Multiple Congenital Peripheral Pulmonary Artery Stenoses

Konjenital Multipl Periferal Pulmoner Arter Darlığı

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A 22-year-old male patient with a previous history of surgery for supra-valvular aortic stenosis in childhood was referred to our unit with a diagnosis of pulmonary hypertension for further assessment. Transthoracic echocardiography showed the presence of right ventricular dilatation, moderate tricuspid regurgitation and systolic arterial pressure of 70 mmHg. Right heart catheterization revealed a mean pulmonary artery pressure of 50 mmHg. Magnetic resonance angiography imaging showed bilateral multiple severe stenoses of the peripheral pulmonary artery (Figure 1). A diagnosis of pulmonary hypertension due to multiple peripheral pulmonary artery branch stenosis was made. Based on the previous history of surgery for supra-valvular aortic stenosis, the case was deemed to have a congenital condition.

Peripheral pulmonary artery stenosis is defined as an obstruction of the pulmonary artery

from the pulmonary trunk to the peripheral artery, and was first reported by Oppenheimer in $1938^{(1)}$. Peripheral pulmonary artery stenosis has been a disease with limited treatment options. In contrast with our case, most congenital pulmonary artery stenoses are located in the main or proximal segmental pulmonary artery branches, and therefore surgery or balloon angioplasty may represent the treatment modalities of choice⁽²⁾.

REFERENCES

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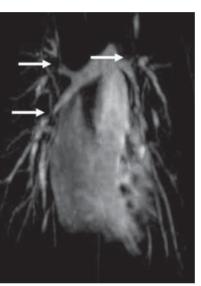


Figure 1. Bilateral multiple severe stenoses of the peripheral pulmonary artery (Arrows).



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