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Isolated Left Inferior Pulmonary Vein Anomalous Connection in an Older Woman (Rarest Variant of Partial Pulmonary Anomalous Venous Connection)

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Received: Aug 24, 2020
Accepted: Oct 01, 2020
Published Online: Oct 05, 2020
Journal: Journal of Clinical Images
Publisher: MedDocs Publishers LLC
Online edition: http://meddocsonline.org/

Clinical image description

A 75-year-old woman with a past medical history of hypertension was referred because moderate effort dyspnea, Right Bundle Branch Block (RBBB) and frequent supraventricular premature beats.

Cardiac Magnetic Resonance (CMR) revealed a dilated RV (iEDV 97ml/m2) with normal ejection fraction (RVEF 64%), and without Atrial Septal Defects (ASD). By phase contrast sequence we obtained a Qp: Qs=2:1 (augmented). CMR SSFP cine sequences (PANEL A1, A2, A3) showed a Partial Pulmonary Anomalous Venous Connection (PPAVC) with the Left Inferior Pulmonary Vein (LIPV) draining into the Innominate Vein (InV) through a Vertical Vein (VV); the other main pulmonary veins were normally connected to the Left Atrium (LA). Cardiac Computed Tomography (CCT) (PANEL B1, B2, B3) confirmed an isolated LIPV anomalous connection, and demonstrated that there were as well two small pulmonary veins from the left upper lobe that drained into the VV. Right heart catheterization revealed normal pulmonary artery pressures (sys: 33 mmHg/ dias: 8 mmHg/ mean: 18 mmHg) and pulmonary vascular resistance (1.7 Woods units).

PPAVCs are rare congenital anomalies in which one or more (but not all) of the pulmonary veins connect to a location other than the left atrium, and the most common pathological variants involve the right pulmonary veins and are accompanied by an ASD. When the PPAVCs affect the left-sided pulmonary veins, the commonest pattern is the Left Superior Pulmonary Vein (LSPV) connected to the innominate vein through a vertical vein [1].

Cite this article: Avila-Sanchez DA, Barreiro-Perez M, Cambronero-Cortinas E, Garcia-Lanzas F, Sanchez PL. Isolated Left Inferior Pulmonary Vein Anomalous Connection in an Older Woman (Rarest Variant of Partial Pulmonary Anomalous Venous Connection). J Clin Images. 2020; 3(1): 1066.

The anomaly presented by our patient (isolated LIPV anomalous connection) is extremely rare and has not been described in previous pathological reports [2] or in the largest cohort of patients with PPAVCs studied by CCT [3]. The right-sided heart dilatation and Qp: Qs = 2: 1 are indications for surgical repair, regardless of age; however, our patient decided to continue a conservative strategy with close regular follow-up.

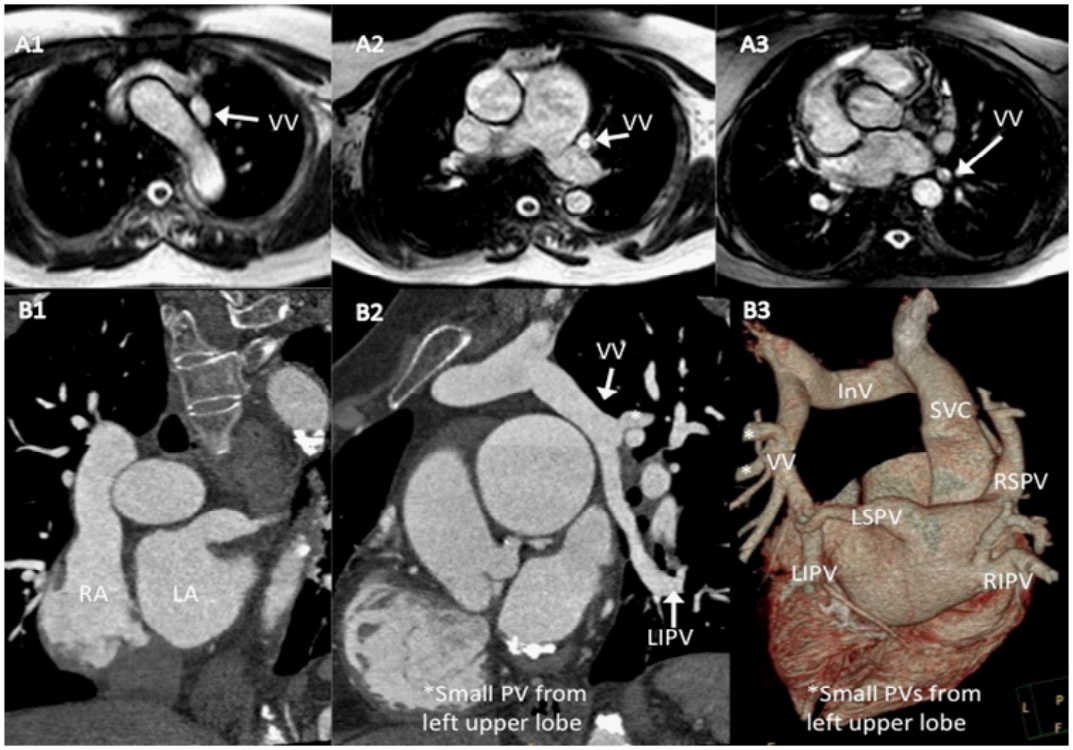

Figure 1: Panel A1, A2 and A3: Cardiac magnetic resonance SSFP cine images. Vertical vein lateral to the aortic arch prior to draining into the left brachiocephalic vein (innominate vein). Main pulmonary artery severely dilated (46mm).

Panel B1, B2 and B3: Cardiac computed tomography images. Intact interatrial septum. Partial pulmonary anomalous venous connection characterized by a left inferior pulmonary vein draining into the innominate vein through vertical vein.

Inv: Innominate Vein; LA: Left Atrium; LIPV: Left Inferior Pulmonary Vein; LSPV: Left Superior Pulmonary Vein; LIPV RA: Right Atrium; PV: Pulmonary Vein; RIPV: Right Inferior Pulmonary Vein; RSPV: Right Superior Pulmonary Vein; SVC: Superior Venous Cavous; VV: Vertical Vein.

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