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GAVIN PUBLISHERS

Journal of Surgery

da Silveira LMV, et al. J Surg: JSUR-1182.

Case Report

DOI: 10.29011/2575-9760. 001182

Pulmonary Hypertension Secondary to Reverse Flow in Left Superior Vena Cava Presenting as Late Evolution of a Pulmonary Anomalous Partial Venous Communication Surgical Correction

Lucas Molinari Veloso da Silveira[1*], Ana Paula Tagliari[2], Bruno S. Matte[3], Adriano Nunes Kochi[4], Murilo Foppa[5]

[1]Faculty of Medicine, Federal University of Rio Grande do Sul (UFRGS), Porto Alegre, Brazil

[2]Department of Cardiovascular Surgery, Hospital of Clinics of Porto Alegre (HCPA), UFRGS, Porto Alegre, Brazil

[3]Department of Cardiology, HCPA, UFRGS, Porto Alegre, Brazil

[4]Cardiologist and Electrophysiologist, Department of Cardiology, HCPA, UFRGS, Porto Alegre, Brazil

[5]Cardiologist and Echocardiographist, Department of Cardiology, HCPA, UFRGS, Porto Alegre, Brazil

[*]**Corresponding author:** Lucas Molinari Veloso da Silveira, Hospital of Clinics of Porto Alegre, Cardiovascular Surgery Service, Rua Ramiro Barcelos, 2,350, CEP: 90.035-903, Porto Alegre - RS, Brazil. Tel: : +55-53999991595; Fax: +55-5133598001; Email: lucasmolinari93@gmail.com

Citation: da Silveira LMV, Tagliari AP, Matte BS, Kochi AN, Foppa M (2018) Pulmonary Hypertension Secondary to Reverse Flow in Left Superior Vena Cava Presenting as Late Evolution of a Pulmonary Anomalous Partial Venous Communication Surgical Correction. J Surg: JSUR-1182. DOI: 10.29011/2575-9760. 001182

Received Date: 03 December, 2018; **Accepted Date:** 12 December, 2018; **Published Date:** 18 December, 2018

Abstract

Partial anomalous pulmonary venous connection is a congenital abnormality that affects less than 1% of the population. Treatment of this malformation is usually surgical, and it has an excellent result. However, the presence of another vascular abnormality, for example, a persistent left superior vena cava, may be a complicating factor to corrective surgeries. In this case, we report an unusual evolution of a patient submitted to surgical correction of partial anomalous pulmonary venous connection. This patient presented with severe pulmonary hypertension and left to right shunt due to reverse flow in a persistent left superior vena cava, which was not diagnosed during the previous cardiac surgeries.

Keywords: Congenital Heart Disease; Pulmonary Hypertension; Pulmonary Veins

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a congenital abnormality characterized by fusion failure of the embryologic pulmonary venous system with the left atrium. This entity is a rare congenital anomaly described in 0.4 to 0.7% of autopsies [1]. The definitive management of PAPVC consists in surgical separation of the pulmonary venous system from the systemic venous system, through redirection of an aberrant pulmonary venous drain into the left atrium [2]. During surgical repair of this pathology, about 16% of the patients present another congenital vascular anomaly, for example, a persistent left superior vena cava (PLSVC) [3]. PLSVC is the most common congenital venous anomaly, with an incidence of approximately 0.3% to 0.5% in the general population, and up to 10% in patients with other concomitant congenital heart diseases [3].This case aims to report an unusual evolution after PAPVC surgical correction in a patient who presented concomitantly PLSVC.

Patient Characterization and Description of The Technique Employed

A 51-year-old female patient presented to a cardiac surgery outpatient clinic with progressive dyspnea for about 5 years and no other complaints. At physical examination, there was a systolic heart murmur 3+/6+ at the left lower sternal border and apex, and no other relevant findings. This patient had already undergone two previous cardiac surgeries. First when she was 6 years-old (medical report summary indicating an interatrial communication closure), and the second at 39 years old, when she presented a right lower pulmonary blood flow drainage into the coronary sinus. The surgery was performed by unroofing the coronary sinus into the left atrium and occluding the coronary sinus ostium in the right

atrium, in order to redirect the right pulmonary blood flow to the left atrium. Soon after discharge of the second surgery, the patient had syncope due to an atrioventricular block and was treated with a permanent pacemaker implant.

At the time of the current outpatient evaluation, she presented a transesophageal echocardiogram (TEE) demonstrating right heart overload (right ventricle diameter = 4.9cm), pulmonary hypertension (trans-tricuspid gradient = 73 mmHg) and left-to-right shunt (Qp/Qs = 2.2). The anatomic source of shunting was not readily identified, even with agitated saline injection in a peripheral vein. A dilated coronary sinus and PLSVC were also observed during TEE.

A chest angiotomography demonstrated right heart and common pulmonary artery dilatation (diameter = 36 mm), compatible with pulmonary hypertension. It also demonstrated interventricular septum rectification, and disclosed a PLSVC, receiving flow from the hemiazygous vein and draining into the coronary sinus (Figure 1). In this exam was possible to detect signs of elevated coronary sinus pressure and hemiazygous vein dilatation. It was also noted collateral vessels draining from LSVC to right superior vena cava (Video 1). Magnetic resonance imaging was not performed since the patient had a non-conditional pacemaker.

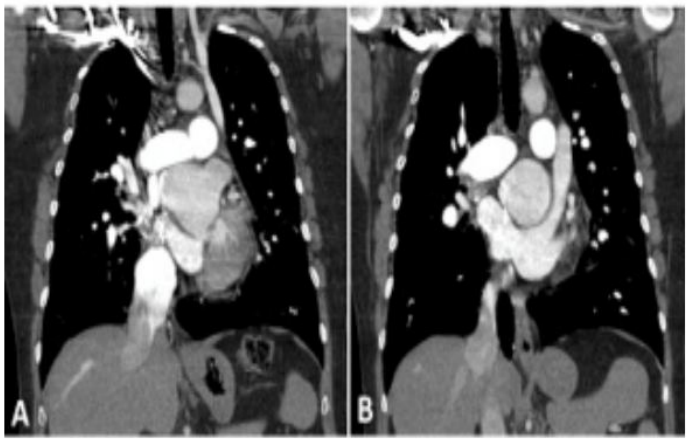

Figure 1: (A): Contrast-enhanced computed tomography coronal image demonstrating the presence of a persistent left superior vena cava. **(B):** Diversion of right pulmonary drainage to the left atrium through the coronary sinus

Right cardiac catheterization was performed and evidentiated a mean pulmonary arterial pressure of the 37 mmHg and a left to right shunt (Qp/Qs = 2.3). Venography performed through contrast injection via the basilic vein disclosed a persistent LSVC connecting to the coronary sinus, observing a reverse flow from the coronary sinus to the LSVC and hemiazygos vein (Figure 2). Oximetry was collected from the lower portion of the LSVC and demonstrated 97.7% oxygen saturation, confirming the presence of a shunt with retrograde flow in the LSVC. Considering the high surgical risk and the adequate symptomatic response to sildenafil, it was opted to maintain the patient in pharmacological management. This case demonstrates an unusual evolution after a PAPVC surgical correction, attributed to the presence of another concomitant congenital vascular malformation not identified during the pre-operative investigation.

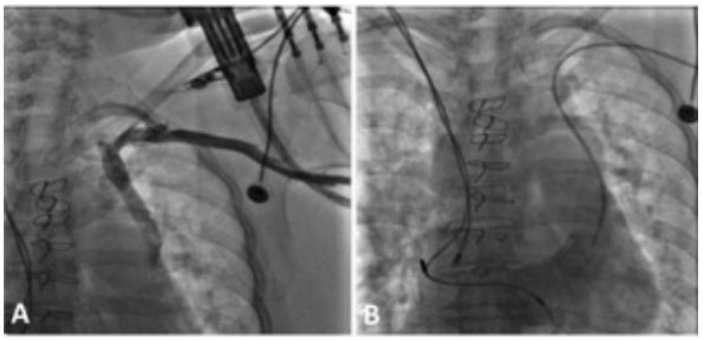

Figure 2: (A): Coronary cineangiography demonstrating the presence of a persistent left superior vena cava. **(B)**: The drainage of the left superior vena cava into the coronary sinus.

Discussion

PAPVC is a congenital abnormality characterized by fusion failure of the embryologic pulmonary venous system with the left atrium [1]. This results in one to three pulmonary veins draining to structures other than the left atrium. Considering the rule of proximity in embryology: right pulmonary veins usually drain to vena cava or right atrium; while left pulmonary veins drain to coronary sinus, left superior vena cava or innominate vein [4]. Other congenital vascular abnormality is the PLSVC. The development of major systemic veins results from growth and involutions of different venous drainages systems during fetal development [5]. The closure failure of the left anterior cardinal vein will result in a PLSVC, which usually drains the left subclavian and jugular veins into the right atrium via an enlarged coronary sinus [6]. Data about the concomitance of these two abnormalities are scarce and the true incidence of this association remains unknown. However, a case series of patients submitted to surgical correction of PAPVC showed that 15.7% of them presented a PLSVC [3].

In the reported case the patient was submitted to a PAPVC surgical correction according to the traditional technique, in which pulmonary veins are routed to the left atrium by unroofing the coronary sinus, and the ostium of the coronary sinus is closed [7]. However, this correction was prejudiced by the misdiagnosis of a PLSVC during pre and intraoperative moments. The misdiagnosis, although uncommon and avoidable, can occur if the presence of a PLSVC is not considered in the case. Data from a selected

pediatric population submitted to cardiac surgeries for congenital heart disease revealed that in the preoperative period almost 20% of the PLSVC diagnosis could be missed by echocardiography-only investigation [5], which is the first-line imaging tool for PLSVC diagnosis [8]. Lu S. et al. comment, for example, a case of partial anomalous pulmonary venous connection misdiagnosed as primary pulmonary arterial hypertension because the two high right superior pulmonary veins drained into the superior vena cava and limited the shunt of patent foramen ovale [9].

Abnormal venous connections can be identified by computed tomography or magnetic cardiac resonance or, even so, intraoperatively. The identification of PLSVC during surgery is made by the dissection of the thymic fat and visualization of the left innominate vein. However, in adult patients referred to surgery, this abnormality may be missed if not carefully and properly researched. The presence of a PLSVC may be a benign finding with no clinical repercussions in patients with no other anomaly of the anatomy, but, in those patients that with unroofed coronary sinus, it can be associated with arterial desaturation and paradoxical systemic embolism [5]. In the presented case, due to surgical correction of PAPVC, our patient developed an anatomy similar to those patients who congenitally present an unroofed coronary sinus. Besides, the surgery performed lead the drainage of the PLSVC to the left atrium, which traditionally cause a right-to-left shunt [6]. Nevertheless, in our patient a left-to-right shunt was disclosed during investigations. This fact, as in the case reported by Chen, et al. [10], occurred because of an elevated left atrium pressure, which resulted in a reverse flow (caudal-to-cranial) on the PLSVC. A similar case, however in an infant, was reported by Quarti A. et al. In this description, disconnected left superior vena cava draining into the left atrium through the left appendage, associated with atrial septal defect and right pulmonary veins anomalous connection was successfully corrected by rerouting the pulmonary venous return into the left atrium and transposing the left vena cava on the right appendage [11].

In this case we report an unusual evolution after a PAPVC surgical correction in a patient presenting severe pulmonary hypertension and a left-to-right shunt, which occurred due to the presence of a concomitant vascular anomaly, which was not diagnosed neither in the preoperative period, nor in the intraoperative period.

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