

An unusual type of single coronary artery anomaly in a patient with hypertrophic obstructive cardiomyopathy

Hipertrofik kardiyomiyopatisi olan hastada ilginç bir tek koroner arter anomalisi

Hürkan Kurşaklıoğlu, Atilla İyisoy, Turgay Çelik, Cem Barçın, Sedat Köse, Ersoy Işık

From the Department of Cardiology, Gulhane Military Medical Academy, Etilik, Ankara, Turkey

Introduction

When there is a single coronary aortic ostium for all the coronary blood flows, the condition is called a single coronary artery (1-3). Indeed, this kind of anomaly is a group of anomalies with one common property of a single aortic ostium, showing great variations in coronary origination and distribution (4). In this report, we present a case of an unusual type of single coronary artery in a patient with hypertrophic obstructive cardiomyopathy.

Case report

A 22-year-old man presented to our institution with exertional chest pain, which had started one month ago. He had been smoking a pack-a-day for the last 4 years. In his family history, his brother died of a sudden death at the age of 24. On admission, his arterial blood pressure was 130/70 mmHg; pulse rate was 78 bpm. A systolic murmur with a grade of 2/6 on the left border of sternum, which increased in intensity with standing, was noted. Electrocardiography showed symmetric T-wave inversion in the leads V1-V6. Echocardiographic examination revealed that the wall thickness of interventricular septum was 18 / 25 mm and the thickness of left posterior wall was 7 / 13 mm in diastole and systole, respectively. There was a systolic gradient of 68 mmHg in the left outflow tract. A moderate mitral insufficiency and systolic anterior motion of anterior leaflet (grade 2) were also noted. The findings of left ventriculography performed with Judkins' technique via right femoral artery were consistent with hypertrophic cardiomyopathy with a systolic gradient of 59 mmHg between left ventricle and aorta (Fig. 1). Coronary angiography revealed that right coronary artery arose as a terminal extension of the left anterior descending artery, traversed in the posterior interventricular sulcus retrogradely, and finally gave several minor branches to the right side of the heart (Fig. 2). There were no luminal stenoses in the coronary arteries. Moreover, aortography showed one coronary ostium in the left sinus of Valsalva and no ostium in the right sinus of Valsalva (Fig. 3).

Discussion

In the general population, the incidence of single coronary artery is approximately 0.024% (1,3). There are several classifications for single coronary artery. However, the most validate in the literature is the classification by Lipton et al (1). According to this classification, the single coronary artery arising from the left sinus of Valsalva is called L-type and its origination from the right side is called R-type. In this classification, the case in which right coronary artery arises as a terminal extension of left circumflex artery is called as Group L I. According to the course of right coronary artery, in case of the right coronary artery passing anterior to the right ventricular infundibulum and reaching the right atrioventricular groove, it is called Group L IIA; if the right coronary artery passes between the right ventricular infundibulum and aorta, then it is called Group L IIB. On the other hand, if the single coronary artery passes posterior to the aortic root toward the left atrioventricular groove, it is called Group L IIP. The unique features of our case did not fit to this classification. In our case, the ostium of single coronary artery was in the left sinus of Valsalva. The right coronary artery continued as a terminal extension of the LAD, not as a branch of left main, circumflex artery or LAD. One point that should be stressed in the differential diagnosis in case of one coronary artery continuing as a terminal extension of any other vessel is the situation in which the vessel occluded and collateralized from one of the other vessels. There was no suspicion that the right coronary artery was not occluded in our case, since there were no history of previous myocardial infarction, wall motion abnormality on the echocardiography, and more importantly the largest diameter of the artery traversing in the posterior interventricular sulcus was the part, which combined with the LAD and it was getting smaller while traversing up in the sulcus.

Our case was an unusual type of single coronary artery anomaly in a patient with hypertrophic obstructive cardiomyopathy. Although the patients with hypertrophic obstructive cardiomyopathy almost always have small vessel disease (5), there is no specific coronary artery anomaly in association with hypertrophic obstructive cardiomyopathy. So, we consider that the asso-

ciation of single coronary artery with hypertrophic obstructive cardiomyopathy is only a co-incidence. Atmaca et al. (6) reported a case of non-obstructive hypertrophic cardiomyopathy with a single coronary artery, in which the right coronary artery was arising as an extension of circumflex artery. However, there is not enough data in order to define if there is an association of this kind of single coronary artery with hypertrophic cardiomyopathy.

In general, single coronary artery anomaly may not cause any disorder in the blood distribution if there is no stenotic lesion. However, it may rarely cause significant ischemia depending on the course of proximal vessels. Hence, we considered that hypertrophic obstructive cardiomyopathy could be the cause of the ECG changes in our case. The patients with hypertrophic obstructive cardiomyopathy can suffer from classical chest pain even if there are no lesions in their coronary arteries (7-9). Unfortunately, there was no data about the reason for the sudden death of his brother. But, the reason of sudden death could probably be related to hypertrophic obstructive cardiomyopathy with familial transmission rather than coronary artery anomaly.

In conclusion, our case is the first reported single coronary artery anomaly with right coronary artery traversed as a terminal extension of the LAD in a patient with hypertrophic obstructive cardiomyopathy.

References

1. Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology* 1979;130:39-47.
2. Sharbaugh AH, White RS. Single coronary artery. Analysis of the anatomic variation, clinical importance, and report of five cases. *JAMA* 1974;230:243-6.
3. Shirani J, Roberts WC. Solitary coronary ostium in the aorta in the absence of other major congenital cardiovascular anomalies. *J Am Coll Cardiol* 1993;21:137-43.
4. Bittner V, Nath HP, Cohen M, Soto B. Dual connection of the left anterior descending coronary artery to the left and right coronary arteries. *Cathet Cardiovasc Diagn* 1989;16:168-72.
5. Maron BJ, Wolfson JK, Epstein SE, Roberts WC. Intramural ("small vessel") coronary artery disease in hypertrophic cardiomyopathy. *J Am Coll Cardiol* 1986;8:545-57.
6. Atmaca Y, Dandachi R, Oral D. A rare variant of single coronary artery and non-obstructive hypertrophic cardiomyopathy. *J Invasive Cardiol* 2002;14:469-70.
7. Pasternac A, Noble J, Streulens Y, Elie R, et al. Pathophysiology of chest pain in patients with cardiomyopathies and normal coronary arteries. *Circulation* 1982;65:778-89.
8. Elliott PM, Koski JC, Prasad K et al. Chest pain during daily life in patients with hypertrophic cardiomyopathy: an ambulatory electrocardiographic study. *Eur Heart J* 1996;17:1056-64.
9. Maron BJ. Hypertrophic cardiomyopathy. *Curr Probl Cardiol* 1993;18:639-704.

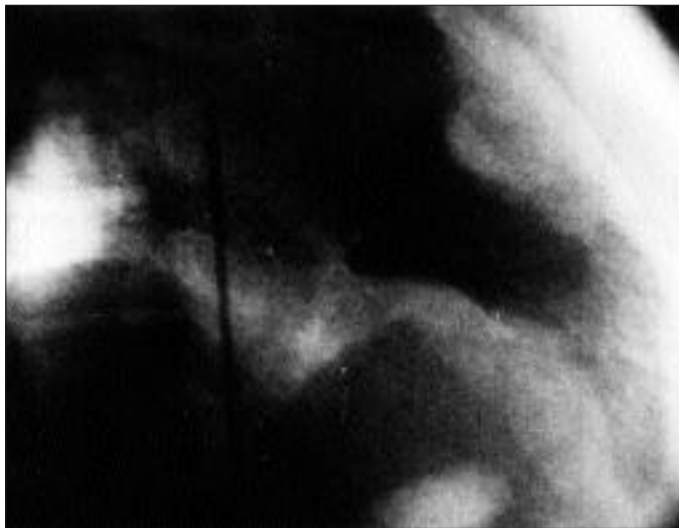


Figure 1. The left ventriculography demonstrates the view taken in the left oblique projection, which is consistent with hypertrophic cardiomyopathy

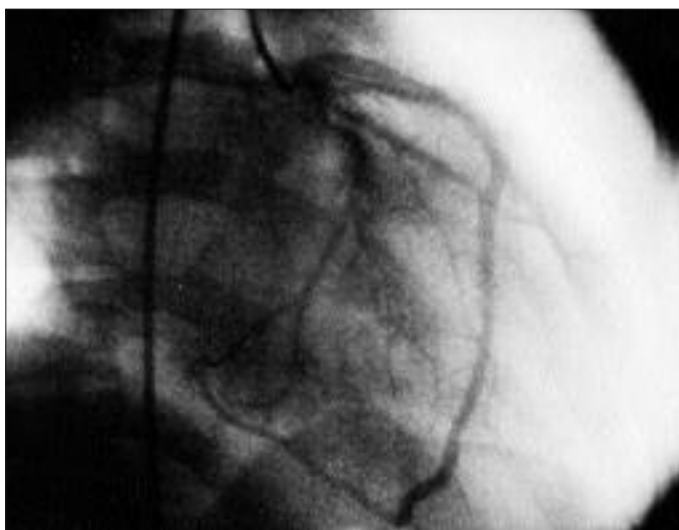


Figure 2. Coronary angiography reveals the single coronary artery in which the right coronary artery originates as a terminal extension of the left anterior descending artery and travels in the posterior interventricular sulcus retrogradely

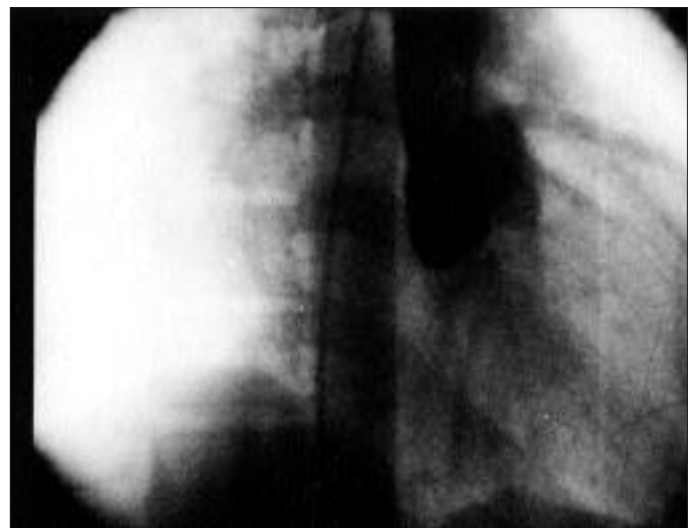


Figure 3. Aortography shows one coronary ostium in the left sinus of Valsalva and no ostium in the right sinus of Valsalva