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Isolated persistent left superior vena cava with giant coronary sinus presenting as atrial tachycardia

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Introduction: Persistence of a left-sided superior vena cava (PLSVC) with absent right superior vena cava (isolated PLSVC) is a rare venous malformation commonly associated with congenital heart disease or alterations of the cardiac situs. We describe an unusual case of a patient with isolated PLSVC presenting with persistent atrial tachycardia and congestive heart failure.

Case presentation: A 40-year-old male presented with palpitations, gradually progressive shortness of breath, abdominal distension and bilateral swelling of feet of 3 months duration. On physical examination, he was tachypneic with a pulse rate of 140/min which was regular and low volume. The jugular venous pressure was elevated. There was hepatomegaly and bilateral pedal edema. Investigations: ECG showed atrial tachycardia at a rate of 140/min. Transthoracic echocardiogram revealed situs solitus, dilated right atrium (RA) and right ventricle (RV) with moderate RV systolic dysfunction. Mild tricuspid regurgitation (TR) was noted. The coronary sinus (CS) was dilated (3.4 cm) with spontaneous echo contrast.

Injection of agitated saline from a left antecubital vein was performed to confirm PLSVC draining into the CS. This resulted in opacification of the CS earlier than into RA confirming the above.

Injection of agitated saline into right antecubital vein (figure A) also resulted in early appearance of the saline contrast in the CS prior to its appearance in the main RA cavity, confirming the absence of a right superior vena cava. Interatrial and interventricular septi were intact.

The diagnosis of absent right superior vena cava and isolated persistent left superior vena cave were confirmed by cardiac CT (Figure B). The persistent atrial tachycardia was probably due to the stretching of the dilated RA and dilated CS.

Conclusion: Isolated persistent left superior vena cave is a rare condition and needs bilateral saline injection and CT to confirm its diagnosis. The condition is usually discovered incidentally, however, in our case it presented with persistent supra ventricular tachycardia and right sided heart failure.
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