

Severe Form of Persistent Thebesian Veins Mimicking Left Ventriculography Presenting with Myocardial Ischemia

Miyokardiyal İskemi Oluşturan ve Sol Ventrikülografiyi Taklit Eden Kalıcı Tebesian Venlerin Ciddi Formu

 Mustafa Serkan Karakas¹,  Veysel Tosun¹,  Necmettin Korucuk¹,  Unal Guntekin¹,  Remzi Yılmaz¹

¹Akdeniz University Medical Faculty, Department of Cardiology. Antalya, Turkey.

Abstract

Coronary artery fistula is a rare congenital anomaly. Thebesian veins are multiple coronary artery microfistula that comunicates coronary arteries and ventricles. Most patients with this anomaly are asymptomatic, but some patients may be symptomatic and have a presentation of myocardial ischemia, thrombosis and embolism, heart failure and arrhythmias. We present a case with exertional dyspnea and angina whose myocardial perfusion scintigraphy demonstrated inferior and apical defect with complete reversibility. Coronary angiography showed no coronary stenosis, but capillary blush draining into the left ventricular cavity through multiple microfistulae from the left anterior descending artery, circumflex artery and right coronary artery mimicking left ventriculography. Because of these small multiple fistulas are managed medically, the patient was discharged on a beta blocker, aspirin and ranolazine therapy. The patient stated a reduction in his symptoms one month later.

Keywords: Thebesian veins, coronary artery fistula, myocardial ischemia

Özet

Koroner arter fistülü nadir bir konjenital anomalidir. Tebesian venler, ventrikülle koroner arterler arasında bağlantı oluşturan çoklu koroner arter mikrofistül-leridir. Hastaların çoğu asemptomatik olmakla birlikte bazı hastalar semptomatik olabilmekte ve miyokardiyal iskemi, tromboz ve emboli, kalp yetmezliği ve aritmilerle gelebilmektedir. Yazımızda efor dispnesi ve anginası bulunan, miyokard perfüzyon sintigrafisinde geri dönüşlü inferior ve apikal defekt saptanan bir vaka sunulmuştur. Hastanın koroner anjiyografisinde koroner darlık saptanmadı ancak sol ön inen arter, sirkümfleks arter ve sağ koroner arterden çoklu mikrofis-tüller aracılığıyla sol ventriküler kaviteye boşalan ve sol ventrikülografi görüntüsü oluşturan kapiller boyanma izlendi. Küçük mikrofistüllerden dolayı hastaya medikal tedavi verildi. Hasta beta bloker, aspirin ve ranolazin tedavisi ile taburcu edildi. Bir ay sonraki kontrolde hasta yakınmalarında azalma olduğunu belirtti.

Anahtar Kelimeler: Tebesian venler, koroner arter fistülü, miyokardiyal iskemi

Introduction

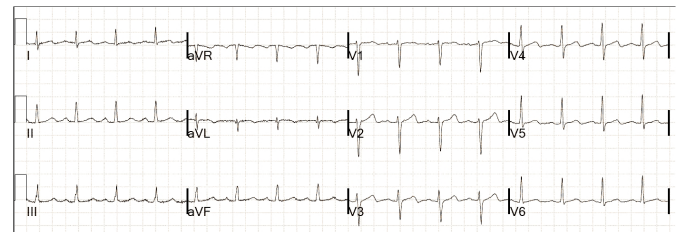
Coronary artery fistula (CAF) is a rare congenital anomaly and its incidence is 0.08% to 0.3% in the adult population referred for cardiac catheterization (1). There is a connection between one or more of the coronary arteries and a cardiac chamber or great vessels. Thebesian veins or venae cordis minimae are multiple coronary artery microfistula that comunicates coronary arteries and ventricles.

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, endocarditis, endarteritis and arrhythmias (2,3). In the literature, coronary artery fistula presenting ischemic heart disease is rare (4). Here, we reported a case of persistant Thebesian veins in a patient presenting angina and exertional dyspnea.

Case

A 70-year-old male admitted to cardiology department with exertional dyspnea and chest pain. He was taking proton pump inhibitor as medical therapy. His physical examination was normal. Electrocardiography showed sinus rhythm and there was biphasic T waves in V3-V4-V5-V6 derivations (Figure 1).

Figure 1: Biphasic T waves in V3, V4, V5, V6 derivations on the electrocardiography.



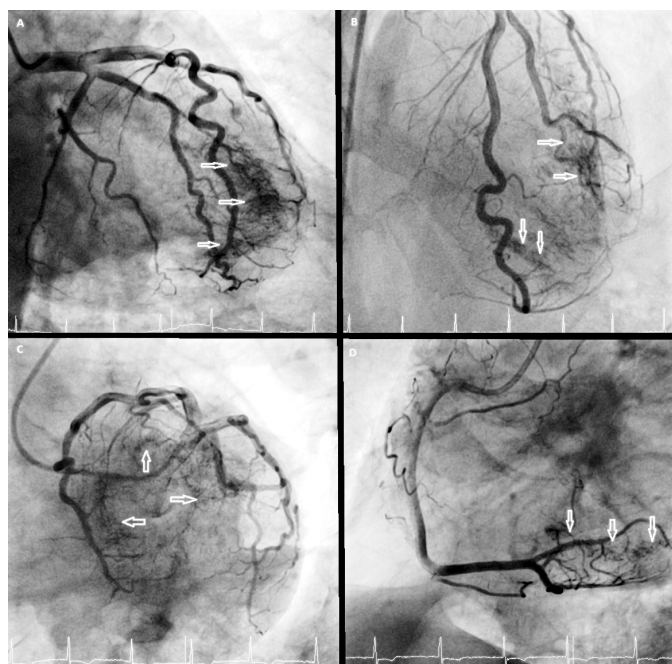
Corresponding Author: Mustafa Serkan Karakas, Akdeniz University Medical Faculty, Department of Cardiology. Dumlupınar Boulevard, Antalya, Turkey.

E-Mail: mserkan19@hotmail.com

Alınma Tarihi: 10.11.2015 / **Kabul Tarihi:** 01.10.2019 / **Yayınlanma Tarihi:** 15.06.2021

Echocardiography demonstrated minimal concentric left ventricle (LV) hypertrophy, normal LV chamber size and normal LV systolic function without the regional wall motion abnormality. We planned myocardial perfusion scintigraphy for ischemia and it was reported as ischemia in inferior and apical segments. Coronary angiography demonstrated epicardial coronary plaques in coronary arteries, but appearance of capillary blush draining into the left ventricular cavity, during both left main and right coronary artery injection and mimicking left ventriculography (Figure 2). We concluded that this appearance suggests prominent thebesian veins communicating between the coronary arteries and the ventricle. Because of small multiple fistulas are managed medically, he was discharged on a beta blocker, aspirin and ranolazine therapy. He stated a reduction in his symptoms one month later.

Figure 2: Multiple microfistula draining into the left ventricle from the LAD (A), LAD and Cx (B,C) and RCA (D) mimicking ventriculography with contrast hand injection.



Discussion

Coronary artery fistula (CAF) is a rare congenital anomaly. There is an abnormal connection that directly links one or more coronary arteries to a heart chamber or to major thoracic vessels (5). CAF occurs equally in men and women and have a 0.002% incidence among general population, 0.08% to 0.3% in the adult population referred for cardiac catheterisation (1,6,7).

The natural history of CAF is variable. Ata et al. showed that approximately half of patients with coronary artery fistulas are asymptomatic (8). Clinical presentation of CAF varies widely. It is dependent on the size of the fistula, age of patient and presence of myocardial ischemia (9). Chest pain, exertional dyspnea and fatigue may develop. The mechanism of symptoms appeared to be coronary steal phenomenon and diastolic overload (10). Steal phenomenon suggests that there is a decrease in myocardial perfusion on exertion due to the inability of coronary flow reserve. The most common complication is myocardial ischemia due to coronary steal phenomenon which occurs in 15% of patients with CAF (11). In our case, myocardial ischemia induced by a coronary steal phenomenon and decrease in intracoronary diastolic pressure especially during exercise may cause his angina. Congestive heart failure and arrhythmia could be caused by excessive loading of cardiac

chambers. Intravascular thrombosis, infective endocarditis, and rarely hemopericardium due to rupture of aneurysmal coronary artery might be other complications (12).

In CAF patients drainage most frequently occurs into the right ventricle because of drainage is generally carried out to low pressure chambers (13). Fistulas terminating in the left ventricle are rare (12). In our case, all three coronary arteries had persistent thebesian veins and communicating to the left ventricle, mimicking ventriculography.

Conflicting evidence from case reports exists about the benefits of medical therapy for these patients. As a general practise, prophylactic low dose aspirin is suggested. For treatment of angina beta blockers and calcium channel blockers may be prescribed. Some case reports suggest that ranolazine reduces angina episodes (14). Additionally, it should be remembered nitrate therapy may exacerbate ischemia by increasing leakage to the left ventricle in these patients.

Conflict of Interest: Authors state no conflict of interest

References

1. Stierle U, Giannitsis E, Sheikhzadeh A, Portraz J et al. Myocardial ischemia in generalized coronary artery-left ventricular microfistulae. *Int J Cardiol* 1998;63:47-52.
2. Alkhulaifi AM, Horner SM, Pugsley WB, Swanton RH et al. Coronary artery fistulas presenting with bacterial endocarditis. *Ann Thorac Surg* 1995; 60:202-4.
3. Skimming JW, Walls JT. Congenital coronary artery fistula suggesting a steal phenomenon in a neonate. *Pediatr Cardiol* 1993;14:174-5.
4. Chattopadhyay S, Wahab N, O'Reilly K. Persistent thebesian veins presenting with myocardial ischemia. *Can J Cardiol* 2010;26:e31-2.
5. Ashraf SS, Shaukat N, Fisher M, Clarke B. Bicornary-pulmonary fistulae with coexistent mitral valve prolapse: A case report and literature review of coronary-pulmonary fistula. *Eur Heart J* 1994;15:571-4.
6. Vlodaver Z, Neufeld H, Edwards J. Coronary arterial variations in the normal heart and in congenital heart disease. London: Academic Press, Inc.; 1975 43-78.
7. Luo L, Kebede S, Wu S, Stouffer GA. Coronary artery fistulae. *Am J Med Sci* 2006;332:79-84.
8. Ata Y, Turk T, Bicer M, Yalcin M, et al. Coronary arteriovenous fistulas in the adults: natural history and management strategies. *J Cardiothorac Surg* 2009;4:62
9. Mechleb W, Abboud L, Mattar C, Haddadin T. Case report and review of literature: coronary artery fistulae. *Tenn Med* 2013;106:39-41
10. Stierle U, Giannitsis E, Sheikhzadeh A, Potratz J. Myocardial ischemia in generalized coronary artery-left ventricular microfistulae. *Int J Cardiol* 1998;63:47-52.
11. Valente AM, Lock JE, Gauvreau K, et al. Predictors of long-term adverse outcomes in patients with congenital coronary artery fistulae. *Circ Cardiovasc Interv* 2010;3:134-9.
12. Blake HA, Manion WC, Mattingly TW, Baroldi G. Coronary artery anomalies. *Circulation* 1964;30:927-40.
13. Abelin AP, Sarmiento-Leite R, Quadros AS, Gottschall CAM. Fistula coronaria. *Rev Bras Cardiol Invas.* 2008;16:242-243.
14. Khoueiry G, Baydoun H, Abi Rafeh N, McCord D, et al. Persistent thebesian vessels involving the right and left ventricles leading to coronary steal phenomena and ischemia. *Congenit Heart Dis.* 2014;9(2):E61-5.