

In our patient, the excellent response and disappearance of systemic disease led us to question the need to continue with treatment, since no clear directives are currently available. During his off-treatment period, the patient presented cerebral progression. This led us to consider 2 hypotheses: either a T790M resistance mutation had occurred, or the tumor was extremely dependent on the EGFR pathway, and when the drug was withdrawn, breakthrough disease developed with progression in a sanctuary site. After molecular analysis, the second option appears more plausible, and raises once again the issue of the best management of long-term survivors receiving anti-EGFR treatment.

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Pulmonary Arterial Hypertension Secondary to Partial Anomalous Pulmonary Venous Return in an Elderly Patient[☆]



Hipertensión arterial pulmonar secundaria a un drenaje venoso pulmonar anómalo parcial en una paciente anciana

To the Editor:

Partial anomalous pulmonary venous return (PAPVR) is an uncommon congenital abnormality that can be diagnosed in adult life, although it is more often detected during childhood.¹ It consists of abnormal, incomplete pulmonary venous return to the systemic venous circulation (superior vena cava, azygous vein, coronary sinus, brachiocephalic vein, inferior vena cava, etc.) causing

left-to-right shunt. PAPVR is more common in the right side, and is often associated with other congenital abnormalities, such as heart defects (particularly atrial septal defect), or an abnormally developed airway.² We report a case of a patient in her seventies with left PAPVR that began with signs and symptoms of pulmonary arterial hypertension (PAH).

The patient was a 76-year-old woman with no significant medical history who consulted due to progressive dyspnea, edema in the lower limbs, and discomfort in the chest and abdomen. Chest radiography showed cardiomegaly and scant bilateral pleural effusion. Signs of right heart overload were observed on electrocardiogram, and D-dimer was slightly elevated. A chest CT angiogram ruled out pulmonary thromboembolism, but unexpectedly revealed a PAPVR in which the veins of the left upper lobe drained to the brachiocephalic vein via a vertical vein (Fig. 1A). Radiological signs of severe

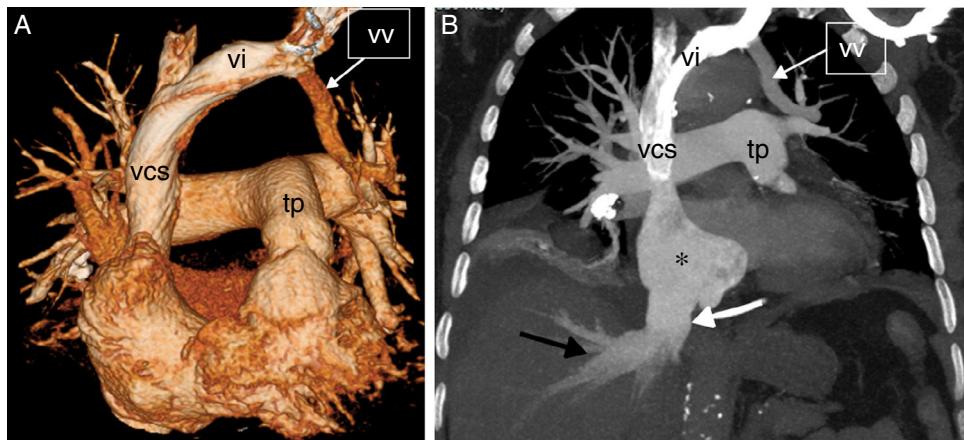


Fig. 1. (A) Computed tomography with 3D volume rendering showing a vertical vein (vv), formed by the confluence of pulmonary veins of the left upper lobe, running toward the brachiocephalic vein (vi); tp: pulmonary artery trunk; vcs: superior vena cava. (B) Coronal computed tomography with maximum intensity projection (MIP) showing signs of arterial/pulmonary hypertension and tricuspid valve insufficiency: dilation of right atrium (asterisk), inferior vena cava (white arrow) and suprahepatic veins (black arrow) congestion. Note the vertical vein (vv), the brachiocephalic vein (vi), and the superior vena cava (svc).

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PAH were also observed, with dilation of the right heart cavities and congestion of the inferior vena cava and the suprahepatic veins (Fig. 1B).

PAPVR is a rare congenital abnormality that is usually diagnosed in children, although it may also be detected in adults, particularly if they develop PAH. Less than 10% of PAPVRs are left-sided, and up to 80% are associated with atrial septal defect (ASD). This case reminds us that PAPVR, if untreated, leads to volume overload of the right heart, tricuspid valve insufficiency, PAH, and finally right ventricular failure.^{3,4} Our case is interesting for several reasons: the advanced age of the patient when diagnosed with PAPVR, the left-sidedness of the PAPVR, and the absence of an associated ASD. PAPVR should always be suspected, even in elderly patients, in cases with "primary" or unexplained PAH.

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