Persistent left superior vena cava: An unusual cause of curable pulmonary hypertension

Keywords: Left-to-right-shunt; Persistent left superior vena cava; Pulmonary hypertension; Congenital heart disease; Cardiac CT

Dear Editor,

Persistent left superior vena cava (PLSVC) is the most common venous anomaly of the thorax. Several variations have been described [1,2]. Usually PLSVC drains normally into the right atrium via the coronary sinus. We report a rare case of PLSVC connecting to the left atrium that induced extra-cardiac left-to-right shunt and symptomatic pulmonary arterial hypertension.

A 66-year-old woman was referred for cardiac computed tomography (CT) to investigate an abnormal vessel connecting to the left atrium, identified during catheter ablation of an atrial fibrillation. At the age of 26, the patient underwent surgery for aortic coarctation. She was treated for arterial hypertension. She presented with grade III dyspnea according to the New York Heart Association and exercise-induced tachycardia. CT showed PLSVC connecting to the left atrium between the left appendage and the left superior pulmonary vein. Innominate vein was present. Coronary arteries arose normally from the aorta. There was no septal defect and the coronary sinus was normal. Both atria were dilated with a surface of 35 cm², as well as the right ventricle (end-diastolic diameter of 50 mm). The pulmonary artery was enlarged (43 mm) and bilateral mosaic perfusion was found in the lungs (Fig. 1). A jet of contrast medium flowing from the innominate vein to the right superior vena cava suggested a left-to-right shunt (Fig. 2). Contrast echocardiography revealed pulmonary arterial hypertension. This was confirmed by right heart catheterization that disclosed a mean and a maximum pulmonary arterial pressure of 42 and 70 mmHg, respectively. Right heart blood flow was increased (cardiac index: 6.1 L/min/m²). No other cause of pulmonary arterial hypertension was found, so that surgery was decided. Peroperative findings confirmed the anatomic configuration and the extracardiac left-to-right shunt (Fig. 3). Arterial blood was flowing through the PLSVC cephalad to the left cerebrobranchial vein. Tourniquet occlusion of the PLSVC at its upper extremity resulted in correction of the blood flow. When the lower end of the PLSVC was occluded, no bulging of the coronary sinus appeared so there was no evidence of an associated coronary sinus atresia. Then, definite occlusion of the abnormal vessel was performed. At six months follow-up, the patient had a grade II dyspnea. Cardiac magnetic resonance (MR) imaging showed no residual shunt; Qp/Qs ratio was balanced. The caliber of the left brachiocephalic vein dropped from 18 to 8 mm, right arterial surface normalized at 21 mm², cardiac blood flow was decreased at 2.9 L/min/m². Echocardiography showed decreased pulmonary arterial hypertension with a maximum pulmonary arterial hypertension of 45 mmHg.

To our knowledge, this is the second reported case of left-to-right shunt caused by a PLSVC without coronary sinus atresia [3]. In addition, this is the first reported case in which PLSVC induced a left-to-right shunt complicated of symptomatic pulmonary arterial hypertension. Our patient had a history of aortic coarctation, which is associated with PLSVC [4]. However, at the time of surgery, no CT or MR imaging has been performed and the diagnosis of associated PLSVC was overlooked [5]. Most common PLSVC variations include, connection to the coronary sinus, with or without coronary sinus ostium atresia or unroofed coronary sinus, and connection to the left appendage with arterial septal defect [1]. Rare cases of connection to the roof of the left atrium associated with coronary sinus atresia and right-to-left shunt have been reported. The latter may be complicated by paradoxical embolism, cyanosis, catheterisation complication and iatrogenic myocardial venous infarction in case of associated coronary sinus ostium atresia. PLSVC connected to the left atrium usually induces extracardiac right-to-left shunt.
The authors declare that they have no competing interest.

References