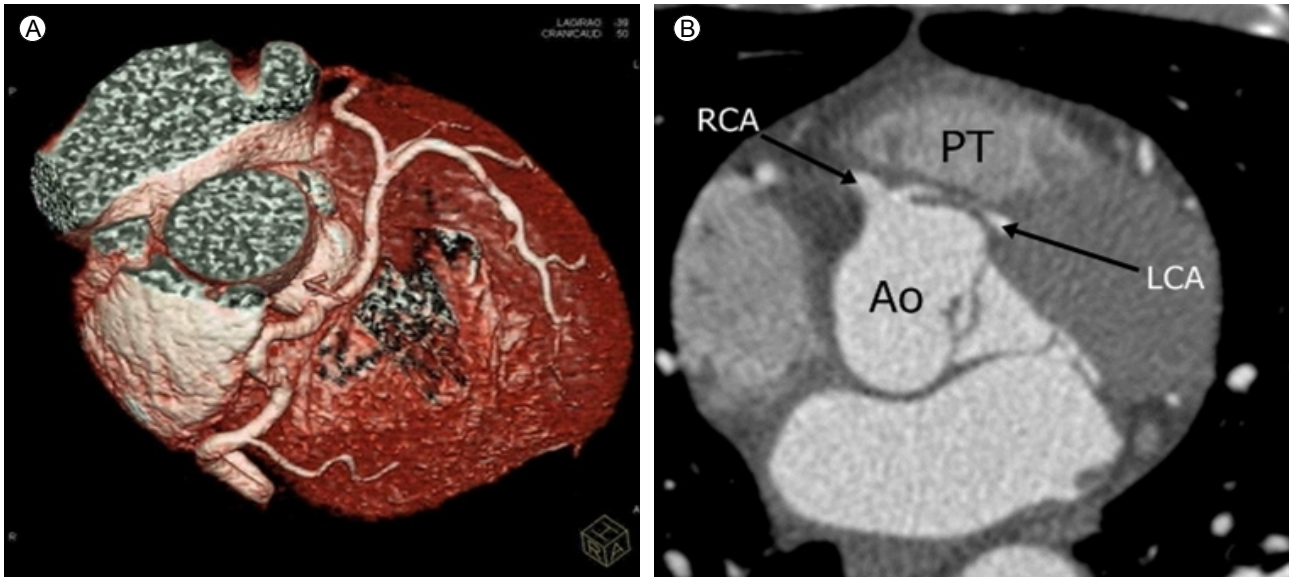


Figure 2. Coronary computed tomography (CT) angiography. (A) Coronary CT angiography 3D image. A long anomalous left main coronary artery arising from the right coronary sinus of the valsalva with separate orifices. Two separate orifices were in very close proximity but with independent origins in the right coronary sinus of the valsalva. (B) LCA arising from the right coronary sinus of the valsalva. The course of the left main coronary artery was between the aorta and the pulmonary trunk (interarterial pathway). The proximal segment of the LCA showed periodic narrowing during ventricular systole. The RCA arises normally from the right coronary valsalva sinus; a mild calcified plaque was seen in the mid-RCA. RCA, right coronary artery; PT, pulmonary trunk; Ao, aorta; LCA, left coronary artery.

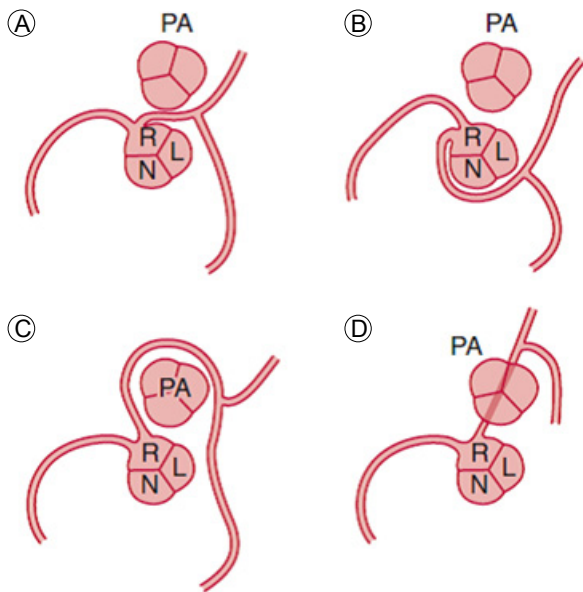


Figure 3. Four subtypes of anomalies in the left coronary artery arising from the right coronary sinus of the valsalva [10]. (A) Interarterial, between the aorta and the pulmonary artery (PA). (B) Retroaortic, beneath the right ventricular outflow tract. (C) Prepulmonic, beneath the right ventricular outflow tract. (D) Septal, beneath the right ventricular outflow tract. R, right coronary sinus; L, left coronary sinus; N, noncoronary sinus.

cardiac ischemia and cardiac sudden death are common in patients with interarterial pathway [7,9,10]. Most patients with an anomalous LCA arising from the right coronary sinus of the valsalva have no symptoms, and anomalous coronary artery is diagnosed at autopsy. Approximately 18-30% of patients with an anomalous coronary artery have symptoms as syncope, dizziness, chest pain, and dyspnea. These symptomatic patients could be diagnosed early with an anomalous coronary artery, and coronary artery bypass surgery might decrease the risk of fatal events.

The methods used to screen patients with a suspected anomalous coronary artery are important. First, echocardiography might establish a diagnosis of anomalous coronary artery. In addition, coronary angiography, coronary CT angiograms, and coronary magnetic resonance angiography might be helpful for establishing a diagnosis of anomalous coronary artery.

In the current case, the middle-aged patient had underlying diseases such as diabetes mellitus and hypertension, but had experienced no coronary symptoms previously. However, he was

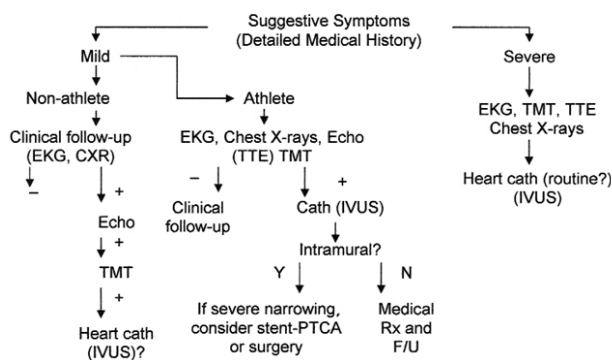


Figure 4. The proposed diagnostic protocol for adult patients who are at risk of coronary artery anomalies [7]. —, negative test result; +, positive test result; CXR, chest x-ray; Echo, echocardiography; EKG, electrocardiogram; F/U, follow-up; IVUS, intravascular ultrasound; N, no; PTCA, percutaneous transluminal coronary angioplasty; Rx, treatment; TMT, treadmill test; TTE, transthoracic echocardiography; Y, yes.

transferred to the hospital with a brief episode of syncope, chest pain, and palpitation. Many evaluations were performed, including a brain CT, an electrocardiogram, CIMT, a treadmill test, a head-up tilt table test, and echocardiography, but all yielded normal findings, except for diastolic dysfunction. Coronary angiography showed no significant stenosis of the RCA; however, an anomalous LCA arising from the right coronary sinus of the aorta without stenosis was found. A multi-detector coronary CT angiography was used to evaluate of exact pathway and its subtype. It was a type A (interarterial) pathway of an anomalous LCA arising from the right coronary sinus of the aorta.

After identifying and classifying the coronary anomalies, determining the clinical severity is important. Therefore, provocation tests are indispensable for evaluating the clinical relevance. The relationship between the coronary anomaly and the presenting symptoms is frequently unclear. However, the aim is to ascertain patient prognosis to define and begin the appropriate treatment. In this case both a treadmill and head-up tilt table test were negative. Additional stress studies such as stress echocardiography or fraction flow reserve examinations might have been helpful. However, these tests were not performed because the clinical severity was not significant (Fig. 4) [7].

Surgical intervention is indicated in cases with documented myocardial ischemia when the LCA arises from the right sinus

of the aorta and has an intramural course or passes between the great arteries. Surgical revascularization using coronary artery bypass grafting, unroofing, or coronary reimplantation yields favorable results. Coronary artery stenting also has good short-term results, and could be the only option for critically ill patients. In the current case, the patient had no symptoms, and so he wanted to receive only medical treatment. Therefore, we prescribed a calcium channel blocker and a β -adrenergic blocker to reduce the dp/dt .

The patient remains in good health and is receiving regular follow-up; he has had no further medical events. However, we will recommend that he undergo immediate surgical correction if he experiences any cardiac ischemic symptoms or syncopal episodes during subsequent follow-up examinations.

Anomalous coronary arteries are rare, and so are missed easily until fatal symptoms occur. Congenital anomalies of the coronary artery present with various symptoms, including syncope, myocardial ischemia, and sudden cardiac death, depending on the surrounding structural location, origin, and pathway of the coronary artery. In this case, we reported an anomalous left coronary artery arising from the right coronary sinus of the aorta. The case presented with syncope in a middle-aged male, who has lived in good health with medical treatment for 2 years without further events.

중심 단어: 관상동기형; 관상동맥굴; 실신

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