Severe pulmonary arterial hypertension in an adult patient with total anomalous pulmonary venous connection operated in infancy

Hipertensiune arterială pulmonară severă la un pacient adult cu o anomalie totală de conectare venoasă pulmonară operată în copilărie

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Abstract

The goal of total anomalous pulmonary venous connection repair is to obtain an unobstructed communication between the pulmonary veins and the left atrium and removing intracardiac shunting. However, pulmonary venous obstruction or stenosis may be seen in 5-10% of patients, is usually evident in the first 6 months following surgery and may lead to pulmonary congestion, pulmonary arterial hypertension, and late mortality. In such cases, early intervention may be indicated before irreversible secondary changes occur. We present the case and the therapeutic approach of an adolescent patient with total anomalous pulmonary venous drainage to the superior vena cava operated in infancy who developed pulmonary venous obstruction and secondary severe pulmonary arterial hypertension.

Keywords: ***Total anomalous pulmonary venous connection; pulmonary venous obstruction; surgical repair; pulmonary arterial hypertension***

Rezumat

Scopul corectării chirurgicale a anomaliei totale de conectare venoasă pulmonară este obținerea unei comunicări neobstruate între venele pulmonare și atriul stâng și înlăturarea șuntului intracardiac. Cu toate acestea, obstruarea venelor pulmonare sau stenoza acestora pot fi observate la 5-10% din pacienți, de obicei în primele 6 luni postoperator; acestea pot duce la stază pulmonară, hipertensiune arterială pulmonară și mortalitate tardivă. În asemenea cazuri, este indicată intervenția precoce, înainte de producerea unor modificări ireversibile. Se prezintă modalitatea de abord terapeutic în cazul unui adolescent cu anomalie totală de drenare venoasă pulmonară în vena cavă superioară, operat la vârsta de sugar, care a dezvoltat ulterior obstrucție venoasă pulmonară și hipertensiune arterială pulmonară secundară severă.

Cuvinte-cheie: ***anomalie totală de drenare venoasă pulmonară; obstrucția venelor pulmonare; corectare chirurgicală; hipertensiune arterială pulmonară***

Introduction

Obstructed pulmonary venous drainage – either in association with total anomalous pulmonary venous drainage, congenital stenosis, or post-repair stenosis – is associated with poor outcome and late mortality[(1)]. Although improved surgical techniques have led to significantly better outcomes, the risk of progressive pulmonary venous obstruction continues to be a clinical problem. In such cases, symptoms are related to the number of stenotic veins as well as the severity of the stenosis. Because post-repair stenosis typically involves fibrotic scar tissue extending from the site of anastomosis, surgical or percutaneous options are usually not feasible or ineffective[(2)].

Case Report

We present the case of a 16-year-old boy with a supracardiac total anomalous pulmonary venous connection (TAPVC) and an atrial septal defect operated at 5 months of age: the left pulmonary venous collector was connected to the left pulmonary veins and the right inferior pulmonary vein was connected to the left atrium using a single patch technique which also closed the atrial septal defect. No surgery was performed on the right superior pulmonary vein which drained high into the superior vena cava.

The patient has now a New York Heart Association functional class III/IV with no chest pain or syncope. Physical examination showed central cyanosis, severe scoliosis, normal heart sounds and murmurs and baseline oxygen saturation of 75% with associated secondary erythrocitosis. 12-lead electrocardiogram evidenced sinus rhythm with right ventricular hypertrophy and the chest radiography showed pulmonary venous congestion, pulmonary infiltrates and severe scoliosis (Figure 1A). Meanwhile, transthoracic echocardiography noticed a normal left ventricle with no mitral or tricuspid regurgitation, no residual interatrial shunt, a dilated and hypertrophic right ventricle and severe pulmonary arterial hypertension. High-resolution computed tomography (CT) (Figure 1B) showed a thin left pulmonary artery, thickening of the pulmonary interstitium and a compensatory growth of the left bronchial circulation with occlusion of the left pulmonary venous drainage. Meanwhile, right heart catheterization evidenced a mean

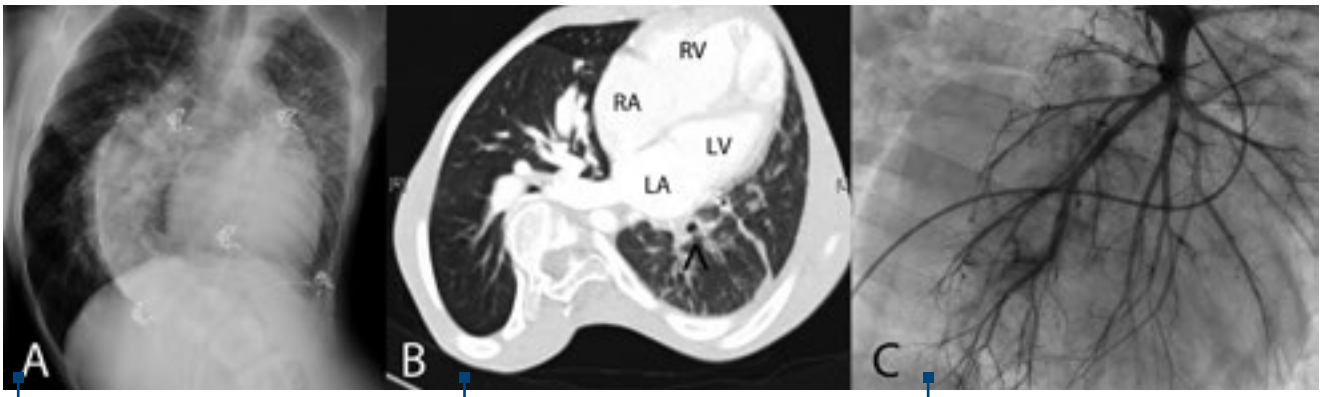

Figure 1A. *Chest radiography showing pulmonary venous congestion, interstitial lesions and pleural changes in the left lung with associated severe scoliosis.*

Figure 1B. *High-resolution computed tomography angiography showing interstitial edema in the left lung and obstruction at the left pulmonary veins level (arrow head).*

Figure 1C. *Selective left pulmonary artery angiogram evidencing a thinned left pulmonary artery. LA: left atrium, RA: right atrium, LV: left ventricle, RV: right ventricle.*

right atrial pressure of 8 mmHg, a right ventricle pressure of 100/4 mmHg, a right arterial pulmonary pressure of 110/65/89 mmHg, a left arterial pressure of 98/78/89 mmHg, a left ventricle pressure of 133/5 mmHg and a mean left pulmonary capillary wedge pressure of 20 mmHg. At the same time, oxygen saturation at the pulmonary artery was 55% and at the aorta was 72%. Meantime, pulmonary angiography (Figure 1C) demonstrated a narrow but smooth left pulmonary artery. The basal six minute walk test, with oxygen administration (3 l/min), showed a low exercise tolerance (168 meters) with severe oxygen desaturation (from 82 to 53%). Meanwhile, the spirometry demonstrated a moderate restrictive pattern (FEV1 1,01 L, FVC 1,23 L, TLC: 2,29 L). Because of his long-standing pulmonary vein occlusion, with suspected irreversible pulmonary artery changes, and severe scoliosis surgical repair of the venous obstruction and lung transplant were contraindicated.

As circulatory alterations provoked by pulmonary venous hypertension share some characteristics with other forms of pulmonary hypertension, bosentan treatment was started at a dose of 62.5 mg every 12 hours during the first month and then increased to 125 mg every 12 hours in association with nasal cannulae oxygen administration at low flow rates (3–4 l/min). Also, due to his poor clinical improvement, sildenafil 20 mg dose every 8 hours was added. Six months later, the patient is able to walk 162 meters in the six minutes walking test , basal saturation with oxygen supply (3 l/min) is 88%, decreasing to 70% after walking, and he is more comfortable despite he often uses wheelchair over medium distances. No side-effects have been seen in relation to his treatment for pulmonary arterial hypertension such as lung oedema.

Discussion

TAPVC is a relatively uncommon congenital heart defect (< 1%). The principle of operative repair is to establish an unobstructed communication between the pulmonary venous confluence and the left atrium. However, in those patients with a TAPVC to the superior vena cava the variants that are difficult to repair include drainage of all of the pulmonary veins directly to the superior vena cava (SVC) without a pulmonary venous confluence, drainage of one or more pulmonary veins to the high SVC and drainage of one or more pulmonary veins to a small right SVC usually in association with a persistent left SVC[3].

Postoperative pulmonary venous obstruction tends to appear in the first 6 months after TAPVC[2] surgery. Nonetheless, it is unknown whether traumatic manipulations of the pulmonary veins at surgery or the abnormal architecture of the pulmonary vasculature typically observed in patients with TAPVC may render these vessels more vulnerable to developing intimal hyperplasia and obstruction[3]. Tachypnea, tachycardia, or cyanosis may indicate pulmonary venous obstruction. However, diagnostic tests may include chest radiography to show pulmonary venous congestion, interstitial lesions, pleural changes, and absence of cardiomegaly; CT angiography to evidence septal lines, centro-acinar opacities and mediastinal lymphadenopathies; magnetic resonance imaging to see the situation of the drainages and pulmonary angiography to demonstrate the venous return after selective injection into both pulmonary arteries. On the contrary, although echocardiography has been used to identify the anomalous connection of the pulmonary venous confluence with the systemic venous system, it does not usually provide adequate assessment of pulmonary vein stenosis. ■

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