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CASE REPORT

Coronary Sinus Atresia With Persistent Left Superior Vena Cava: Unusual Clinical Presentation and Endovascular Management

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Abstract Atresia of the coronary sinus (ACS) is a rare congenital anomaly. When associated with persistent left superior vena cava (PLSVC), this defect could have no significant hemodynamic effect, and the patient might remain asymptomatic. However, vascular interventions might induce changes or complications that could show the anomaly. Appropriate management requires a good understanding of this condition. We present the first reported case of ACS and PLSVC occurring after thrombosis of the innominate vein (IV) after central venous catheter placement. The patient presented with atypical subacute chest pain and recurrent extrasystoles. Diagnosis and characterization of vascular anomalies was made by computed tomography phlebography, and the patient was successfully managed by endovascular recanalization of the IV.

Keywords Congenital coronary anomalies · Coronary sinus atresia · Left superior vena cava · Catheters and venous catheterization · Stent

Introduction

Congenital anomalies of the superior vena cava system, including myocardial drainage, represent a large and heterogeneous group of anomalies, the physiological consequences of which may vary from no symptoms to the most severe form of right-to-left shunt inducing systemic arterial deoxygenation. In the majority of asymptomatic patients, the physiopathologic effect of such anomalies or anatomic variants is negligible. However, vascular interventions, such as surgical ligation or placement of an intravascular device, might induce changes in the preexisting anatomic and physiologic equilibrium with unexpected consequences depending on the nature of anomalies and their physiologic supplies.

We present a case of asymptomatic patient with undiagnosed congenital atresia of the coronary sinus (ACS) associated with persistent left superior vena cava (PLSVC) who developed clinical symptoms of myocardial congestion caused by occlusion of the innominate vein (IV) after central catheter placement.

Case Report

A 73-year-old patient, with no previous medical history except gastrectomy 20 years earlier, was admitted to the Emergency Department with acute abdominal pain and ileus. Laparotomy confirmed small-bowel obstruction, which was treated by adhesiolysis. During the hospital stay, a central venous catheter was implanted through the left subclavian vein for intravenous therapies (Fig. 1) and left in place for 13 days. The patient was discharged with no complication and free from symptoms. Three months later, he presented with atypical subacute chest pain. No significant electrocardiographic changes were seen expect frequent extrasystoles. Cardiac enzymes were at the normal level. Echocardiography evidenced normal left-ventricle function and showed multiple vascular dilatations on

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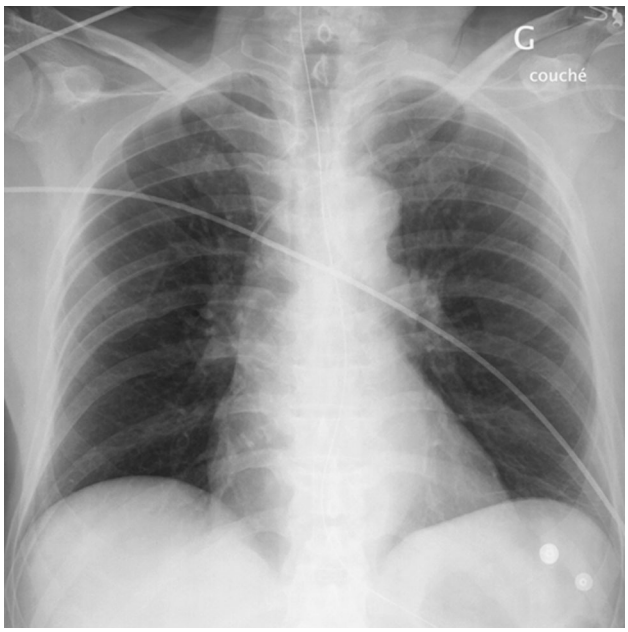

Fig. 1 Chest X-ray that shows the left subclavian central venous catheter. Notice that the catheter tip was at the junction between the innominate vein and the superior vena cava

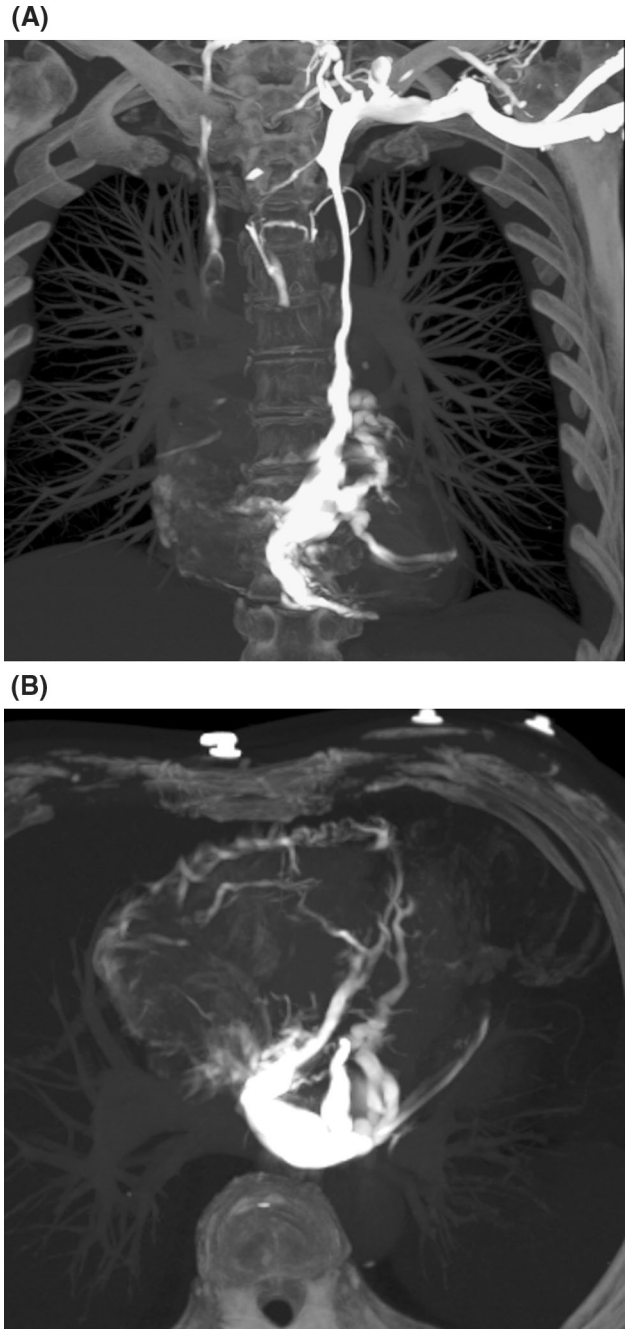

Fig. 2 CT phlebography. **A** Thin-slab reconstruction using the maximum intensity projection algorithm in the coronal view showing occlusion of the IV and patent left superior vena cava connected to the coronary sinus, which is connected to right atrium. Notice the high-grade ostial stenosis of the coronary sinus. **B** Axial view reconstruction using the same algorithm shows dilatation of the coronary sinus and its affluents

computed tomography (CT) phlebography (Fig. 2) that were identified as multiple dilatations of the coronary sinus and its myocardial affluent veins, including the septal veins. These findings had received a first diagnosis of vascular malformation, and the patient was referred to us to evaluate potential endovascular management. Actually the initial CT phlebography as well as further investigations, including cardiac magnetic resonance (MR) imaging, showed occlusion of the IV, PLSVC, and high-grade stenosis of the coronary sinus ostium. The PLSVC drained into the coronary sinus blood mainly from the left upper extremity. The ostial stenosis of the coronary sinus corresponded to CSA associated with PLSVC. Induced occlusion of the IV, probably after central venous catheter placement/removal, resulted in progressive dilatation of the myocardial vein.

To restore pre-existing venous drainage in this rare configuration of congenital anomaly (i.e., ACS) and anatomic variant (i.e., PLSVC), a mechanical guidewire-based recanalization procedure was performed. With the patient under local anesthesia and administered 5,000 IU of heparin, recanalization was first unsuccessfully attempted by way of the left brachial approach. Then a combined approach access, brachial and right femoral, as previously described by Qanadli et al. [1], provided successful retrograde recanalization of the IV with a 0.018-in. nitinol Pointer guidewire (PBN Medicals, Stenlose, Denmark). A 260-cm length hydrophilic 0.035-in. Radiofocus wire (Terumo European N.V., Leuven, Belgium) was then placed through a 4F hydrophilic catheter (Terumo European N.V.). After unsatisfactory balloon angioplasty with Fox Plus balloons (6/40 mm and then 9/40 mm [Abbott

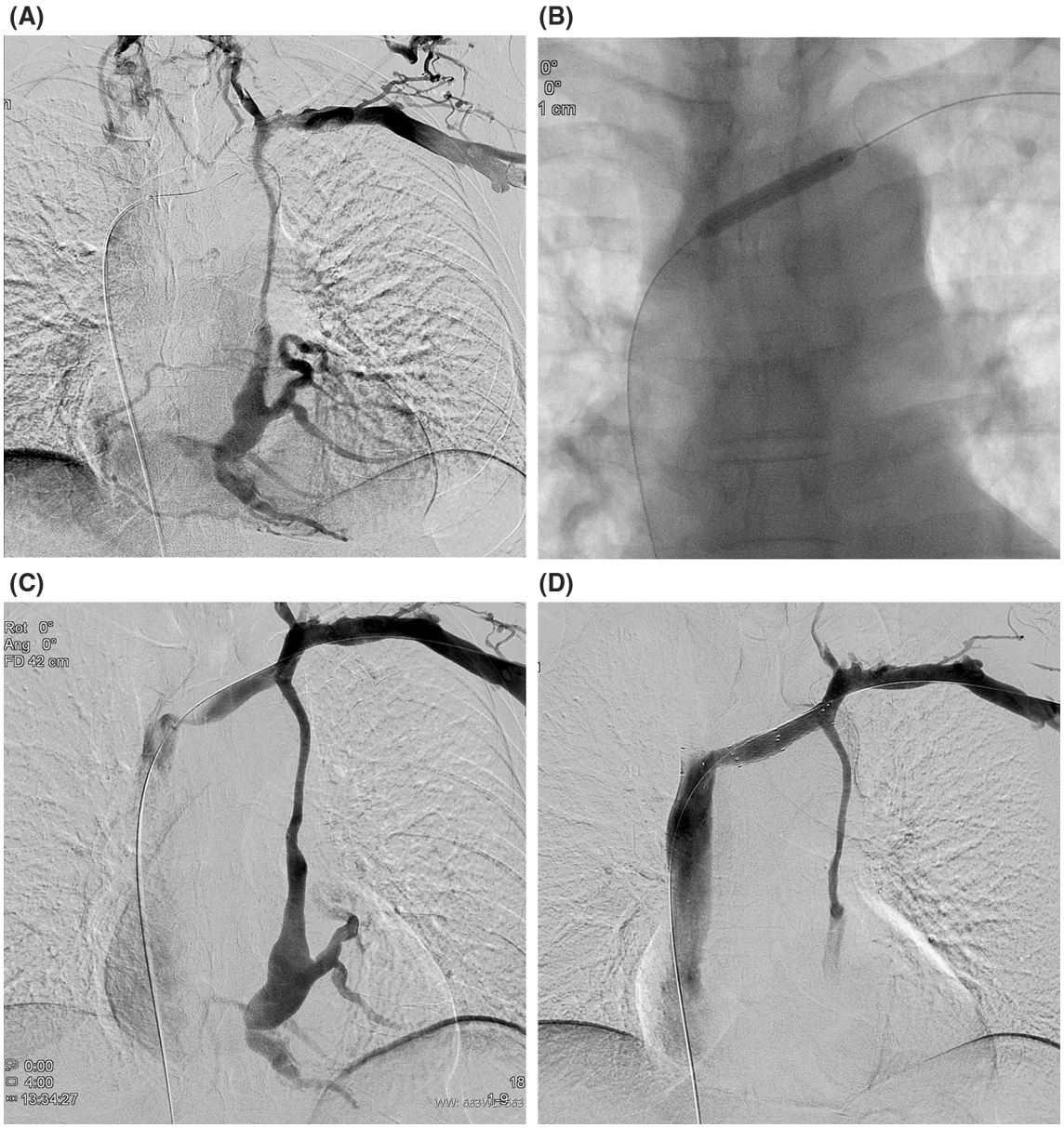

Fig. 3 Retrograde endovascular recanalization procedure. **A** The high-grade stenosis of the innominate vein. **B** Balloon angioplasty after successful recanalization. **C** Angiogram after 6- and 9-mm balloon angioplasty. Significant residual stenosis was seen. **D** Final angiogram after IV stenting. Immediate venous drainage was preferentially to the IV with hesitant antegrade flow in the coronary sinus and the superior vena cava

Laboratories Vascular Enterprises, Beringen, Switzerland]), two self-expandable stents (Sinus XL 12/40 and 14/40 mm [Optimed, Ettlingen, Germany]) were inserted into the IV (Fig. 3), thus restoring flow in the IV and decreasing accessory left-side venous drainage in the PLSVC. The patient was discharged as asymptomatic with aspirin and clopidogrel for 3 months. Eleven months later, he experienced new-onset related to in-stent restenosis, which was successfully treated by balloon angioplasty. The patient was asymptomatic during subsequent 32-month follow-up.

Discussion

ACS is a rare anomaly of the coronary vasculature previously reported as case reports and usually discovered at autopsy. A classification of coronary sinus abnormalities was suggested by Mantini et al. [2], namely, enlarged coronary sinus, absent coronary sinus, atresia of right atrial ostium of the coronary sinus, and hypoplasia of the coronary sinus. Anatomically, ACS consists of membranous occlusion with or without segmental interruption of the coronary sinus [3]. This defect is an intrinsically benign

anomaly because it is usually associated with unroofed coronary sinus and/or PLSVC. PLSVC is the most common variant of systemic venous drainage and is found in 0.3–0.5 % of the general population and ≤0 % of patients with a congenital cardiac anomaly [4]. In 70–90 % of patients with PLSVC, there is also a right superior vena cava, which may or may not communicate with the PLSVC [4]. The PLSVC commonly drains into the coronary sinus. It is the most common thoracic venous anomaly and is usually asymptomatic. While in the typical form it is often hemodynamically nonsignificant, its discovery may have clinical significance nonetheless.

When a functional LSVC is associated with the coronary sinus atresia, it can drain blood retrogradely from the coronary system by way of the IV vein to the right SVC. The PLSVC is the only vessel draining the coronary sinus branches in half of cases [5]. Subsequently, the coronary venous blood still physiologically drains to the right atrium without hemodynamic obstruction or shunt. However, any condition that could preclude interrupting coronary sinus drainage might lead to coronary venous hypertension with subsequent myocardial congestion and even death [5, 6].

Venous thrombosis is a frequent complication of intravascular central catheters. Once the device has been pulled, most of the residual venous thrombosis will resolve spontaneously; however, under unfortunate circumstances proximal inflow congestion can result [7]. In our patient, IV occlusion persisted after central catheter removal. However, no significant typical symptoms of central thoracic vein occlusion were observed, and the patient complained of atypical chest pain. Considering the clinical condition before central venous catheter insertion, we hypothesized that symptoms were related to myocardial vein congestion caused by the IV occlusion. Then we planned to recanalize the IV to restore, at least partially, the coronary sinus drainage by the LPSVC to the right atrium as it existed physiologically before the onset. The performed percutaneous procedure, with a successful clinical long-term result, proved this hypothesis. In our knowledge, such a clinical presentation, as well as its endovascular management, has never been reported.

Clinical implications with respect to vascular access and arrhythmia management should be well understood to prevent complications in asymptomatic patients. CT phlebography [8], as well an MR angiography, has proven extremely helpful in characterizing vascular anomalies as seen in our case and allowed us to establish the correct diagnosis and plan appropriate treatment.

In conclusion, being familiar with such congenital anomalies could help avoid complications during endovascular maneuvers, specifically those that could induce venous thrombosis, such as placement of central catheters, pacemakers, and defibrillators leads. Integrating potential clinical expressions of these anomalies helps to guide the appropriate treatment. Percutaneous endovascular interventions are feasible and safe approaches to manage a patient with induced complications.

Conflict of interest The authors declare that they have no conflicts of interest.

References

1. Qanadli SD, El Hajjam M, Mignon F, de Kerviler E, Rocha P, Barré O et al (1999) Subacute and chronic benign superior vena cava obstructions: endovascular treatment with self-expanding metallic stents. Am J Roentgenol 173:159–164
2. Mantini E, Grondin CM, Lillehei CW, Edwards JE (1966) Congenital anomalies involving the coronary sinus. Circulation 33:317–327
3. Santoscoy R, Walters HL 3rd, Ross RD, Lyons JM, Hakimi M (1996) Coronary sinus ostial atresia with persistent left superior vena cava. Ann Thorac Surg 61:879–882
4. Biffi M, Bertini M, Ziacchi M, Martignani C, Valzania C, Diemberger I et al (2009) Clinical implications of left superior vena cava persistence in candidates for pacemaker or cardioverter-defibrillator implantation. Heart Vessels 24:142–146
5. Jha NK, Gogna A, Tan TH, Wong KY, Shankar S (2003) Atresia of coronary sinus ostium with retrograde drainage via persistent left superior vena cava. Ann Thorac Surg 27(6):2091–2092
6. Yokota M, Kyoku I, Kitano M, Shimada M, Mizuhara H, Sakamoto K et al (1989) Atresia of the coronary sinus orifice. Fatal outcome after intraoperative division of the drainage left superior vena cava. J Thorac Cardiovasc Surg 98:30–32
7. Teichgraber UK, Gebauer B, Benter T, Wagner HJ (2003) Central venous access catheters: radiological management of complications. Cardiovasc Intervent Radiol 26:321–333
8. Qanadli SD, El Hajjam M, Bruckert F, Judet O, Barré O, Chagnon S et al (1999) Helical CT phlebography of the superior vena cava: diagnosis and evaluation of venous obstruction. Am J Roentgenol 172:1327–1333