

**Giant pulmonary artery
aneurysm in a patient with
mixed connective tissue disease**

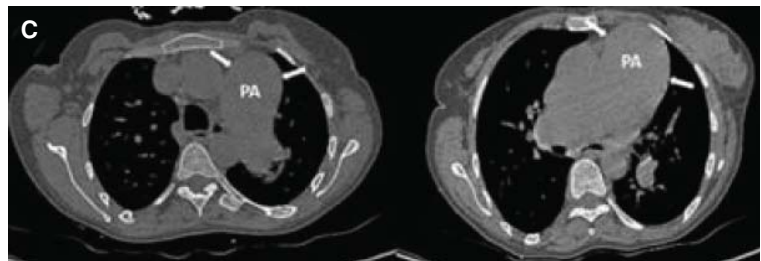
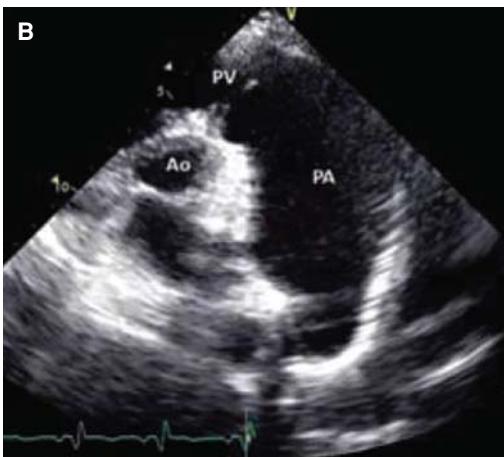
**Mikst bağ dokusu hastalığı
olan bir olguda dev
pulmoner arter anevrizması**

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A 40-year-old woman presented with complaints of severe headache, clouded consciousness, palpitation, and dyspnea. Physical examination revealed a blood pressure of 210/110 mmHg, rhythmic heart sound and a heart rate of 87 beats/min. A 3/6 systolic murmur was heard at the apex, while a 2/4 diastolic murmur was heard at the pulmonary focus. Basal lung sounds were found to be reduced. Sclerotic lesions were observed on both hands and on the face of the patient, and erythematous lesions on her nose. Bullous keratopathy was noticed in the eyes. The patient complained of increasing difficulty in swallowing for the past three years. She was admitted to the cardiology intensive care unit for follow-up due to the diagnosis of hypertensive encephalopathy. Neurologic consultation was performed and recommendations from the Neurology Department were included in her therapy. The patient's consciousness increasingly improved with antihypertensive therapy. Her x-ray revealed hilar fullness and strain findings at the base of the lungs, while the area consistent with the pulmonary artery was observed to be dilated (Figure A). Transthoracic

echocardiography revealed a hypertrophic left ventricle, normal systolic functions, mild mitral regurgitation, 3rd degree tricuspid regurgitation, and a pulmonary artery pressure of 75 mmHg. The interatrial septum was found to be aneurysmatic, while the interventricular and interatrial septum found to be were displaced to the left. The right ventricular diameter was 4.8 cm, while the pulmonary artery was measured as 6.3 cm at the widest point and evaluated as aneurysmatic, and a 2nd degree pulmonary regurgitation was observed (Figure B). A thoracic computed tomography performed for detailed evaluation of the pulmonary artery showed pulmonary diameter of 6.5 cm at the widest point (Figure C), and the lung findings were suggested to be an involvement consistent with a collagenous tissue disease. Analysis of autoimmune and serologic markers demonstrated antinuclear antibody (ANA) IFAT as positive, the ANA subgroup antiribonucleoprotein (RNP) as positive, rheumatoid factor as 260 IU/ml, while the CRP (sensitive) was measured as 101 mg/dl. Treatment with iloprost was initiated for the medical treatment-resistant pulmonary hypertension. The patient was discharged with medical therapy and regular follow-up schedule.



Figures. (A) Lung image showing hilar fullness, strain findings at the base of the lung, and dilation of the pulmonary conus. (B) Transthoracic echocardiography image of pulmonary artery aneurysm measured as 6.3 cm at the widest point in the parasternal axis view. (C) Thoracic computed tomography image of two different sections of a giant pulmonary artery aneurysm measured as 6.5 cm at the widest point.