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Severe Form of Persistent Thebesian Veins Presenting as Ischemic Heart Disease

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Coronary artery fistula is a rare congenital anomaly. Most patients with this anomaly are asymptomatic, but some may develop heart failure, myocardial ischemia or arrhythmias. We report a case of a patient who presented with myocardial ischemia secondary to persistent Thebesian veins. Coronary angiography demonstrated a marked capillary blush draining into the left ventricular cavity through multiple microfistulae from the left anterior descending artery, left circumflex artery and right coronary artery. The patient was discharged without chest pain and was medically maintained with a beta-blocker and angiotensin converting enzyme inhibitor. **(Korean Circ J 2012;42:714-717)**

KEY WORDS: Coronary vessel anomalies; Coronary angiography; Arteriovenous fistula; Myocardial ischemia.

Introduction

A coronary arterial fistula is a connection between one or more of the coronary arteries and a cardiac chamber or great vessel, having bypassed the myocardial capillary bed. The feeding artery of the fistula may drain from a main coronary artery or one of its branches and is usually a dilated and tortuous artery terminating in one of the cardiac chambers or great vessels. Coronary arterial fistulas are usually asymptomatic, especially when they are hemodynamically insignificant. Complications can arise from a coronary 'steal' phenomenon occurring from the adjacent myocardium causing myocardial ischemia, thrombosis and embolism, heart failure, rupture, endocarditis, endarteritis and arrhythmias.¹⁾²⁾ There have been many reported cases of coronary artery fistulas,³⁻⁵⁾ but only few reports of Thebesian veins in Korea.⁶⁾⁷⁾ Here, we presented a unique case of

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extensive persistent Thebesian veins in an old patient presenting as acute coronary syndrome.

Case

An 83-year-old woman presented with progressive exertional chest pain and dyspnea. She was an active smoker (1 pack per day during 50 years) with a past medical history of depression requiring pharmacotherapy. The patient's blood pressure was 145/48 mm Hg with a pulse rate of 62 beats per minute. Cardiac murmur was not auscultated on physical examination. Although her initial electrocardiography (ECG) showed sinus rhythm without ST segment changes, after 2 hours of observation, T-wave inversions on lead V2, V3 and V4 were recorded (Fig. 1). Troponin I level was not elevated throughout her admission. The patient was treated for an acute coronary syndrome and her symptoms were relieved with intravenous nitroglycerin. Coronary angiography demonstrated a marked capillary blush draining into the left ventricular (LV) cavity due to extensive multiple microfistulae arising from the left anterior descending artery (LAD), left circumflex artery and right coronary artery (RCA) producing a left ventriculography (Fig. 2). The left main coronary artery (LMCA) was severely dilated (up to 10 mm) and the LAD was diffusely calcified with a 40% narrowing at the proximal LAD. RCA was also markedly dilated with a discrete 30-40% luminal narrowing at the posterior descending artery and posterior lateral artery bifurcation. Echocardiography demonstrated concentric LV hypertrophy, normal LV chamber size and normal LV systolic function

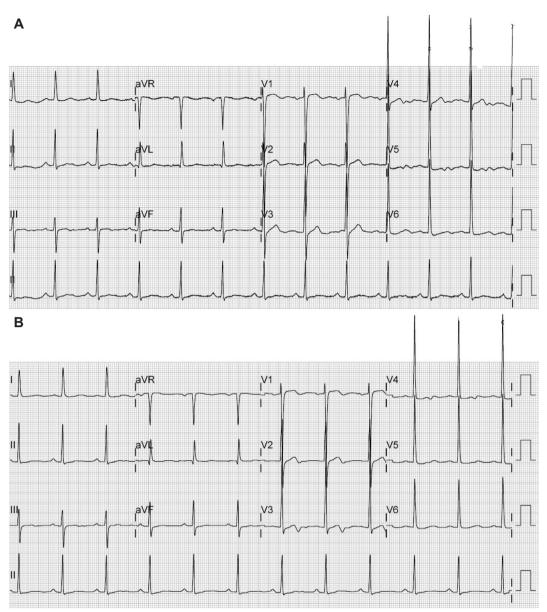


Fig. 1. Twelve-lead ECG showing sinus rhythm with T-wave inversion in V 2-4 2 hours after admission (B) compared to initial ECG (A). ECG: electrocardiography.

without the regional wall motion abnormality. She remained asymptomatic and was discharged on a beta-blocker and angiotensin converting enzyme (ACE) inhibitor. Two months later, the T-wave inversions at V2, V3 and V4 were normalized.

Discussion

Coronary artery fistula is a rare congenital anomaly with an incidence of 0.2% in patients undergoing diagnostic cardiac catheterization.⁸⁾ Drainage most frequently occurs into the right ventricle. Fistulas terminating in the left ventricle are rare. Thebesian sinusoids that supply nutrients in the embryonic heart are responsible for coronary artery to ventricular shunts. The pathogenesis of these fistulous communications has been attributed to a persistence of intertrabecular spaces in localized areas of the myocardium.⁹⁾ Embryonic sinusoids are formed from endothelial protrusions that extend into the epicardial surface joining the coronary arteries. This network normally becomes obliterated into a capillary network and the Thebesian vessels. A fistulous communication may be due to a failure of these intertrabecular spaces to fuse. In the present case, all the three coronary arteries had persistent Thebesian veins. The shunt flow was so large that the LMCA was severely dilated and coronary angiography was sufficient to display ventriculography. Her wide pulse pressure and low diastolic pressure might be possibly

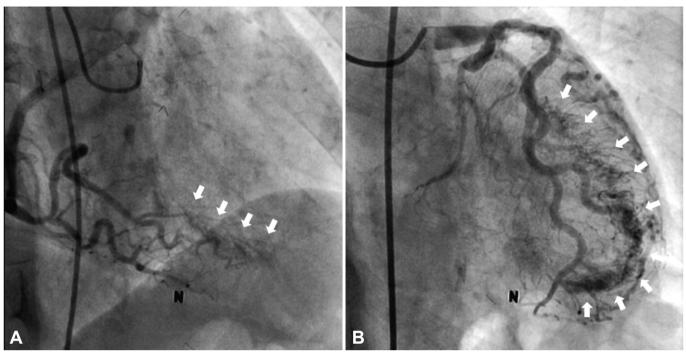


Fig. 2. Coronary angiogram showing right coronary artery (A) and left main coronary artery (B). Arrows indicate multiple arteriosinousoidal fistulae filling with contrast agent on injection, visualizing the endocardial border (A and B).

due to the coronary fistula.

Approximately half of all patients with coronary artery fistulae remain asymptomatic; the other half develop congestive heart failure, infectious endocarditis, myocardial ischemia induced by a coronary steal phenomenon, or rupture of an aneurysmal fistula.⁹⁾ In coronary artery fistulae to the left ventricle, most common presenting symptoms are chest pain, exertional dyspnea and fatigability.¹⁰⁾

Treatment of a large single fistula involves surgical correction or coil embolization whereas small multiple fistulae are usually managed medically similar with the present case, although there are controversies on optimal medications. For example, there was a report that showed myocardial ischemia aggravation following nitrate therapy.¹¹ Nitrate therapy probably increases ischemia by both increasing leakage into the left ventricle and decreasing the overall coronary artery flow to the myocardium. However, chest pain was relieved after the intravenous nitrate administration in the present case which suggested that mechanism of ischemia from persistent Thebesian veins remains to be elucidated. In addition to nitrate, we prescribed a beta-blocker and ACE inhibitor to reduce cardiac workload and afterload and to increase systemic flow. Further discussion and research are required to seek for the optimal medical therapy for this severe form of Thebesian veins.

Because our patient showed typical symptoms of exertional chest pain and ECG findings consistent with myocardial ischemia and responded to medical treatment for acute coronary syndrome, we did not perform additional tests to exclude other causes of chest pain. However, further examination including upper gastrointestinal endoscopy, computed tomography and ergonovine provocation test could be considered for persistent symptoms or signs suggestive of other causes of chest pain.

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