[PICTURES IN CLINICAL MEDICINE]

Long-surviving Anomalous Origin of the Right Pulmonary Artery from the Ascending Aorta Complicated with Pulmonary Arteriovenous Fistula

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Picture.

A 41-year-old woman was referred to our hospital due to exertional dyspnea. Contrast-enhanced computed tomography (CT) showed anomalous origin of the right pulmonary artery from the ascending aorta (AORPA) and polyarteriovenous fistula in the right lung field (Picture, arrow heads). Right heart catheterization showed slight elevation of the left mean pulmonary pressure (26 mmHg), with a normal pulmonary capillary wedge pressure (13 mmHg). The patient elected to undergo follow-up rather than surgical repair and is now 48 years old, making her the longest-surviving uncorrected patient with AORPA recorded in the literature (1). AORPA is a rare congenital heart disease with a high mortality rate, resulting in pulmonary hypertension and heart failure without surgical repair because the right

pulmonary artery is exposed to pressure overload from the systemic circulation, and the left pulmonary artery is exposed to volume overload from the systemic venous return (2). In this case, pulmonary polyarteriovenous fistula may be serving as a shunt, reducing the pulmonary arterial resistance and preventing right pulmonary arterial hypertension, leading to this patient's unusually long and ongoing survival.

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