Anomalous single coronary artery presenting as typical angina pectoris: a case report

Tipik angina pektorise neden olan tek koroner arter anomalisi: Olgu sunumu

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Single coronary artery is described as an isolated coronary artery originating from the aortic root through a single ostium in the absence of another ostium, where isolated coronary is the only source of blood supply to the whole heart. We present a 53-year-old woman whose coronary angiography for typical chest pain revealed an isolated single coronary artery. On coronary angiography, the whole coronary system originated by a single trunk from the right sinus of Valsalva. Multislice computed tomography showed that the left anterior descending, circumflex, and right coronary arteries arose from a single ostium in the right sinus of Valsalva without a left main trunk. Despite maximal medical therapy, her chest pain persisted. The patient did not accept surgical treatment proposed for correction of the anomaly.

Key words: Chest pain/etiology; coronary angiography; coronary vessel anomalies; sinus of Valsalva/abnormalities.

The incidence of coronary artery anomalies which are most frequently found incidentally during routine coronary angiography is 0.6-1.3%. [1-3] Single coronary artery is described as an isolated coronary artery originating from the aortic root through a single ostium in the absence of another ostium. The coronary from single ostium is the source of blood supply to the whole heart. [4] The incidence of a anomalous single coronary artery in the community was found to be 0.02%. [5,6] It is usually asymptomatic with a benign course. However, certain types of single coronary arteries may lead to severe cardiac events such as sudden death and myocardial infarction, particularly during exercise. [7] Isolated single coronary artery may also present with a clinical presentation involving chest pain, rhythm abnormalities, syncope, or congestive heart failure. [8,9] We present a patient who presented typical chest pain and who was diagnosed with anomalous coronary artery originaTek koroner arter, aort kökünden tek koroner ostiyum waracılığıyla çıkan ve ikinci bir ostiyumun gözlenmediği izole koroner arter olarak tanımlanır. Tek koroner arter bütün kalbin kan akımının sağlanmasından sorumludur. Bu yazıda tipik göğüs ağrısı nedeniyle koroner anjiyografi yapılan ve tek koroner arter saptanan 53 yaşında kadın hasta sunuldu. Koroner anjiyografide tüm koroner sistemin sağ sinüs Valsalva'dan çıkan tek gövdeden kalp bölgesine yayılım gösterdiği saptandı. Çokkesitli bilgisayarlı tomografide, sol ön inen, sirkumfleks ve sağ koroner arterlerin sol ana gövde olmaksızın tek ostiyum aracılığı ile sağ sinüs Valsalva düzeyinde tek gövdeden köken aldıkları izlendi. Takiplerinde yoğun medikal tedaviye rağmen göğüs ağrısının devam etmesi nedeniyle hastaya cerrahi tedavi önerildi; ancak, hasta cerrahi tedaviyi kabul etmedi.

Anahtar sözcükler: Göğüs ağrısı/etyoloji; koroner anjiyografi; koroner damar anomalisi; Valsalva sinüsü/anormallik

ting from a single trunk in the right sinus of Valsalva.

CASE REPORT

A 53-year old woman presented with chest pain at our outpatient clinic. Her past medical history revealed that she had been having complaints of malaise since her youth age and had been getting tired earlier than her age mates. The patient had been complaining of chest pain for the past two years, located around the middle of the chest and characterized by radiation to the left, increasing on exertion and resolving with a-10-minute rest period. In addition, the pain was reported to have continuously worsened within the past one year. The patient who underwent a stress myocardial perfusion scintigraphy and coronary angiography at another center also complained about dyspnea and palpitation, in addition to the chest pain. The results of stress myocardial perfu-

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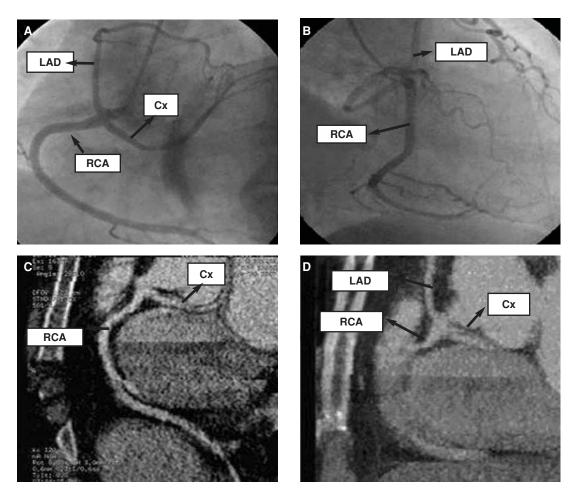


Figure 1. (A) Angiographic image of the origin of the whole coronary system from a single truck in the right sinus of Valsalva extending to the heart region. **(B)** Multislice computed tomography showing the whole coronary system arising by a single ostium in the right sinus of Valsalva from a single trunk without the left main trunk. LAD: Left anterior descending artery; Cx: Circumflex artery; RCA: Right coronary artery.

sion scintigraphy were interpreted as a suboptimal test within normal range due to the inability to reach the target heart rate. On the other hand, the patient was scheduled for medical therapy following diagnosis of a single coronary artery anomaly on coronary angiography. The patient was hypertensive with a family history as a risk factor for coronary artery disease. She had been taking aspirin 100 mg, cilazapril 5 mg, carvedilol 25 mg and amlodipine 10 mg for the past two years. Physical examination revealed a blood pressure of 140/90 mmHg and heart rate of 70 bpm which was regular. No abnormal findings were observed during the physical examination. Electrocardiography revealed a nonspecific negative T-wave in the inferior leads. The chest X-ray was normal. No pathological finding was reported from the two-dimensional echocardiography. Reevaluation of the coronary angiography performed at the external center revealed the whole coronary system to have originated by a single trunk from the right sinus of Valsalva (Figure 1a).

A multislice computed tomography was performed to monitor the course of the coronary artery, and the left anterior descending (LAD), the circumflex, and the right coronary artery were observed to originate from a single trunk through a single ostium in the right sinus of Valsalva without the right main trunk (Figure 1b). The right coronary artery was observed to follow its normal course, with an open lumen and normal dimensions. The LAD artery was also observed to meet its normal course by turning left immediately from the anterior border of the pulmonary trunk, after taking an anterior and an upward course form the trunk. On the other hand, the circumflex artery took its normal course at the posterior part of the trunk extending to the left between the aortic root and the left atrium. Adjuvant treatment with nitrate and trimetazidine was initiated. The patient was considered for surgery due to persistent chest pain despite intensive medical treatment during subsequent follow-up periods. However, the pati-

Table 1. Angiographic classification of single coronary artery[10]

	Code	Description
Ostial location	Right (R)	Right sinus of Valsalva
	Left (L)	Left sinus of Valsalva
Anatomical distribution	I	Single coronary artery with normal right or left coursing (RC or LC)
	II	After leaving the right or left sinus the single coronary artery crosses at the base of the heart as a large transverse trunk in order to supply the contralateral coronary artery.
	III	Single coronary artery arising from the right sinus, with the LAD and the circumflex arteries arising from separate coronary artery trunks instead of a single trunk just immediately at the exit.
Course of the transverse branch	Α	Anterior to the large vessels (anterior to the right ventricle)
	В	Between the aorta and pulmonary artery
	Р	Posterior to the large vessels
	S	Septal type (above to the interventricular septum)
	С	Combined type

ent refused surgery and is still being followed symptomatically.

DISCUSSION

Many congenital coronary artery anomalies are asymptomatic and are only incidentally found during routine angiography. [1] The most commonly seen coronary artery anomalies include the LAD and circumflex arteries arising from separate ostia in the left sinus of Valsalva, left circumflex arteries arising from the right sinus of Valsalva or the right coronary artery, and coronary artery fistulae (CAF). Isolated single coronary artery anomaly is one of the most rarely seen coronary anomalies and constitutes 2-4% of all coronary artery anomalies. [5-10] Single coronary artery anomalies are classified according to the site of origin from the left and right coronary arteries, anatomical distribution on the ventricular surface, and according to its relationship with the ascending aorta and the pulmonary artery (Table 1).[10] Our case was found to be consistent with RIIIA. Coursing of the coronary artery between the aorta and the pulmonary artery in this anomaly involves a very high risk of sudden death. On the other hand, very severe adverse conditions may be seen even in the absence of coronary artery coursing between the aorta and the pulmonary artery, since it may lead to cardiac ischemia, congestive heart failure, ventricular fibrillation, or sudden death. [10] Despite the relatively low incidence of atherosclerotic heart diseases in young patients with coronary anomalies compared to their elder counterparts, exertional or spontaneous sudden death is more common. [9] The prognosis in patients with single coronary varies according to the anatomic distribution. While the prognosis may be very good in some patients, sudden death may be observed in others, whereas about 15% of patients become asymptomatic from severe heart diseases before the age of 40. [4,11] The mortality rate of patients with left coronary artery originating from the right coronary sinus before the age of 20 years is very high (59%). Death normally occurs after tiring physical activities since the dilated vessel is compressed between the aorta and the pulmonary artery during exercise. [4,12] There is a high risk of mortality unless surgical treatment is administered under conditions where the artery courses between the right ventricular outflow and the aorta.[11] The patient's age and type of anomaly are very important for the determination of treatment in asymptomatic patients. There is a need for surgical treatment in symptomatic patients diagnosed at a younger age, where the course of the artery is found to be very dangerous. [13] Taylor et al. [9] reported the risk of sudden death to be very high (82%) in patients with single coronary artery, where there is interarterial coursing of the left main coronary artery. In one case report by Raynard et al.[14] and two case reports by Ono et al. [15] interarterial coursing of the LAD was presented; left internal mammary artery-LAD bypass was reported to be very effective in these patients. Surgical approach involves reimplantation of the anomalous artery into the coronary sinus or an appropriate bypass. However, coronary bypass in these patients is considered to be a more effective and safer procedure.[16] On the other hand, there is currently no guidelines or consensus regarding the management of patients with single coronary artery which are not associated with atherosclerosis and the other congenital heart diseases. [17] Our patient had typical chest pain, and the decision to administered surgery was made since there was no regression despite intensive medical treatment. However, the patient refused surgical treatment.

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In conclusion, chest pain may be observed in patients with single coronary artery anomaly in the absence of atherosclerosis. Further case series and prospective studies are required to establish the treatment method in these patients.

REFERENCES

- 1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 1990;21:28-40.
- 2. Kardos A, Babai L, Rudas L, Gaál T, Horváth T, Tálosi L, et al. Epidemiology of congenital coronary artery anomalies: a coronary arteriography study on a central European population. Cathet Cardiovasc Diagn 1997; 42:270-5.
- 3. Gowda RM, Chamakura SR, Dogan OM, Sacchi TJ, Khan IA. Origin of left main and right coronary arteries from right aortic sinus of Valsalva. Int J Cardiol 2003; 92:305-6.
- 4. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. Circulation 2002;105:2449-54.
- 5. Desmet W, Vanhaecke J, Vrolix M, Van de Werf F, Piessens J, Willems J, et al. Isolated single coronary artery: a review of 50,000 consecutive coronary angiographies. Eur Heart J 1992;13:1637-40.
- von Kodolitsch Y, Franzen O, Lund GK, Koschyk DH, Ito WD, Meinertz T. Coronary artery anomalies Part II: recent insights from clinical investigations. Z Kardiol 2005;94:1-13.
- Frescura C, Basso C, Thiene G, Corrado D, Pennelli T, Angelini A, et al. Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease. Hum Pathol 1998; 29:689-95.
- Click RL, Holmes DR Jr, Vlietstra RE, Kosinski AS, Kronmal RA. Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival-a re-

- port from the Coronary Artery Surgery Study. J Am Coll Cardiol 1989;13:531-7.
- 9. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol 1992;20:640-7.
- 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.
- 11. Popma JJ. Coronary angiography and intravascular imaging. In: Zipes DP, Libby P, Bonow RO, Braunwald E, editors. Heart disease: a textbook of cardiovascular medicine. 7th ed. Philadelphia: Elsevier Saunders; 2005. p. 423-55.
- 12. Basso C, Corrado D, Thiene G. Congenital coronary artery anomalies as an important cause of sudden death in the young. Cardiol Rev 2001;9:312-7.
- 13. Doğan SM, Gürsürer M, Aydın M, Göçer H, Çabuk M, Dursun A. Myocardial ischemia caused by a coronary anomaly left anterior descending coronary artery arising from right sinus of Valsalva. Int J Cardiol 2006; 112:e57-9.
- Roynard JL, Cattan S, Artigou JY, Desoutter P.Anomalous course of the left anterior descending coronary artery between the aorta and pulmonary trunk: a rare cause of myocardial ischaemia at rest. Br Heart J 1994;72:397-9.
- 15. Ono M, Brown DA, Wolf RK. Two cases of anomalous origin of LAD from right coronary artery requiring coronary artery bypass. Cardiovasc Surg 2003;11:90-2.
- 16. Koşar F, Ermiş N, Erdil N, Battaloğlu B. Anomalous LAD and CX artery arising separately from the proximal right coronary artery-a case report of single coronary artery with coronary artery disease. J Card Surg 2006;21:309-12.
- 17. Braun MU, Stolte D, Rauwolf T, Strasser RH. Single coronary artery with anomalous origin from the right sinus Valsalva. Clin Res Cardiol 2006;95:119-21.