An Unusual Form of Coronary Artery Fistula: A Small Aneurysm of Vieussens’ Arterial Ring Communicating with the Pulmonary Artery

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Vieussens’ arterial ring (VAR) is the connection between the conus branch of the right coronary artery and the proximal right ventricular branch of the left anterior descending coronary artery. VARs are found in 48% of the population; however, pathologic VAR is rare. We experienced a case of pathologic VAR that involved a fistula connecting to the main pulmonary artery.

Key words: 1. Coronary vessel anomalies
2. Fistula

CASE REPORT

A 53-year-old male presented chest discomfort and palpitations. Electrocardiography performed on admission revealed sinus tachycardia with no ischemic change. His blood pressure was 143/80 mmHg and pulse rate was 95 beats/min. Laboratory results including complete blood count and cardiac enzymes were within the normal range. There was no cardiomegaly or evidence of active lung lesion on chest radiography. Transthoracic echocardiography revealed normal ejection fraction, chamber size, and valve function, and regional wall motion abnormality was not present. However, there was abnormal blood flow into the pulmonary artery on the parasternal short axis view, suggesting a coronary artery fistula (CAF). The patient underwent coronary angiography, which revealed a coronary fistula between the proximal left anterior descending coronary artery (LAD) and the pulmonary artery (Fig. 1). The right coronary artery (RCA) also had a congenital anomaly. The right conal artery, originating from a separate orifice of the right coronary sinus and draining into

Fig. 1. Coronary angiography. Left coronary angiography demonstrated an anomalous coronary fistula from the proximal left anterior descending coronary artery to the main pulmonary artery (arrow).
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Fig. 2. Coronary angiography. Right coronary angiography demonstrated the conus branch from a separate coronary ostium. The right conal branch has a fistula to the pulmonary artery (arrow).

Fig. 3. Three-dimensional reconstruction of computed tomography coronary angiography. The right conal branch had a separated coronary ostium and was connected to the proximal left anterior descending coronary artery via Vieussens’ arterial ring.

Fig. 4. Three-dimensional reconstruction of computed tomography coronary angiography. The right conal branch and the proximal left anterior descending coronary artery formed Vieussens’ arterial ring (arrow), which showed aneurismal dilatation prior to entering the main pulmonary artery.

MDCT showed an abnormal connection between the right conal artery and the LAD draining to the pulmonary artery via a 6.2-mm aneurysm. In other words, the right conal artery originating from a separate orifice of the right coronary sinus had an abnormal connection with the RCA via a short vessel just distal to its origin. It passed up and over the right ventricular outflow tract to provide flow to the aneurysm, which was connected to the LAD via a highly tortuous artery. There was also a fistula between the aneurysm and the main pulmonary artery (Figs. 3, 4). These features were compatible with the diagnosis of an aneurysm of VAR. The patient underwent an echocardiographic stress test and treadmill test to investigate cardiac ischemia; these tests showed no ischemic change. Surgical resection was offered as a treatment option; however, the patient refused treatment for personal reasons and is being followed closely.

DISCUSSION

CAFs are abnormal connections between coronary arteries and cardiac chambers or great vessels, which have an incidence of approximately 0.87% [1]. Most fistulae originate
from the right coronary or the left anterior descending artery, and both coronary arteries are the site of the CAF in approximately 5% of the cases. In the present case, the fistula, which showed aneurismatic change, was fed by both the right conal artery and the LAD, and drained into the main pulmonary artery. These findings are compatible with the diagnosis of an aneurysm of VAR with a fistula to the pulmonary artery. Many studies have investigated CAFs and have reported that they have several possible origins; however, reports of CAFs of VAR are rare. VAR was first described in 1706 by Raymond de Vieussens [2] and is described as a connection between the conus branch of the RCA and the proximal right ventricular branch of the LAD. It is thought to be a remnant of the embryonic conotruncal ring and is found in 48% of the population [3]. However, the primary pathological features of VAR are rare. When significant stenosis occurs in the LAD or RCA, VAR will dilate and provide collateral blood flow to the left or right arterial system [4]. In our case, however, although there was no significant stenosis in the coronary artery system, hypoplastic RCA was observed. Atallah et al. [5] reported a case of VAR with a congenitally hypoplastic left coronary arterial system. In the case considered here, they demonstrated that VAR could not be the product of collateral circulation due to obstructive lesions but was the result of congenitally hypoplastic left arterial circulation. In consideration of these features, in our case, VAR was also deemed to be unrelated to collateral circulation due to obstructive lesions; rather, it was the result of a congenitally hypoplastic right coronary system. There have been previous reports of an aneurysm of VAR; however, reports of a fistula between VAR and the main pulmonary artery are rare. The management of CAFs involving the pulmonary artery via VAR is no different from that of a typical CAF, although this remains controversial. However, a majority of these opinions advocate surgical management for symptomatic patients because percutaneous intervention can cause a secondary ischemic event due to the accompanying atherosclerosis. Further, the results of numerous surgical modalities are reported to be excellent.

**CONFLICT OF INTEREST**

No potential conflict of interest relevant to this article was reported.

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