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Coronary Sinus Ostial Atresia with Coronary Sinus Thrombosis

A 74-year-old male with a significant medical history including cocaine use disorder, neurogenic bladder with suprapubic Foley catheter, heart failure with reduced ejection fraction, atrial fibrillation, deep vein thrombosis, sick sinus syndrome with pacemaker, chronic kidney disease stage III, chronic obstructive pulmonary disease, coronary artery disease, and type 2 diabetes mellitus was found unresponsive outside an assisted living facility. On arrival, the patient was hypotensive with a blood pressure of 80/45 mm Hg. He complained of midline spine tenderness but denied fever or abdominal pain. Electrocardiogram was unremarkable. Laboratory tests were notable for elevated white blood cell count of 22 000/µL (reference: 4000-11000/µL), acute kidney injury with a creatinine level of 3.48 mg/dL (baseline 1.4 mg/dL, reference: 0.6-1.2 mg/dL), and elevated brain natriuretic peptide at 13 000 pg/mL (normal: <100 pg/mL). Chest computed tomography (CT) angiography and abdominal CT was performed to exclude acute pathologies and showed no acute pathology. Chest CT angiography revealed significant coronary venous dilatations, persistent left superior vena cava, and a nodular filling defect in the dilated coronary sinus (Figure 1). Venous phase abdominal CT images demonstrated the absence of the coronary sinus ostium and a low attenuation lesion within the dilated coronary sinus, consistent with thrombus (Figure 2). Transesophageal echocardiography confirmed the diagnosis and low-molecular-weight heparin was initiated. Follow-up echocardiography after 4 weeks confirmed a significant decrease in thrombus size.

Coronary sinus atresia is a rare congenital defect characterized by the absence or abnormal closure of the coronary sinus, the primary vein that collects deoxygenated blood from the myocardium and drains it into the right atrium.^{1,2} This condition can cause venous blood from the coronary circulation to be rerouted through alternate pathways such as the Thebesian veins or a persistent left superior vena cava, as in the present case. The hemodynamic consequences of coronary sinus atresia can be significant and potentially cause increased pressure in the coronary veins, which may lead to impaired coronary circulation and myocardial ischemia, even in the absence of coronary artery disease.³ Coronary sinus atresia may predispose individuals to coronary sinus thrombosis, although exceptionally rare. Stagnant blood flow can create conditions favorable for thrombus formation. Coronary sinus thrombosis is a significant clinical condition, as it can obstruct the drainage of deoxygenated blood from the myocardium, exacerbating issues of impaired coronary circulation.^{4,5} While less common, a thrombus in the coronary sinus can potentially dislodge and travel to the pulmonary circulation, leading to a pulmonary embolism.6

Coronary sinus atresia is often associated with other congenital anomalies, such as a persistent left superior vena cava, atrial septal defects, and complex cyanotic congenital heart diseases. Patients might present with signs of heart failure, chest pain, or arrhythmias or may be asymptomatic and discovered incidentally during imaging or surgery.^{2,3} Diagnosis of coronary sinus atresia typically involves imaging techniques. Echocardiography, particularly with Doppler imaging, can help visualize abnormal blood flow patterns. Cardiac magnetic resonance imaging and CT



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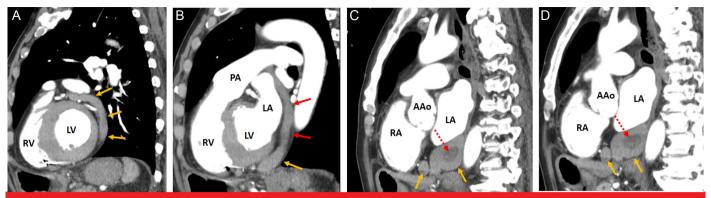


Figure 1. Consequence sagittal chest computed tomography angiography images (A-D) demonstrate the dilated coronary veins (yellow arrows), coronary sinus thrombosis (dashed red arrows), and persistent left superior vena cava (red arrows). RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; AAo, ascending aorta; PA, pulmonary artery.

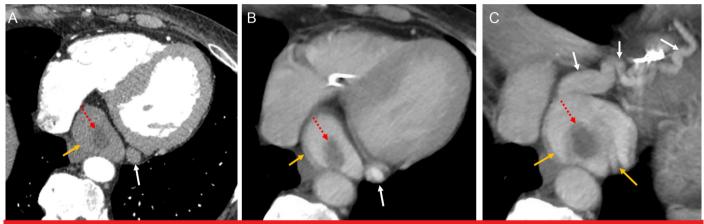


Figure 2. Axial chest computed tomography (CT) angiography (A) and venous phase CT (B) images show enlarged coronary sinus (yellow arrows), a low attenuation filling defect within the coronary sinus (dashed red arrows), and lower part of the persistent left superior vena cava (white arrows). Axial venous phase CT (C) image at the level of posterior interventricular sulcus shows enlarged coronary sinus (yellow arrows), a low attenuation filling defect within the coronary sinus consistent with thrombus (dashed red arrow), and dilatation of the middle cardiac vein (white arrows).

angiography provide detailed anatomical information, while cardiac catheterization may be used for definitive diagnosis and to assess hemodynamic significance.^{3,4}

Management of coronary sinus atresia depends on the severity of symptoms and associated defects. In asymptomatic cases, regular monitoring may be sufficient. Symptomatic patients or those with significant associated defects may require surgical intervention to reroute blood flow and alleviate symptoms. Coronary sinus atresia significantly increases the risk of coronary sinus thrombosis, which can have severe clinical consequences. In patients with coronary sinus thrombosis, anticoagulation therapy and follow-up are essential. Early detection and appropriate management are crucial for improving outcomes. 1.6

Informed Consent: Informed consent was obtained from the patient for the anonymous use of materials taken from him in all kinds of research following the necessary procedures.

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