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Case Report

Persistent right superior vena cava in a patient with dextrocardia: Case report and review of the literature

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ABSTRACT

Introduction: Systemic venous circulation anomalies are uncommon; they are often incidental findings during echocardiography.
Case: A 56-year-old man, with dextrocardia, was evaluated for dyspnea. The patient's medical history included diabetes mellitus requiring insulin treatment, hypertension, and tobacco use. Physical examination revealed normal jugular venous pulsations and clear lungs. Cardiac examination revealed normal heart sounds, and grade II/VI systolic ejection murmur over the right precordium. Echocardiography revealed normal chamber size and systolic function, without significant valvular lesions. The coronary sinus was dilated. It was evaluated using intravenous agitated saline contrast to rule out anomalous venous drainage or shunting. When injected into the left antecubital vein, contrast appeared initially in the right atrium followed by the right ventricle. However, when injected into the right antecubital vein, contrast appeared initially in the dilated coronary sinus followed by the right atrium and right ventricle. There was no evidence of intracardiac shunting. These findings were consistent with persistent right superior vena cava in the setting of situs inversus dextrocardia, with normally draining left superior vena cava.
Conclusion: Persistent superior vena cava connection to the coronary sinus is often incidental but an important finding which helps in planning safe invasive procedures.
<**Learning objective:** Understand the importance of identifying anomalous venous connections with regard to catheter-based procedures. Appreciate the incidence of these vascular anomalies in the normal population and in congenital heart disease. Understand how echocardiography with intravenous agitated saline contrast can be helpful in the diagnosis of such anomalous venous connections.>

Introduction

Anomalies of the systemic venous circulation are not uncommon in patients with congenital heart disease, and can pose a risk if unrecognized prior to diagnostic or therapeutic catheter procedures requiring the use of systemic veins. We present a case of a 56-year-old man with dextrocardia and situs inversus who was found to have a persistent right superior vena cava (PRSVC) by 2D echocardiography using intravenous agitated saline contrast performed to evaluate his dilated coronary sinus.

Case report

A 56-year-old man, with situs inversus dextrocardia, was evaluated for dyspnea. The patient's medical history included diabetes mellitus requiring insulin treatment, hypertension, and tobacco use. Physical examination revealed normal jugular venous pulsations and clear lungs. Cardiac examination revealed normal right-sided point of maximal impulse, normal heart sounds, and grade II/VI systolic ejection murmur over the right precordium. Electrocardiogram revealed sinus rhythm and dextrocardia, without ischemia (Fig. 1). Chest X-ray revealed no acute pathology (Fig. 2). Cardiac catheterization revealed normal coronary arteries and normal ejection fraction. Echocardiography revealed normal chamber size and left ventricular systolic function, without significant valvular lesions. There was an incidental dilated coronary sinus (Fig. 3), which was evaluated using intravenous

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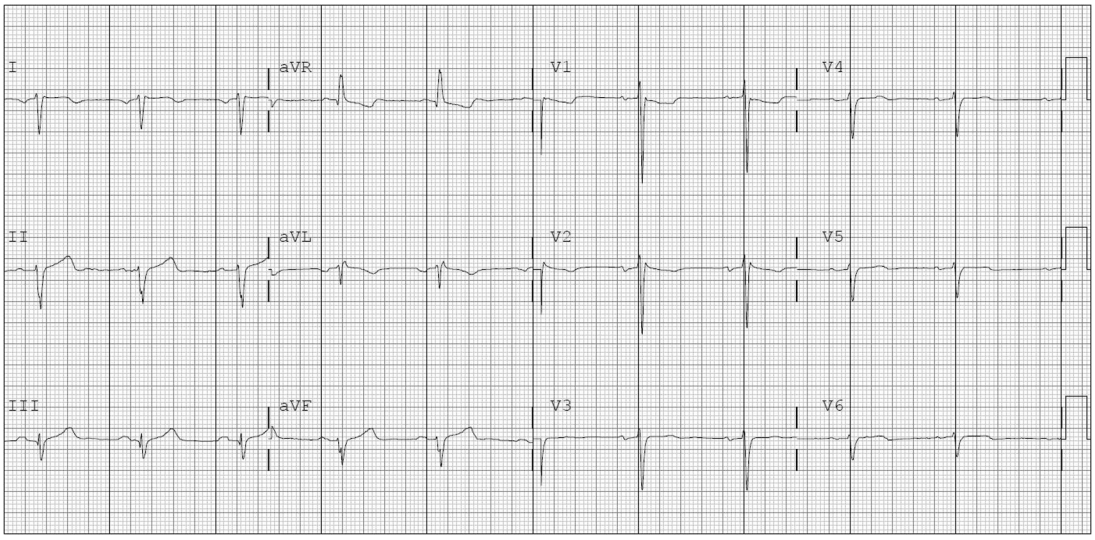

Fig. 1. Electrocardiogram showing sinus rhythm with features of dextrocardia.

(IV) agitated saline contrast to rule out anomalous venous drainage or shunting. When injected into the left antecubital vein, the contrast appeared initially in the right atrium followed by the right ventricle (Fig. 4). However, when injected into the right antecubital vein, the contrast appeared initially in the dilated coronary sinus followed by the right atrium and right ventricle (Fig. 5). There was no evidence of intracardiac shunting. These findings were consistent with persistent right superior vena cava in the setting of situs inversus dextrocardia, with normally draining left superior vena cava. Due to the absence of an obvious cardiac cause of his dyspnea and his tobacco use history, pulmonary function tests were performed, which revealed moderate restrictive lung disease and increase in airway resistance. This was thought to explain his symptoms; therefore, pulmonary clinic follow-up was arranged for further diagnosis and therapy.

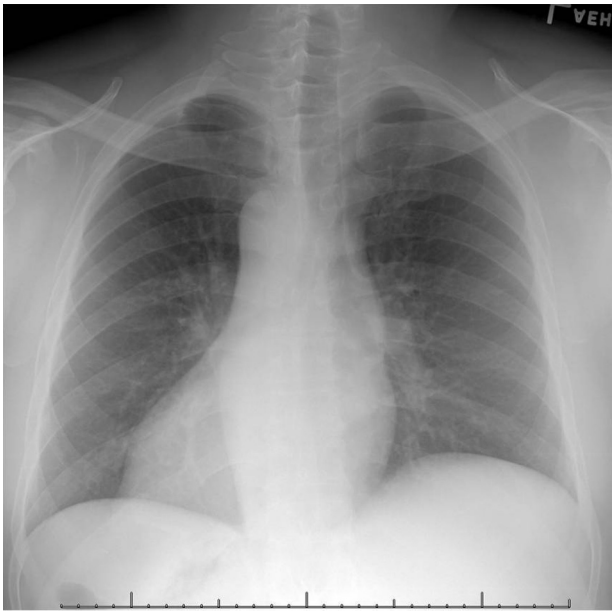

Fig. 2. Chest X-ray with situs inversus dextrocardia showing part of the stomach air on the right side.

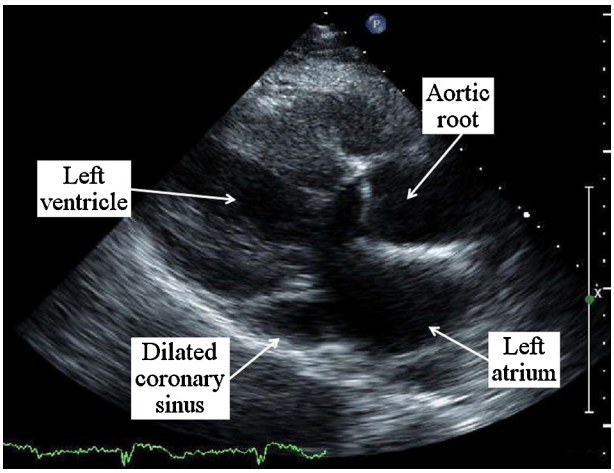

Fig. 3. Parasternal long-axis 2D echocardiographic view showing the dilated coronary sinus, which measured 2.5 cm in transverse diameter.

Discussion

Anomalous connection of systemic venous circulation to the coronary sinus has been previously reported. The most common is a persistent left superior vena cava (PLSVC), which has been reported in approximately 0.5% of the general population, and about 3–10% in patients with congenital heart disease [1]. These anomalies are usually asymptomatic and are discovered incidentally. They have also been reported in patients with dextrocardia, a rarer anomaly in itself. Reports of PLSVC, in addition to pertinent studies of anomalous superior vena cava connections in dextrocardia, will be reviewed to demonstrate the clinical implications of these often incidental findings, especially in relation to catheter-based procedures. The following four case reports will demonstrate the difficulties encountered during pacemaker insertion, and central venous line and Swan–Ganz placement in patients with previously unrecognized PLSVC.

Bhatti and colleagues [2] reported a PLSVC draining into a dilated coronary sinus, associated with anomalous left hepatic vein drainage into the right atrium, in an 86-year-old man undergoing pacemaker implantation for complete heart block. The left