Ventricular septal defect with bidirectional shunting in a patient with congenitally corrected transposition

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Huge main pulmonary arterial thrombus in a child with increased lipoprotein (a) level

Lipoprotein (a) yükseklüğü olan bir çocukta pulmoner arteriyel dev trombus

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Pulmonary arterial thrombosis is an extremely rare clinical condition both in children and in adults. Lipoprotein (a) [Lp (a)] is an atherogenic
lipoprotein particle which displays adjunctive thrombotic properties by inhibition of the fibrinolytic pathway.

A 9-year-old boy with no previous history of cardiac or pulmonary disease was referred to our hospital for investigation of dyspnea and tachypnea that had started one month ago. In the echocardiogram and computed tomography (CT)-angiogram, a large thrombus was seen in the main pulmonary artery bifurcation which almost completely obstructed the right pulmonary artery and partially the left one with tricuspid regurgitation and dilation of the right chambers of the heart (Fig. 1, 2).

Thrombophilia screening was normal except high Lp (a) concentration (1.33 g/L, normal: 0.01-0.30 g/L). We performed family screening for Lp (a) and found a high Lp(a) level (0.9 g/L) in his father. While the patient was on the heparin therapy, a sudden increase in his tachypnea, dyspnea and anxiety occurred and he complained of pleuritic chest pain. Since pulmonary perfusion scintigraphy with Tc99 macroaggregated albumin revealed bilaterally perfusion defects in the lungs, the patient was immediately referred for thrombectomy and the thrombus was removed totally (Fig. 3). As a result, he was discharged from the hospital under warfarin and low-dose aspirin therapy.

Elevated Lp (a) is a very rare cause of venous and arterial thromboembolism and should be checked in such cases both to determine the etiology of the thrombus and for detecting the other family members with increased Lp (a) level and thromboembolism risk.

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