Rare variant of mixed total anomalous pulmonary venous drainage (TAPVD) in a 55-year-old patient

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A 55-year-old man presented with recent-onset oedema of lower extremity. He had cyanosis and clubbing since childhood, but his exercise tolerance was relatively good. Transthoracic echocardiography (TTE) demonstrated significantly dilated and hypertrophied right ventricle, moderate pulmonary stenosis and an atrial septal defect (ASD) of 27 mm with right-to-left shunt (Panels A–C and Supplementary data online, Video S1). Peak pressure gradient of tricuspid regurgitation (PG-tr) and of the pulmonary valve (PG-pv) were 67 and 54 mmHg, respectively. A pulmonary vein was connected into the remarkably dilated coronary sinus (CS) (Panels D–E and Supplementary data online, Video S2 and S3). A vertical vein was detected in the suprasternal view (Panel F and Supplementary data online, Video S4). No pulmonary veins were found to be connected into the left atrium. The diagnosis of mixed total anomalous pulmonary venous drainage (TAPVD) with intracardiac and supracardiac connections was suggested from TTE. Enhanced computed tomography (CT) confirmed this diagnosis (Panels G–I). The left-side pulmonary veins (LPV) joined to a vertical vein draining into the left innominate vein and then the superior vena cava. The right-side pulmonary veins (RPV) formed a confluence draining into the CS.

Mixed TAPVD with intracardiac and supracardiac drainage is a rare condition which has been reported in just a few cases of infants. The long survival of this adult patient is benefited from moderate pulmonary stenosis and large ASD, as well as the absence of pulmonary venous obstruction. Considering the risk associated with surgical correction of this defect, the patient was medically managed.

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