

Apikal hipertrofik kardiyomiyopati ile bir hastada koroner arter ile pulmoner arter arasındaki fistülün perkütan yolla kapatılması

Percutaneous closure of the coronary artery-pulmonary artery fistula in a patient with apical hypertrophic cardiomyopathy

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Özet– Elli dört yaşında kadın hasta, son 6 aydır olan eforla gelişen göğüs ağrısı yakınmasıyla başvurdu. Hastanın transtorasik ekokardiyografisinde apikal bölgede hipertrofi görüldü. Ayırıcı tanı amacıyla yapılan kardiyak manyetik rezonans görüntülemesinde apikal hipertrofik kardiyomiyopati saptandı. Göğüs ağrısı nedeniyle yapılan miyokart perfüzyon sintigrafisinde anterior bölgede iskemi bulundu. Koroner anjiyografide koroner arterler normal olup sol ön inen arter proksimalinden pulmoner artere doğru uzanan arteriovenöz fistül (AVF) gösterildi. Mevcut yakınmalarının AVF'ye bağlı koroner çalma sendromu nedeniyle geliştiği düşünüldü. Perkütan koroner girişim ile “coil” kullanılarak AVF kapatıldı. Hastanın göğüs ağrısı düzeldi. Koroner AVF bulunan hastalarda koroner çalma sendromu miyokart iskemisinin nedeni olabilmektedir.

Summary– A fifty-four-year-old female patient was admitted to our unit with exertional chest pain of six months duration. Transthoracic echocardiography showed apical hypertrophy. Cardiac magnetic resonance imaging performed for differential diagnosis revealed apical hypertrophic cardiomyopathy. The patient underwent myocardial perfusion scintigraphy with the indication of chest pain which showed anterior ischemia. Coronary angiography revealed an arteriovenous fistula (AVF) extending from the proximal segment of the left anterior descending artery to the pulmonary artery. The patient's existing complaints were attributed to a coronary steal syndrome secondary to the coronary AVF. The AVF fistula was closed with a coil and the patient's chest pain improved. In conclusion, coronary steal syndrome may lead to myocardial ischemia in patients with a coronary AVF.

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Abbreviation

AVF Arteriovenous fistula

Coronary arteriovenous fistulas (AVFs) are rarely seen entities which frequently lead an asymptomatic course.^[1] In patients with coronary AVF, bacterial endocarditis, endarteritis, and embolism can develop, and spontaneous aneurysmatic rupture secondary to fistulas can be seen. In these patients, hypervolemia secondary to shunts results in onset of symptoms of heart failure including shortness of breath, and ortopnea. Besides, angina, arrhythmia, and myocardial infarction can develop because of ischemia occurring secondary to a decrease in coronary artery blood flow .^[2] Although repair of a fistula in asymptomatic patients is currently debatable, closure of a fistula is required because of potentially developed complications in symptomatic patients.^[3]

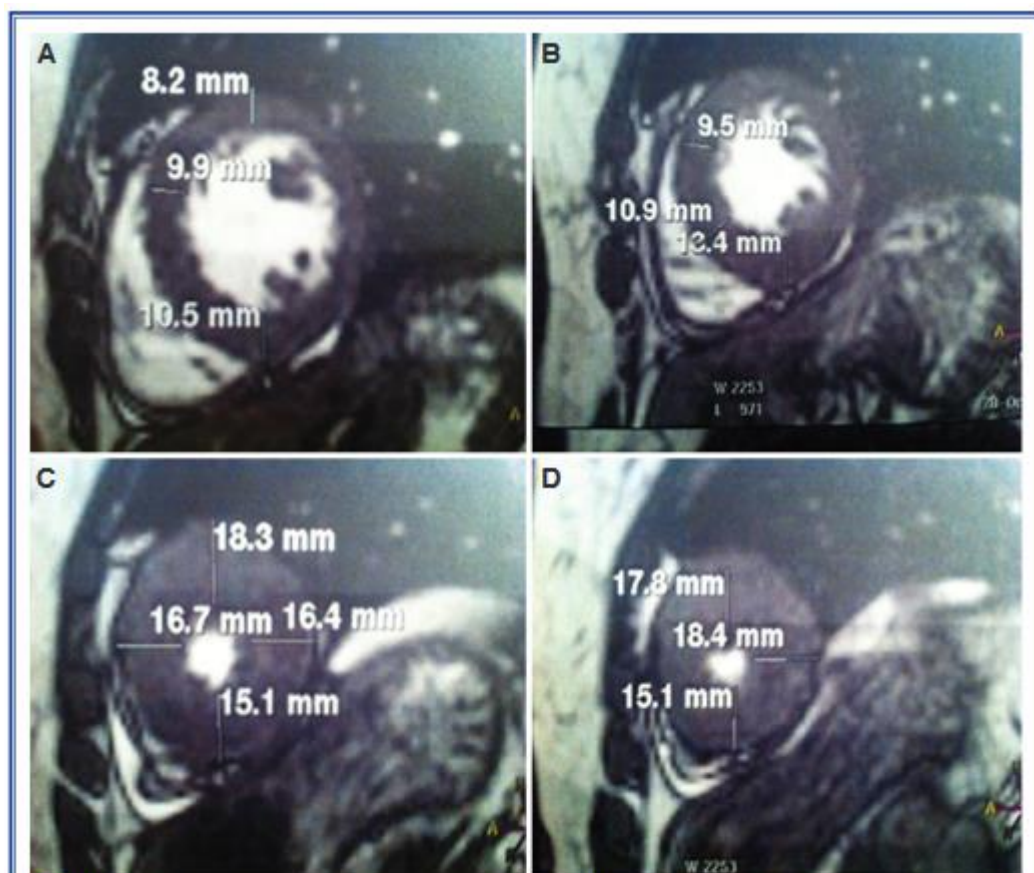


Figure 1. Cardiac magnetic resonance imaging: (A) basal left ventricular wall thickness (B) moderately increased left ventricular wall thickness and (C, D) apical left ventricular wall thickness

In the present article, an AVF extending from the proximal segment of the left anterior descending artery into pulmonary artery was detected on coronary angiograms of the patient who consulted to our clinic with complaints of

chest pain. The preexisting complaints of the patient who developed chest pain because of coronary steal syndrome, regressed after percutaneous closure of the fistula.

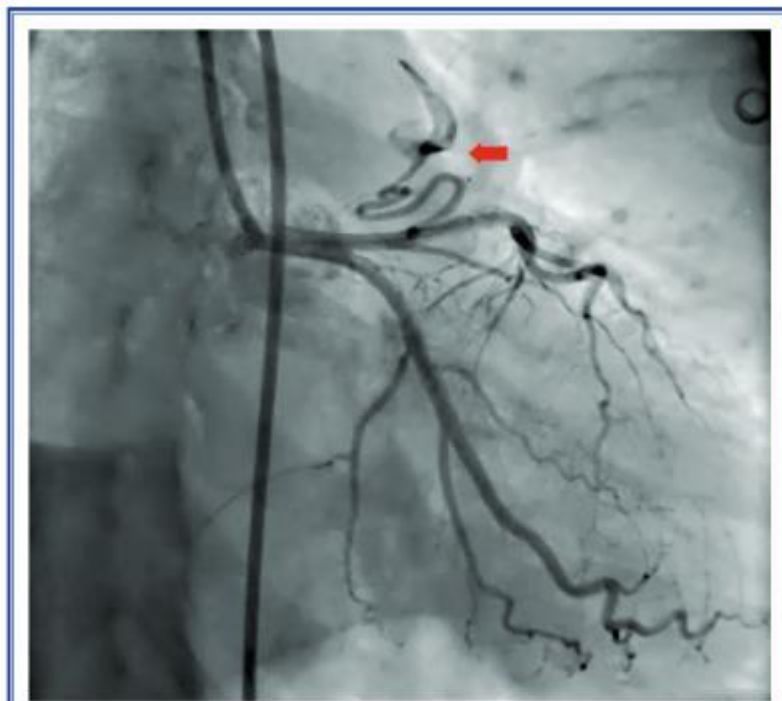


Figure 2. On coronary angiogram, a prominent arteriovenous fistula extending from the left anterior descending coronary artery into pulmonary artery is seen

CASE PRESENTATION

A 54-year-old female patient referred to our clinic with complaints of exertional chest pain occurring occasionally for the last 6 months. These complaints regressed gradually as assessed by auscultatory examinations. Her personal medical history revealed diagnoses of hypertension, and type 2 diabetes mellitus, and she was receiving oral ramipril (5 mg od) , and metformin

(850 mg bid) therapy. Her father, and mother had a history of coronary artery disease. Her physical examination findings were as follows: blood pressure 145/75 mmHg, regular pulse rate: 64/min. On cardiovascular system examination, uninterrupted grade 2/3 heart murmurs were heard over left 2., and 3. intercostal spaces. Electrocardiographic findings were as follows: heart rate, 76/min; normal sinus rhythm, and signs of left ventricular hypertrophy. Laboratory results were as

follows: glycemia, 120 mg/dl; LDL, 125 mg/dl; HDL, 44 mg/dl; triglyceride, 223 mg/dl, and hemoglobin, 15.8 g/dl. Transthoracic echocardiographic examinations revealed normal left ventricular systolic functions, and left ventricular apical hypertrophy(left ventricular wall thickness, 18 mm). For differential diagnosis, the patient was evaluated by cardiac magnetic resonance technique, and predominantly apical hypertrophic cardiomyopathy was detected (Figure 1a-d). The patient complaining of chest pain underwent firstly pharmacologic stress test using

dipyridamole, then myocardial perfusion scanning using a radioactive tracer, ie Tc-99M labeled MIBI. In stress images, moderate degrees of hypoperfusion seen on the anterior wall improved during resting images. Subsequent pharmacological test performed, triggered ischemic manifestations which necessitated coronary angiography. On coronary angiograms, any narrowing or occlusive disease of coronary arteries was not seen. A marked AVF extending from the proximal segment of the left anterior descending artery into the pulmonary artery was detected (Figure 2).

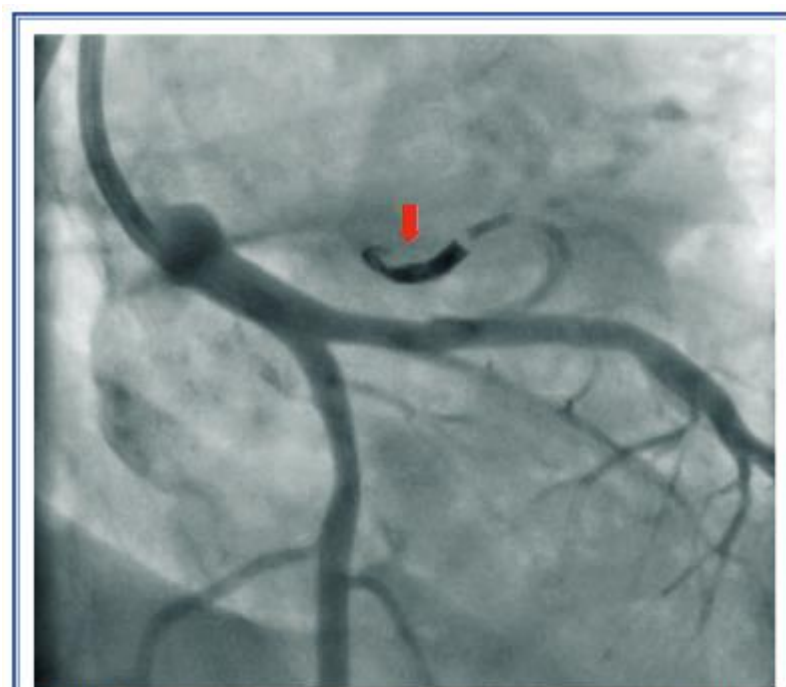


Figure 3 Closure of the arteriovenous fistula via percutaneous coronary intervention using a “coil” , and complete cessation of blood flow through fistula are observed

We attributed onset of existing complaints of the patient predominantly to coronary steal syndrome developed secondary to AVF, and percutaneous coronary intervention was performed to close AVF using 3 coils (Axium™ Helical Detachable Coil [2 mm x 6 cm, and 2 mm x 4 cm]) (Figure 3). Any postprocedural complication did not develop. Clopidogrel (75 mg od), ramipril (10 mg od), and metformin (850 mg tid) therapies were prescribed before discharge. At the control visit performed one month later, the patient reported disappearance of her complaints of chest pain.

DISCUSSION

Coronary AVFs are rarely seen (0.3-0.8 %) entities. They frequently lead an asymptomatic course.^[1] They can be congenital or acquired (traumatic or infectious).^[4] Frequently, they arise from the right coronary artery (55 %) or branches of the left coronary artery.^[2] Though rarely seen, AVFs originating from both coronary arteries have been reported.^[4,5] Coronary AVFs constitute 20 % of all fistulas.^[6] Coronary AVF can form between coronary arteries, and cardiac chambers, coronary sinus, superior vena cava, pulmonary artery or pulmonary veins.^[1,2]

Coronary AVFs are asymptomatic in young patients, while they become symptomatic in advanced ages. Symptoms generally increase dependent on the severity of the left-to-right shunt. In the

coronary steal syndrome, blood flow in non-atherosclerotic coronary arteries is directed into fistula, and other cavities rather than through capillary plexus of myocardium leading to the development of myocardial ischemia. Relative ischemia can develop secondary to apical hypertrophic cardiomyopathy. In our case, since it was thought that pulmonary fistula in the setting of relative ischemia secondary to apical hypertrophic cardiomyopathy further increases severity of ischemia, we decided to close the fistula, and reevaluate her symptoms after the procedure. Other alternatives are also available which facilitate the understanding of the extent of responsibility incurred by the fistula for the development of ischemia. Hemodynamic significance of the shunt secondary to fistula formation can be determined by calculation of systemic/pulmonary blood flow ratio based on blood pressure, and oxymetric measurements via right-, and left catheterization. Besides, during percutaneous coronary intervention, measurements of the fractional flow reserve in the distal left anterior descending artery at baseline, and after closure of the fistula with” coil” can demonstrate hemodynamic improvement in coronary artery. Recurrent myocardial perfusion scintigraphic examinations can reveal whether or not regional ischemia on the anterior myocardial wall is relieved after closure of the fistula.

Even repair of fistulas in asymptomatic patients is still debatable, closure of the fistula is required in symptomatic patients because of potentially developed complications.^[3] Even though gold standard treatment is closure of the fistula using surgical intervention, currently minimal invasive catheterization methods have been employed as an alternative to surgery.^[7] Percutaneous coronary interventions have similar effectiveness, mortality, and morbidity rates comparable to surgical methods.^[7] Development of embolism in previously normal branches of coronary artery during percutaneous coronary intervention has inherent risks of inadvertent migration of the occluder into the heart chambers, and major arteries, and rupture of the fistula.^[7] In conclusion, despite rarity of coronary AVFs, coronary steal syndrome should be conceived in the differential diagnosis of patients with myocardial ischemia.

Conflict of interest: None declared

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Anahtar sözcükler: Arteriyovenöz fistül/epidemiyoloji; göğüs ağrısı; koroner anjiyografi; koroner-subklavya çalma sendromu; koroner damar anomalisi.

Key words: Arteriovenous fistula/epidemiology; chest pain; coronary angiography; coronary-subclavian steal syndrome; coronary vessel anomalies.