Left anterior descending coronary artery originating from the pulmonary artery: a rarity suspected during echocardiography

Ekokardiyografi sırasında şüphelenilen nadir bir anomali: Pulmoner arterden köken alan sol ön inen arter

Necla Özer, M.D., Ali Deniz, M.D., Rıza Doğan, M.D.[1]

Departments of Cardiology and [1]Cardiovascular Surgery, Medicine Faculty of Hacettepe University, Ankara

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly that is often referred to as Bland-White-Garland syndrome. Isolated anomalous origin of the left anterior descending (LAD) or circumflex arteries is even rarer. A 38-year-old woman presented with typical angina of about two-year history. Physical examination findings were normal other than a grade 3/6 systolic murmur. Electrocardiography showed anterolateral ST-segment depression and T wave inversion, indicating anterior ischemia. Color flow transthoracic Doppler echocardiography showed dilated coronary arteries. Parasternal short axis views demonstrated an abnormal flow originating from the common pulmonary artery. Upon suspicion of a coronary anomaly, coronary angiography was performed. Both the left circumflex (Cx) and right coronary arteries (RCA) were found dilated, giving extensive collaterals to the LAD artery, which drained into the main pulmonary artery. Computed tomographic angiography confirmed that the LAD artery originated from the main pulmonary artery. Surgical correction was performed and the LAD artery was re-anastomosed to the aorta. Control coronary angiography performed one week after surgery showed patent LAD artery and diminished collateral supply from the RCA and Cx arteries.

Key words: Coronary vessel anomalies/diagnosis/surgery; echocardiography; pulmonary artery/abnormalities.

Bland-White-Garland sendromu adı verilen ve sol koroner sistemin pulmoner arterden köken aldığı koroner arter anomalisi oldukça nadir bir durumdur. Sadece sol ön inen (LAD) arterin veya sirkumfleks arterin pulmoner arterden çıkması ise daha da nadirdir. Otuz sekiz yaşında kadın hasta, iki yıllık öyküsü olan tipik angina ile başvurdu. Fizik muayene bulguları, 3/6 dereceli sistolik üfürüm dışında normaldi. Elektrokardiyografide, anterior iskemiye işaret eden anterolateral ST-segment çökmesi ve T dalga inversiyonu izlendi. Renkli akım transtorasik Doppler ekokardiyografide koroner arterler genişlemiş bulundu. Parasternal kısa eksen görüntülerde ana pulmoner arterden anormal bir akım dikkat çekmekteydi. Koroner anomali varlığından şüphelenilmesi üzerine yapılan koroner anjiyografide, sol sirkumfleks ve sağ koroner arterlerin genişlemiş olduğu ve bunlarla LAD arter arasında yaygın kollateraller oluştuğu gözlendi; ayrıca, LAD arteri ana pulmoner artere boşalmaktaydı. Bilgisayarlı tomografik anjiyografi ile LAD arterin ana pulmoner arterden köken aldığı doğrulandı. Yapılan cerrahi düzeltme ameliyatında LAD arterin aort ile anastomozu sağlandı. Ameliyattan bir hafta sonraki koroner anjiyografide, LAD arterin açık olduğu ve sol sirkumfleks ve sağ koroner arterlerden kollateral desteğinin zayıfladığı görüldü.

Anahtar sözcükler: Koroner damar anomalisi/tanı/cerrahi; ekokardiyografi; pulmoner arter/anormallik.

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly that is often referred to as Bland-White-Garland (BWG) syndrome. Isolated anomalous origin of the left anterior descending (LAD) coronary artery or circumflex artery is even rarer. In this case report, we described an anomalous LAD artery originating from the pulmonary artery, which was first suspected from echocardiographic views.

CASE REPORT

A 38-year-old woman was admitted to our hospital with typical angina of about two-year history. On physical examination, a grade 3/6 systolic murmur was

Received: September 18, 2007 *Accepted:* December 24, 2007

Correspondence: Dr. Necla Özer. Hacettepe Üniversitesi Tıp Fakültesi, Kardiyoloji Anabilim Dalı, 06100 Ankara, Turkey.
Tel: +90 312 - 305 17 80 Fax: +90 312 - 311 40 58 e-mail: neclaozer@hotmail.com

heard at the cardiac apex. Other physical examination findings were normal. Electrocardiography revealed anterolateral ST-segment depression and T wave inversion indicating anterior ischemia. Dilated coronary arteries were detected in color flow images of transthoracic Doppler echocardiography. Parasternal short axis views demonstrated an abnormal flow originating from the common pulmonary artery (Fig. 1a). Upon suspicion of a coronary anomaly, coronary angiography was performed. Both the left circumflex (Cx) and right coronary arteries (RCA) were found dilated, giving extensive collaterals to the LAD artery, which drained into the main pulmonary artery (Fig. 1b). Coronary computed tomographic (CT) angiography confirmed that the LAD artery originated from the main pulmonary artery (Fig. 2). Surgical correction was planned. During surgery, the LAD artery was re-anastomosed to the aorta without tension. Control coronary angiography performed one week after surgery showed patent LAD artery and diminished collateral supply from the RCA and Cx arteries.

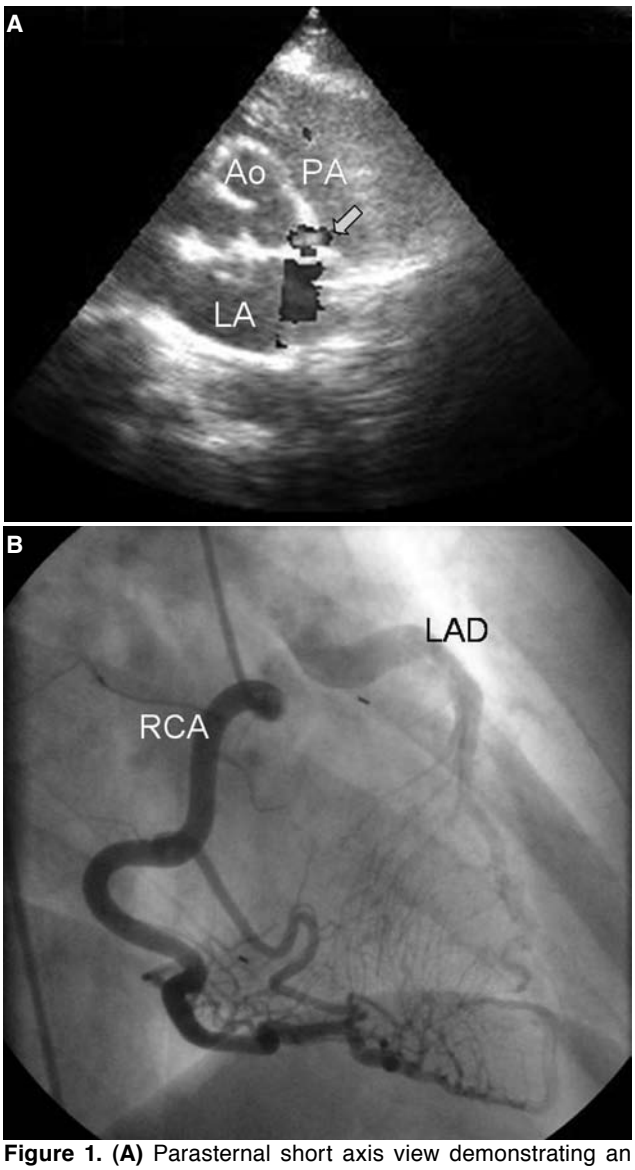

Figure 1. (A) Parasternal short axis view demonstrating an abnormal flow originating from the pulmonary artery (arrow). PA: Pulmonary artery, Ao: Aorta, LA: Left atrium. **(B)** Coronary angiography showing ectatic right coronary artery (RCA) giving collateral vessels to the left anterior descending (LAD) artery.

DISCUSSION

The estimated incidence of BWG syndrome is 1/300,000 live births, accounting for 0.24% to 0.46% of all congenital cardiac anomalies.[1] In the neonatal period, the pulmonary blood pressure diminishes, the ductus arteriosus closes, and the flow in the left coronary artery reverses. The development of collateral circulation between the right and left coronary arteries determines the extent of myocardial ischemia in BWG syndrome; thus, patients with well-established collaterals are classified as the "adult type", and those with no collaterals as the "infantile type".[2]

It is extremely rare that the LAD artery originates from the pulmonary artery.[3] In this type of coronary artery anomaly, the LAD territory is at ischemic risk due to low perfusion pressure, though this risk is less than that seen in ALCAPA. The Cx and RCA arteries are usually ectatic and give collaterals to the LAD artery. In our case, the RCA and Cx arteries gave

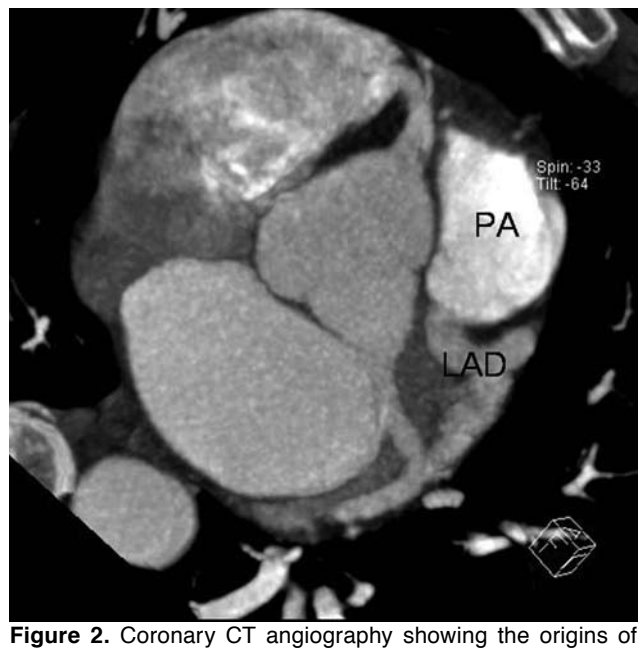

Figure 2. Coronary CT angiography showing the origins of the three coronary arteries. The circumflex and right coronary arteries originate from the left and right sinuses of Valsalva respectively, whereas the left anterior descending (LAD) artery originates from the pulmonary artery (PA).

extensive collaterals to the LAD artery, representing the "adult type" of this anomaly. The symptoms and signs of this anomaly usually include atypical angina and systolic murmur. Pulmonary hypertension may develop in ALCAPA due to left-to-right shunting, left ventricular dysfunction, and mitral regurgitation which is thought to be caused by ischemic papillary muscle dysfunction. In our case, systolic left ventricular dysfunction, significant mitral regurgitation, or pulmonary hypertension were not present. The absence of these features makes anomalous LAD artery from the pulmonary artery a milder form, though this does not exclude the necessity of treatment.

Echocardiographic diagnosis is rare for coronary artery anomalies. In our case, we suspected a coronary anomaly from transthoracic echocardiographic views. Sometimes a mass may also be seen in the right atrium due to a huge right coronary artery aneurysm.[4] Mostly CT coronary angiography is a valuable noninvasive tool to show anomalous coronary arteries.

Surgery is recommended in patients with BWG syndrome, even in the absence of symptoms or a significant left-to right shunt syndrome, due to the risk for ventricular arrhythmias and sudden death. [5] Surgical treatment of ALCAPA has much evolved over the past decades, making several methods a historical interest, such as aortopulmonary anastomosis or banding of the pulmonary artery. The goals of modern surgical management are establishment of a two-coronary system with long-term patency, use of native tissue, and maintenance of potential for normal growth of coronary ostia and arteries. The two-coronary artery system can be established via several techniques, including ligation of the origin of anomalous left coronary artery combined with the establishment of a conduit to bypass the left coronary artery, creation of an aortopulmonary window with an intrapulmonary baffle (Takeuchi operation), and direct reimplantation of the anomalous left coronary artery to the ascending aorta. Isolated LAD anomalies require surgical treatment because of the same risks, though they present less risk due to smaller ischemic region. Methods such as direct transfer (without tension), tubular reconstruction, and in situ transfer can be used in all patients regardless of the site of the anomalous coronary orifice. Whenever possible, direct implantation of the LAD artery to the ascending aorta is a surgical option unless tension develops. Another option is the ligation of the LAD artery ostium and performing aortocoronary bypass graft operation.

Our case may receive attention because this type of LAD artery is extremely rare, and it gave rise to symptoms during adult life, even after two successful pregnancies. Surgical correction is necessary to decrease anterior ischemia.

REFERENCES

1. Malec E, Zajac A, Mikuta M. Surgical repair of anomalous origin of the coronary artery from the pulmonary artery in children. Cardiovasc Surg 2001;9:292-8.
2. BarbetakisN,EfstathiouA,EfstathiouN,Papagiannopoulou P, Soulountsi V, Fessatidis I. A long-term survivor of Bland-White-Garland syndrome with systemic collateral supply: a case report and review of the literature. BMC Surg 2005;5:23.
3. Derrick MJ, Moreno-Cabral RJ. Anomalous origin of the left anterior descending artery from the pulmonary artery. J Card Surg 1991;6:24-8.
4. Arsan S, Naseri E, Keser N. An adult case of Bland White Garland syndrome with huge right coronary aneurysm. Ann Thorac Surg 1999;68:1832-3.
5. Alexi-Meskishvili V, Berger F, Weng Y, Lange PE, Hetzer R. Anomalous origin of the left coronary artery from the pulmonary artery in adults. J Card Surg 1995; 10(4 Pt 1):309-15.