

A giant sinus node artery fistulizing to superior vena cava presenting with steal phenomenon: An unusual case report

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ABSTRACT

Congenital coronary fistulas are rare anomalies. They can be either symptomatic or asymptomatic. Management strategy for coronary fistulas differs depending on the blood flow and symptoms. Nevertheless, it should be considered a serious health problem which may result in unpleasant complications such as congestive heart failure through left-to-right shunt, myocardial ischemia through steal phenomenon, endocarditis, and aneurysmal ruptures. Herein, we present the surgical management of a young female who was referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity caused by a giant sinus node artery fistulizing to the superior vena cava.

Keywords: Coronary artery disease, coronary artery fistula, sinus node artery, vena cava superior.

Congenital coronary fistulas are rare anomalies with an incidence of 0.1 to 0.2%.^[1] First described by Krause in 1865,^[1] they occur as a result of anomalies in the embryology of the coronary circulation. The first surgical procedure was performed by Bjork and Crawford in 1974.^[2] The right coronary artery is the most common site of origin, and the right ventricle and atrium are the most common areas of opening.^[2] Herein, we present the surgical treatment of a rare and giant coronary fistula fistulizing to the superior vena cava through the sinus node artery.

CASE REPORT

A 31-year-old female patient was referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity. Her medical history revealed several treatments applied to relieve extracardiac causes-related complaints last year. Physical examination showed no pathology. Chest X-ray and electrocardiography demonstrated cardiomegaly findings. After cardiac catheterization, an extraordinary tortuous arterial structure extending from the right coronary artery to the superior vena cava was observed (Figures 1 and 2). At the preoperative examination, the aneurysmatic right coronary artery was found to give a sinus node artery branch approximately 2 cm after the separation from the

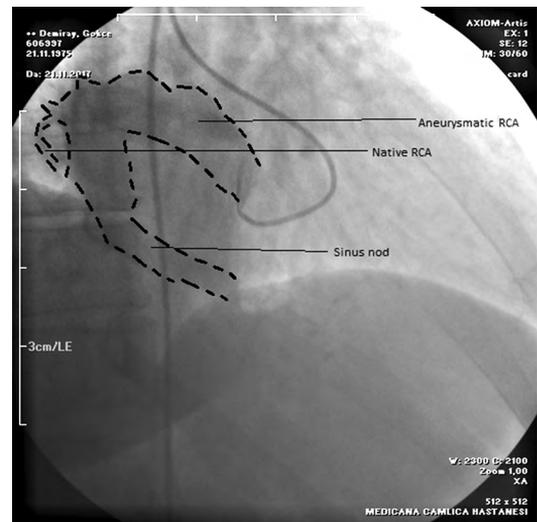


Figure 1. Appearance of fistula tract during coronary angiography.

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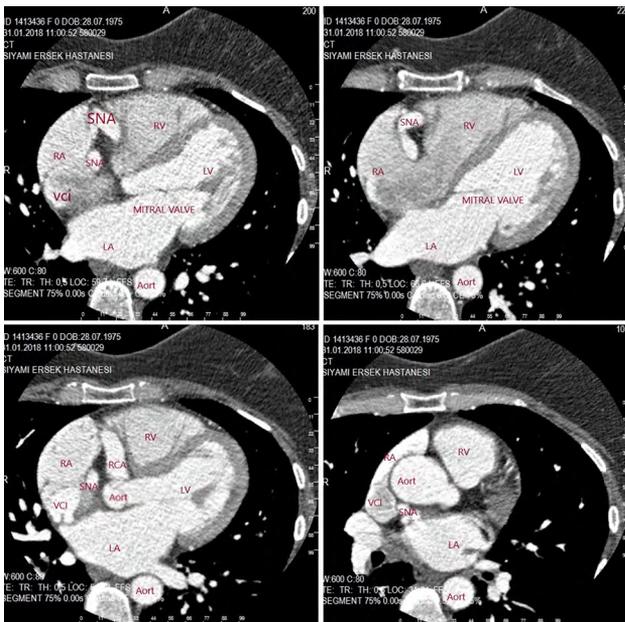


Figure 2. Computed tomography angiography showing aneurysmatic right coronary artery and sinus node artery. Right coronary artery is seen to run in normal diameter after giving the sinus node artery branch. RV: Right ventricle; RA: Right atrium; LV: Left ventricle; LA: Left atrium; SNA: Sinus node artery; VCI: Vena cava inferior.

aorta, and the diameter decreased to 4 mm there. The diameter of the sinus node artery was 1.5 cm. After running about 5 cm, it opened medially into



Figure 4. A view of resected aneurysmatic sinus node artery (lower) and an aneurysmatic segment of right coronary artery (upper).

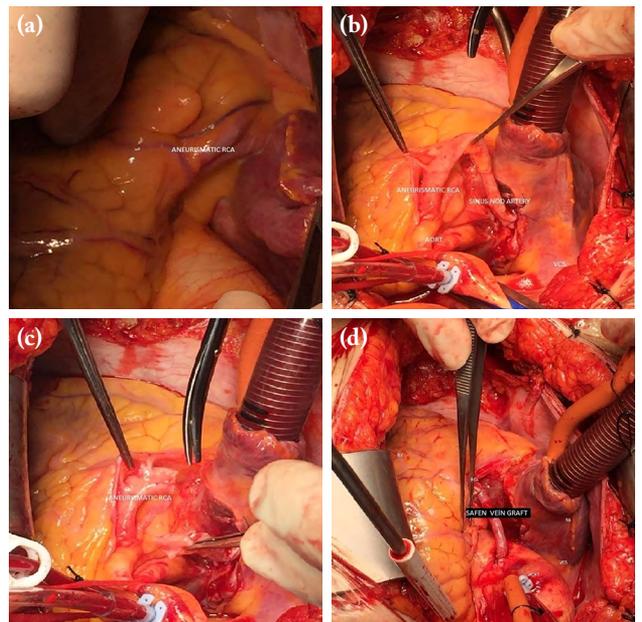


Figure 3. (a) View of the aneurysmatic right coronary artery in the anterolateral segment of the proximal aorta. (b) Aneurysmatic dilatation of the sinus node artery after exploration. (c) The appearance of the undeveloped right coronary artery after separation of the aneurysmatic sinus node artery. (d) Appearance of aorta-right coronary artery bypass with saphenous graft.

the superior vena cava, leading to a murmur of 4/6. A 2×2-cm venous aneurysm formation was observed at the opening (Figure 3a-c). Surgery was decided. A written informed consent was obtained from the patient.

Surgery was performed using standard surgical procedures with median sternotomy and bicaval cannulation. Considering the increased likelihood of rupture, the aneurysmatic proximal 2-cm segment of the right coronary artery was removed (Figures 3d and 4). The saphenous vein graft was interposed between the aorta and distal-free end of the right coronary artery (Figure 3d). The sinus node artery was also aneurysmatic and tortuous. It was excised completely and the point at which it opened into the superior vena cava was sutured. The operation was performed under cardiopulmonary bypass to avoid rhythm disturbances. After giving the sinus node artery, the right coronary artery continued about 10 cm in its normal diameter (Video 1). The postoperative course was uneventful and the patient was discharged on the postoperative fifth day.

DISCUSSION

In the presence of coronary artery disease, angina is a common finding in elderly patients. However, in younger patients and, particularly in females, angina should suggest congenital arteriovenous fistulas. Although coronary fistulas are mostly congenital, they may occur after blunt external thoracic trauma, myocardial infarction, angioplasty or cardiac surgery.^[3] Other congenital anomalies may be also accompanied by coronary fistulas with an incidence of 40%.^[3] They become symptomatic after the third decade of life. The angina is caused by coronary steal. Fistulas opening into the right heart chambers create shunts from left-to-right which may result in congestive heart failure in proportion to the size of the shunt. Such symptoms as angina pectoris, fatigue, shortness of breath, palpitation, and findings suggestive of rupture of an aneurysm, embolism, endocarditis, congestive heart failure can be seen. The degree of symptoms is proportional to the amount of physiological coronary stealing phenomenon developed by the fistula tract.^[4] The frequency of the continuous murmur, the most frequent physical examination finding, ranges from 20 to 80%.^[5] Almost half of the patients have non-specific electrocardiographic changes. Cardiomegaly can be observed on telecardiography due to an increased cardiac output.^[1] For a definitive diagnosis, selective coronary angiography is required. Thus, the origin and termination of the fistula, the path it follows anatomically, and the affected structures can be precisely visualized.

Coronary fistulas mainly originate from the right coronary artery, less frequently from the left coronary artery, or both. They were found to originate from the right coronary artery with a frequency of 19.7% in the Albeyoglu et al.,^[6] 50% in the Levin et al.,^[7] 65% in Lowe,^[8] and 51% in the Wilde and Watt^[9] series. Drainage occurs to the low-pressure heart chambers. Frequency in a decreasing order is as the right ventricle (39%), right atrium (33%), pulmonary artery (20%), left atrium, coronary sinus, vena cava superior, and bronchial arteries.^[1,7] Drainage to the left heart chambers is relatively rare (2%).^[1] Pathophysiological, myocardial ischemia or infarction occurs, when the blood to be directed to the left ventricle is directed to the low-resistance heart chambers through the fistula, creating a steal phenomenon. In fistulas which open to the left ventricle, the blood flow is frequently diastolic. Ischemia occurs, when the pulse pressure

providing the coronary flow is lowered by steal phenomenon.^[6] One of the potential complications is premature atherosclerosis, resulting in an intimal damage caused by high-volume blood flow.^[3] Our case had primarily angina symptoms and tachycardia episodes.

Nonetheless, treatment of asymptomatic fistulas is still controversial. However, most surgeons agree that they should be closed in the presence of a significant shunt or aneurysmal dilatation.^[1] Symptomatic patients need to be treated. Cardiopulmonary bypass may be required or not.^[9] Fistulas which are intramural, short, close to the sinus Valsalva and characterized by an aneurysm are often closed using cardiopulmonary bypass, while extramural and anatomically accessible ones are closed using simple ligation and resection. In addition, closure with the use of percutaneous transcatheter closure devices has become increasingly widespread, particularly in pediatric patients.^[10] Some authors do not consider surgery in asymptomatic patients, while surgery is appropriate for moderate to large-flow fistulas. Small fistulas are likely to spontaneously close as primarily or secondarily. Nevertheless, most authors consider the surgical removal of fistulas to prevent complications and sudden death risk, regardless of whether they are symptomatic or not. The reported operative mortality and morbidity rates are very low. It should be kept in mind that fistula complications may increase surgical morbidity and mortality.

In conclusion, we tried to evaluate giant sinus node artery fistula in the light of literature data. In particular, considering the causes of chest pain and arrhythmia in young patients, the consideration of coronary arteriovenous fistulas is important for the correct diagnosis.

Declaration of conflicting interests

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