A Rare Case: Pulmonary Supravalvular Stenosis With Moderate Gradient Causing An Atrial Right To Left Shunt Across The Reopened Foramen Ovale

Ender bir olgu: Yeniden açılan foramen ovale nedeniyle sağdan sola şanta yol açan orta düzeyde gradyanlı pulmoner supravalvuler stenoz

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Abstract
Supravalvular pulmonary stenosis accompanied by a right to left shunt is a rare echocardiographic finding in adults. We report a case of adult pulmonary supravalvular membranous stenosis with an atrial right to left shunt developed through a previously undiagnosed patent foramen ovale (PFO).The peculiarity of our case is that foramen ovale reopened despite a moderate pulmonary trunk gradient over a six year period. This congenital anomaly is a condition to be differentiated from valvular pulmonary stenosis as both anomalies require different modalities of treatment (surgical or catheter).
Key words: Pulmonary,supervalvular stenosis, foramen ovale.

Özet
Sağdan sola şantın eşlik ettiği supravalvuler pulmoner stenoz erişkinlerde ender görülen bir ekokardiyografi bulgusudur. Önceden tanısı konmamış patent foramen ovaleden gelişen sağdan sola şantın eşlik ettiği supravalvuler pulmoner stenoz gelişen hastanın olgusu sunulmuştur. Bu konjenital anomali, valvuler pulmoner stenozdan tedavi yöntemlerinin ayrı olmasından dolayı ayırıcı tanısı yapılmalıdır.
Anahtar kelimeler: Pulmoner, supervalvular stenoz,foramen ovale.

Case

A 38 years old female patient was admitted for evaluation of exertional dyspnea and simultaneous cyanotic fingers and toes in the absence of chest pain and syncope. She had no symptoms on admission ,however, her physical activity was graded as Class I according to the classification of the New York Heart Association. She had no history of thoracic surgery, catheterization or drug abuse. In physical examination, her blood pressure was 125/85 mmHg, her heart rate was 78/min and a systolic murmur (Levine II/VI) was audible at the left sternal border in the 3rd intercostal space. ECG showed incomplete right bundle branch block. Blood tests were in normal ranges, with absence of alterations related to a high output state like anemia or hyperthyroidism.Transthoracic and transesofagial echocardiography visualized a normal right ventricle without hypertrophy (wall thickness: 4mm) (Figure 1),having an internal diameter of 32mm at basal level from the four-chamber view, a supravalvular membranous-structured stenosed pulmonary artery 1cm above the pulmonic valve and a PFO tunnel on interatrial septum which had been visualized and reported as intact six years ago. In contrast echocardiography,

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bubbles appeared in left atrium passing through the PFO tunnel following second heart beat after the injection Systolic pressure gradient between the right ventricle and the distal main pulmonary trunk was 32 mmHg ,moderate pulmonary stenosis. Her blood gas analysis showed a mild hypoxic state with a pO2:77 mmhg.The patient was diagnosed as having pulmonary

supravalvular membranous stenosis with a right to left shunt through patent foramen ovale. The underlying ethiologies of reopening on foramen ovale in last 6 years could not be identified and she has been referred to a cardiovascular surgery institution for catheterization and surgical evaluation.

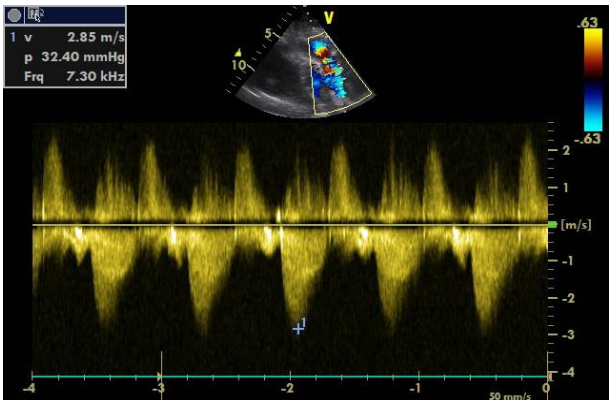

Figure 1. Transthoracic echocardiography findings of a normal right ventricle without hypertrophy (wall thickness: 4mm).

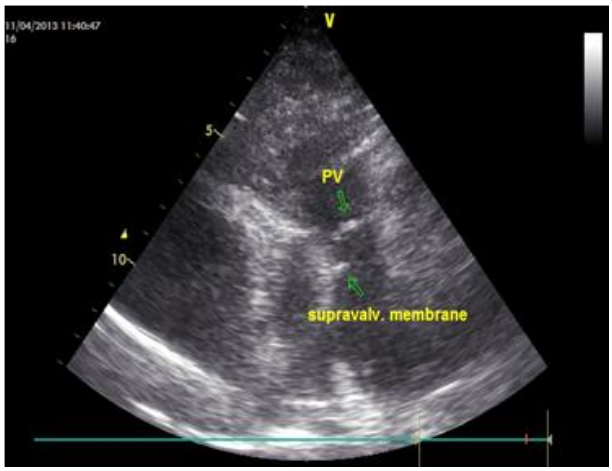

Figure 2. Supravalvular membrane.

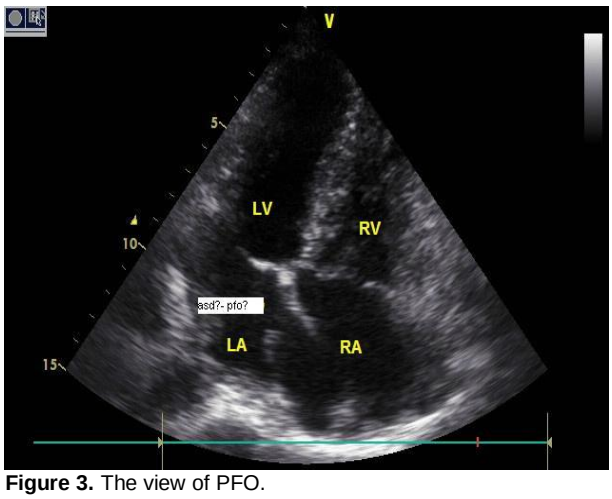

Figure 3. The view of PFO.

Discussion

Supravalvular stenosis of the pulmonary artery is a rare and often missed echocardiographic finding, particularly in adults[2]. Most frequently, the obstruction occurs at the level of the pulmonary valve. Lesions at any level can occur as part of more complex congenital cardiac malformations such as tetralogy of Fallot ,dextro- or levo-transposition of great arteries or atrial septal defect. Isolated type stenosis,on the other hand, is commonly associated with the congenital rubella syndrome[3]. Some patients may have residual or secondary supravalvular pulmonary stenosis or peripheral pulmonary stenosis after surgical procedures such as mechanical pulmonary valve replacement which necessitates an additional interventional therapy, or after an arterial switch operation for transposition [4-6]. However, our patient had no history of any operation or intervention. To the best of our knowledge, no other adult case of pulmonary supravalvular stenosis accompanied by reopened foramen ovale and a moderate degree of transstenotic pressure gradient was reported in the literature.

Cardiac catheterization may reveal the exact mechanisms of right to left shunt and contributes for the decision of percutaneous transluminal balloon dilatation or surgical correction.This type of lesions should be differentiated from valvular pulmonary stenosis as surgical intervention is preferred for the valvular ones[1,6].

This case demonstrates also the utility of echocardiography (transthoracic and transesophageal examination) in the diagnosis and quantification of anomalies of the great cardiac vessels and valves.

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