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Coronary Artery Fistula Concomitant with Bicuspid Aortic Valve Stenosis

Koroner Arter Fistülü ve Biküspit Aort Kapak Stenozu Birlikteliği

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ABSTRACT

Coronary artery fistulas are usually asymptomatic and rare congenital anomalies. Coronary angiography is the main method for the diagnosis of coronary artery fistulas. Non-invasive methods such as echocardiography may also be helpful for the diagnosis. In this paper we reported a case of a coronary artery fistula concomitant with degenerative bicuspid aortic valve stenosis which was diagnosed with transthorasic echocardiography.

Key Words: Coronary artery, fistula, aortic valve, echocardiography.

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ÖZET

Koroner arter fistülleri nadir konjenital anomalilerdendir ve genellikle asemptomatiklerdir. Koroner anjiyografi, koroner arter fistüllerinin tanısında temel yöntemdir. Ekokardiyografi gibi noninvazif yöntemler tanı konulmasında yardımcı olabilir. Bu yazıda transtorasik ekokardiyografiyle tanı konulan koroner arter fistülü ve dejeneratif biküspit aort kapak stenozu birlikteliği olan olgu sunulmuştur.

Anahtar Kelimeler: Koroner arter, fistül, aort kapağı, ekokardiyografi.

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INTRODUCTION

Coronary artery fistulas are anomalous connections between coronary vessels and the cardiac chambers or the great vessels. Clinical presentations of fistulas are variable and depends mostly on the location and size of the shunt. Most of the fistulas are small and they are incidentally diagnosed by coronary angiography. Coronary angiography is the main approach for the diagnosis of coronary artery fistulas. Non-invasive methods such as echocardiography may also be helpful for the diagnosis. In this paper we reported a case of a coronary artery fistula concomitant with degenerative bicuspid aortic valve stenosis which was diagnosed with transthorasic echocardiography.

CASE REPORT

A 58-year old male patient was admitted to our hospital with a one year history of exercise intolerance and presyncope that had been worsened in the last one month. He had no cardiovascular risk factors other than smoking and family history of premature coronary artery disease. On physical examination, the blood pressure was 110/70 mmHg and heart rate was 80/minute. A cardiac auscultation revealed a grade 3/6 crescendo-decrescendo late-peaking murmur in aortic area that radiated to the carotids. Biochemical parameters were entirely normal. A 12-lead electrocardiogram revealed sinus rhythm at 80 beats/minute with left ventricle hypertrophy and repolarization abnormality. These symptoms and physical examination signs were consistent with critical aortic stenosis. An echocardiogram revealed normal left ventricular systolic function and severe aortic stenosis. The aortic valve was bicuspid in configuration and the aortic valve area was 1.1 cm². The gradient was maximum 90 mmHg and mean 63 mmHg at the valve (Figure 1). The ascending aorta was 49 mm. A diastolic flow with a velocity of 0.9 m/second and which was considered to be



Figure 1. Apical five chamber view of aortic valve.

a fistula, was detected at the modified parasternal long axis view of pulmonary artery (Figures 2,3). There was no increase in pulmonary to systemic flow ratio (Qp/Qs). Systolic pulmonary artery pressure was 35 mmHg. Catheterization was planned for aortic valve area, gradient and symptom incompatibility. A coronary angiography revealed non-critical plaques, and a fistular structure extending from both the left anterior descending (LAD) and circumflex arteries to the pulmonary artery, leading to dense opacification of pulmonary arteries (Figure 4). Unfortunately aortic valve could not passed and invasive pressure measurement was not performed. The patient underwent aortic valve replacement with a 21 size St. Jude mechanical valve and ascending aorta repair with 32 mm Perouse vascular dacron greft. During operation, 4 vascular structures with thrills extending from the LAD and circumflex arteries to the pulmonary artery were detected and a ligation was performed. Since discharge, the patient has been followed up without any symptoms.



Figure 2. Modified parasternal long axis view of pulmonary artery.



Figure 3. Pulse wave Doppler imaging of fistula.



Figure 4. Opacification of pulmonary arteries.

DISCUSSION

Coronary artery fistulas are usually asymptomatic and rare congenital anomalies that are discovered accidentally during coronary angiography in most cases. The incidence has been reported to be lower than 0.2% in large coronary angiographic series⁽¹⁾. Around 55% of coronary artery fistulas arise from the right coronary artery, 35% arise from the left coronary artery and 5% from both coronary arteries⁽²⁾. Additionally, 29.8-43% of all coronary artery fistulas drain into the pulmonary artery, 14-40% drain into the right ventricle, 19-20.2% drain into the right atrium, 5.8-19% drain into the left ventricle and 5% drain into the left atrium⁽³⁾. Clinical presentation of a coronary artery fistula depends mostly on the location and size of the shunt. Severe shunts can be a ssociated with pulmonary hypertension, heart failure, bacterial endocarditis or myocardial ischemia due to coronary steal.

Even though coronary angiography remains the diagnostic method of choice for evaluating the anatomy of the coronary fistulas, non-invasive methods such as magnetic resonance imaging (MRI), multidedector computerized tomography (CT) and echocardiography can be used in follow up examinations and subsequent evaluations because of the increased risk this invasive procedures carries⁽⁴⁾. Multidetector CT has been shown to provide a high-resolution anatomic image by using reconstruction methods. Volume rendered images revealed from threedimensional CT data sets provide a good overview of the vascular anatomy⁽⁵⁾. New MRI sequences have improved image guality with better anatomical definition, and MRI has become an alternative method to evaluate anatomy, flow and function⁽⁶⁾. Thus, multidetector CT and MRI are considered a good alternative to coronary angiography. There are studies reporting imaging of coronary artery fistulas by transthoracic echocardiography⁽⁷⁾. In the case presented here, fistula was not considered severe because of the low velocity and the absence of an increase in the Qp/Qs ratio. Coronary angiography was performed because of valve area and gradient incompability and the fistula was found to cause marked filling of the pulmonary arteries. A developed fistular structure was also found during operation. We consider that, despite its use for the diagnosis of fistulas, transthoracic echocardiography may not always be appropriate and coronary angiography, multidetector CT or MRI should be used in the presence of clinical inconsistency.

CONFLICT of INTEREST

None declared.

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