

Percutaneous Intervention of a Persistent Left Superior Vena Cava Draining Into Left Pulmonary Vein and Coarctation of the Aorta

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Abstract

We describe a 54-year-old male with history of type II DM, hypertension and dyslipidemia during admission for bronchopneumonia discovered to have coarctation of the aorta and a persistent left superior vena cava (PLSVC) draining into the left atrium through the left superior pulmonary vein. The latter was thought to contribute to a transient ischemic attack and an episode of chest pain resulting in ST-segment elevation in the inferior leads. He was treated with coarctation stenting and percutaneous exclusion of the PLSVC with a vascular plug.

Keywords: Percutaneous intervention; Coarctation of the aorta; Persistent left superior vena cava; Amplatzer vascular plug II

Introduction

Persistent left superior vena cava (PLSVC) is a relatively common anatomical variant, with an incidence of 0.3% in autopsy studies [1]. In patients with congenital heart defects, a PLSVC is seen in up to 10% of patients; additional cardiac defects include atrial septal defect, bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia, cortriatriatum, tetralogy of Fallot, and heterotaxy syndrome [1-4]. The PLSVC usually drains into the right atrium via the coronary sinus. In the rare case of coronary sinus ostial atresia, the LSVC drains the coronary venous blood flow from the coronary sinus to the

systemic venous circulation. These anomalies cause no shunts [5-9]. However, in 10-20% of cases, it drains to the left atrium either via unroofed coronary sinus, or in a straight line fashion into the roof of the left atrium, or the left superior pulmonary vein [10]. It is in this kind of abnormal connection where there is shunting and occlusion is required [11]. We describe a case of combined coarctation of the aorta and PLSVC draining into the left pulmonary vein treated percutaneously.

Case Report

A 54-year-old male patient with long standing history of type II DM, dyslipidemia and hypertension, with medical therapy of bisoprolol 10 mg daily, valsartan 160 mg daily, HCTZ 25 mg daily, atorvastatin 20 mg nocturnally, insulin and metformin was admitted to a peripheral hospital with bronchopneumonia requiring high dependency unit admission. This was complicated by a transient ischemic attack. Brain computed tomography (CT) was normal. On the second day of admission, he had a significant episode of chest pain. Electrocardiogram (ECG) showed transient ST-segment elevation with spontaneous resolution in the inferior lead. There was mild troponin leak. He was later referred to our center for further workup. His physical exam revealed oxygen saturation at 98% on room air, pulse 70 bpm, BP both upper limb 155/65 mm Hg, and BP lower limb 105/45 mm Hg. There was a brachio-femoral delay. Apex was mildly displaced; he had S1+ S2+ S4 and a systolic murmur at the tip of left scapula; no clicks were appreciated on auscultation.

ECG showed sinus rhythm and left ventricular hypertrophy (LVH) with strain pattern. A 24-h Holter came back with normal sinus rhythm, no atrial fibrillation, no heart blocks or arrhythmia. Transthoracic echocardiogram revealed concentric LVH, borderline left ventricular function of 45-50%, mild to moderate aortic regurgitation, bicuspid aortic valve and a coarctation of the aorta with a 70 mm Hg gradient across it. Brain MRI demonstrated lacunar infarcts and no AV-malformations. Both carotid arteries were free of atherosclerosis on Doppler. Coronary CT angiogram not only demonstrated the tight coarctation but also showed a rare anomaly of a PLSVC draining into the left atrium (Fig. 1). Anomalous pulmonary drainage and pulmonary arteriovenous malformation were excluded. The approach to management of this patient was discussed in a

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Figure 1. Volume rendered 3D image demonstrating the PLSVC draining into the left superior pulmonary vein then into the left atrium and the tight coarctation.

heart team meeting. It was decided to proceed percutaneously.

The patient was brought to the catheterization laboratory. Coronary angiography showed normal coronaries. He did have a significant coarctation with a pressure gradient of 40 mm Hg. Angiographic confirmation of the PLSVC draining into the left superior pulmonary vein was confirmed. A 40 mm CP stent mounted on a 20 × 45 mm BIB balloon was deployed successfully (Fig. 2), guided by fluoroscopy and overdrive pacing. There was complete abolition of the gradient. At a later

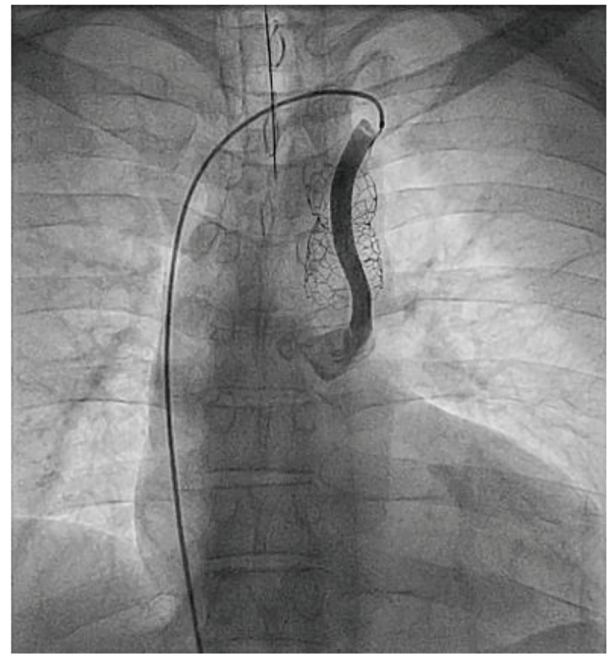


Figure 3. The PLSVC draining into the left superior pulmonary vein then into the left atrium.

date, the patient was brought for PLSVC exclusion therapy. Selective angiography of the PLSVC was performed to delineate vessel course and venous drainage (Fig. 3). Using a marker pigtail measurement of the vessel was obtained. The PLSVC measured 9 mm proximally, 10.5 mm at its mid-point and 8 mm near the entrance to the left upper pulmonary vein. The PLSVC was then temporarily occluded with an 8 mm peripheral balloon to make sure that the patient would tolerate closure of the vessel. There was no significant change in the patient’s hemodynamic measurements, and the decision was made to proceed with transcatheter occlusion of the PLSVC which was achieved using a 12 mm Amplatzer vascular plug II (AGA Medical Corporation, Golden Valley, MN) (Fig. 4).

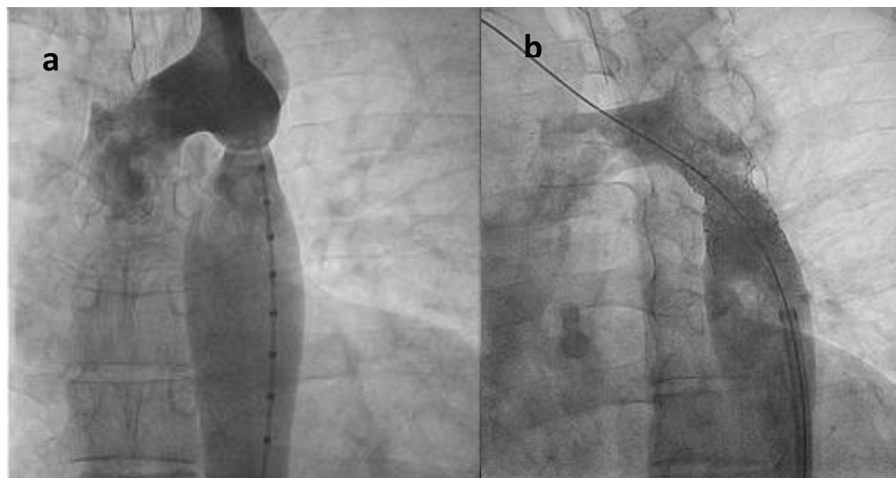


Figure 2. (a) Pre-coarctation and (b) post-coarctation stenting.

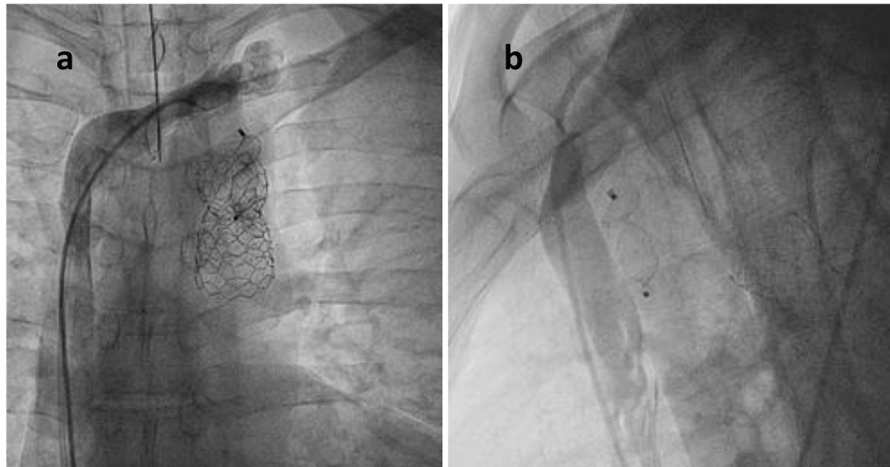


Figure 4. The successful closure of the PLSVC: (a) PA view; (b) lateral view.

Discussion

In the vast majority of patients, a PLSVC drains via an intact coronary sinus to the right atrium, resulting in normal systemic venous return and no clinical sequelae. However, in 8% of patients with a PLSVC, the LSCV drains to the left atrium directly, through an unroofed coronary sinus or pulmonary veins [1, 12-17].

A hemodynamically significant right-to-left shunt may form. Such a shunt may cause significant systemic desaturation and place the patient at risk for a paradoxical embolic stroke, cerebrovascular accident and intracranial abscess [10, 18, 19]. Surgical repair of the PLSVC may be offered if a patient suffers complications due to a right-to-left shunt. The surgical techniques used to repair a PLSVC are well described, and are specific to the patient's particular anatomical defect [19-23]. More recently successful percutaneous closure of PLSVC using the Amplatzer vascular plug device has been reported [4, 10, 24].

Our patient was unique in the sense that he had two major anomalies that have been undetected for a long time though he was not previously well investigated. He however underwent a coarctation stenting and a percutaneous exclusion of the PLSVC with an Amplatzer vascular plug II successfully.

Conclusion

The occurrence of the combination of coarctation and PLSVC draining into the left atrium is extremely rare, both carrying significant morbidity and mortality. Our patient had coarctation of the aorta and a PLSVC that was discovered after our evaluation. The PLSVC draining to the left superior pulmonary vein was thought to precipitate transient ischemic stroke and the episode of chest pain through the paradoxical air embolism from a left hand placed cannula. This case highlights that patients with congenital heart defects may suffer from other associated defects. Percutaneous techniques were very successful in treating both anomalies.

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