Paradoxical arterial hypoxemia in a left-to-right shunt congenital heart disease

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Abstract

The hepatopulmonary syndrome is a rare complication of different types of chronic hepatic diseases with associated portal venous hypertension, resulting in pulmonary vascular dilatation, predominantly in the lower lung fields, and leading to ventilation-perfusion mismatch, arterial hypoxemia and a poor prognosis. We present the case of 42-year-old male patient with an anomalous drainage of the right superior pulmonary vein into the azygos vein and a portal vein cavernomatosis with associated portal venous hypertension who presented severe oxygen desaturation, during exercise, in the context of a hepatopulmonary syndrome.

Keywords: *hepatopulmonary syndrome, hypoxia, portal hypertension, congenital heart disease*

Rezumat

Hipoxemie arterială paradoxală în cadrul unei afecțiuni cardiace congenitale cu șunt stânga-dreapta

Sindromul hepato-pulmonar este o complicație rară a mai multor afecțiuni cronice hepatice care asociază hipertensiune venoasă portală, asociind dilatarea vaselor pulmonare predominant în lobii inferiori și conducând la inegalitatea raporturilor ventilație-perfuzie cu hipoxemie arterială și prognostic rezervat. Prezentăm cazul unui pacient de 42 ani cu un drenaj aberant al venei pulmonare superioare drepte în vena azygos și cavernomatoză venoasă portală asociind hipertensiune venoasă portală, care s-a prezentat pentru desaturare severă în timpul efortului fizic, în contextul unui sindrom hepatopulmonar.

Cuvinte-cheie: *sindrom hepatopulmonar, hipoxemie, hipertensiune portală, afecțiune cardiacă congenitală*

Introduction

Hepatopulmonary syndrome and portopulmonary hypertension occur in a substantial proportion of patients who have advanced liver disease virtually of any aetiology. On the one hand, the hepatopulmonary syndrome results in a defect in arterial oxygenation induced by pulmonary vascular dilatation in the setting of portal venous hypertension[1] and occurs in 25% of the subjects with chronic hepatopathy waiting for a liver transplantation[2]. On the other hand, portopulmonary hypertension occurs in 2-10% of cirrhotic patients due to excessive pulmonary vasoconstriction and leading eventually to right heart failure[3].

Case report

We present the case of a 42-year-old male patient with no cardiovascular risk factors that referred, in the last years, dyspnea on exertion (New York Heart Association functional class II/IV) which occasionally improved with bronchodilator inhalers.

Physical examination showed scoliosis with no cyanosis or murmurs in cardiac auscultation. Laboratory test results were within normal limits. The 12-lead electrocardiogram revealed a sinus rhythm with a right bundle branch block while the transthoracic and transesophageal echocardiogram showed a preserved left and right ventricular function, a normal interatrial septum and a dilatation of the coronary sinus. Magnetic resonance imaging revealed a dilated right ventricle with a preserved ventricular function (telediastolic ventricular volume of 102.6 mL/m^2; telesystolic ventricular volume of 42 mL/m^2, a diastolic diameter of 60 mm and a right ventricular ejection fraction of 48%), a dilation of the pulmonary artery (46 mm) and the coronary sinus (in relation to a left persistent superior vena cava) and an anomalous drainage of the right superior pulmonary vein into the azygos vein.

Cardiac catheterization certified these findings and demonstrated an early filling of the pulmonary veins after the pulmonary arteriography (Figure 1A), an oximetry run between the superior vena cava (95%) and right atrium (84%) and a marked dilatation of the right and left azygos veins (Figure 1B). Pulmonary to systemic blood flow ratio (Qp:Qs) calculated by cardiac catheterization and magnetic resonance imaging showed a result of 1.8. Meanwhile, hemodynamic data (systolic/mean/diastolic preasure) exhibited a right atrium pressure of 3/2/1 mmHg, a right ventricular pressure of 50/5 mmHg, a pulmonary artery pressure of 50/33/18 mmHg and a systemic blood pressure of 128/84/78 mmHg. In turn, the pulmonary output was 8.2 l/min and the aortic output was of 4.4 l/min with a total systemic vascular resistance of 1490.9 dyn·s·cm^{-5} and a total pulmonary vascular resistance of 286.4 dyn·s·cm^{-5}. On the other hand, abdominal computed tomography (CT) and enhanced ultrasound imaging showed a portal vein cavernomatosis with a spontaneous splenorenal shunt. Also, abdominal CT showed a small liver with rounded

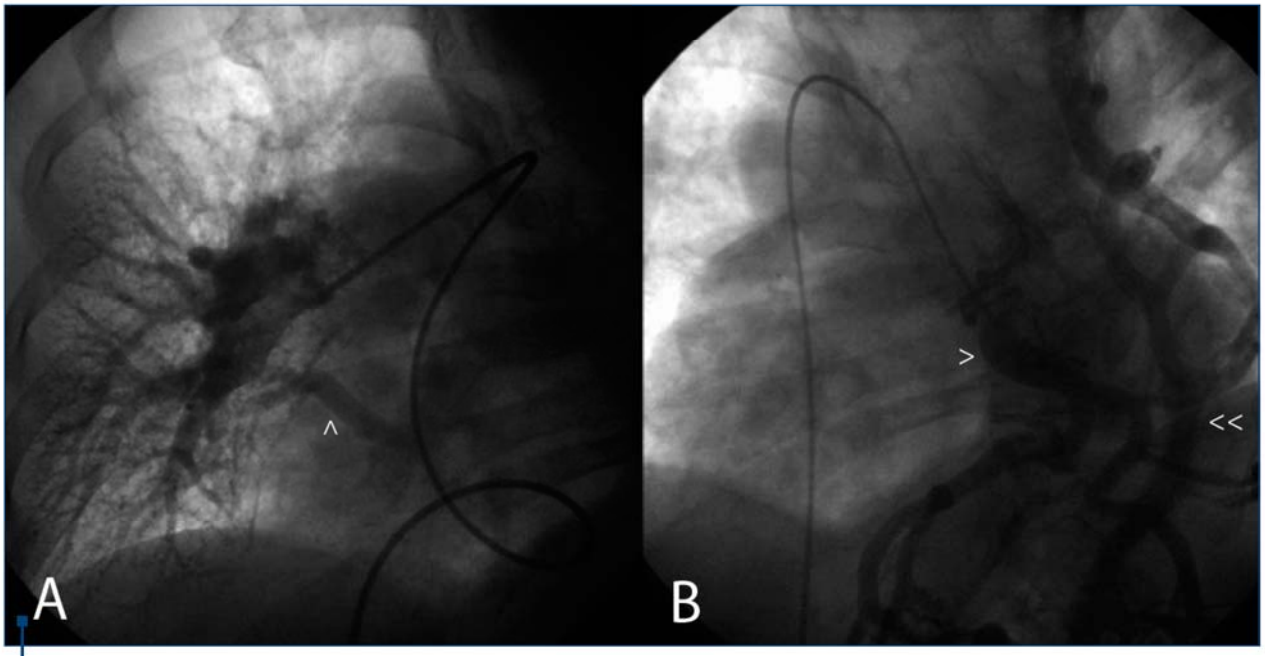

Figure 1. *A: Dilatation of the right pulmonary artery with an early filling of the right inferior pulmonary vein in the context a hepatopulmonary syndrome. Because the pulmonary circulation is a low pressure system it is significantly affected by gravity having the base of the lung a lower resistance than the vessels of the lung apices. This would explain why the right inferior pulmonary vein (arrow head) fills before the pulmonary middle lobe veins. B: Dilatation of the azygos system, right (arrowhead) and left (double arrowhead) azygos veins, draining the left azygos vein into a persistent left superior vena cava.*

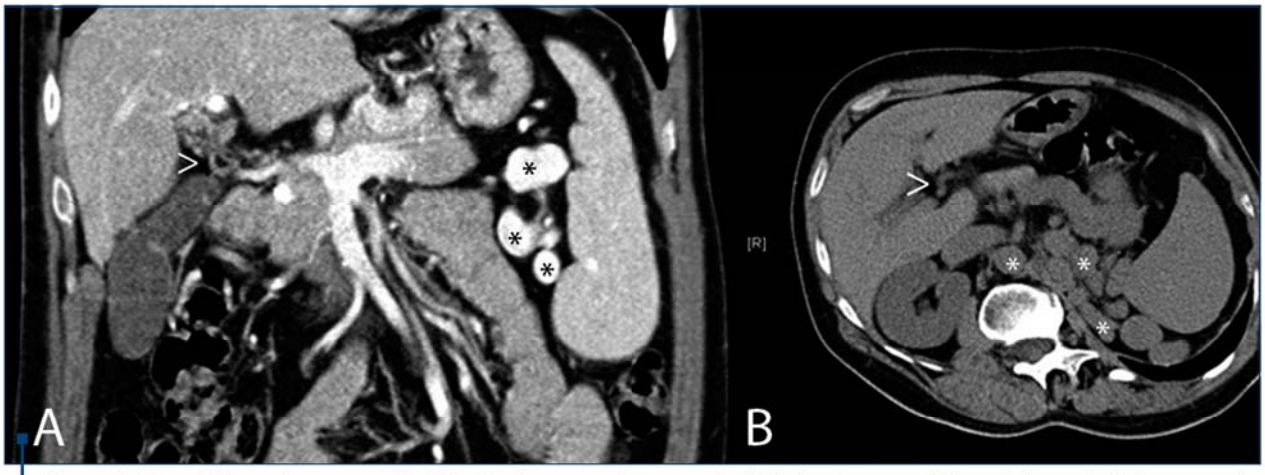

Figure 2. *Coronal (A) and transverse (B) abdominal computed tomography (CT) showing a small liver with rounded edges, portal vein cavernomatosis (arrowhead), spleen and renal varices (asterisks) and mild splenomegaly*

edges, spleen and renal varicose veins and a mild splenomegaly (Figure 2) in relation to portal venous hypertension.

Basal arterial blood gases documented a partial pressure of oxygen (PaO_2) of 72 mmHg, a partial pressure of carbon dioxide ($PaCO_2$) of 34 mmHg, a pH of 7.44, an oxygen saturation of 95% and a bicarbonate (HCO_3) of 23.1 mM/L with a basal alveolar-arterial gradient of 35.2. Pulmonary function testing revealed a moderate degree of restrictive lung disease with a vital capacity (VC) of 56%, a residual volume (RV) of 87%, a total lung capacity (TLC) of 66%, a forced expiratory volume in 1 second (FEV1) of 46%, a forced vital capacity (FVC) of 56%, a FEV1/FVC ratio, also called Tiffeneau index, of 83% and a diffusing capacity (DLCO) of 115%. Exhausting cycle ergospirometry, 8 minutes at 90 watts, showed mild limitation of the aerobic capacity with a peak oxygen consumption of 23 ml/kg per minute (70% of his maximal oxygen consumption (VO_2 max)), a normal ventilatory efficiency and a consumption of 50% of the ventilatory reserve. Oxygen desaturation, determined by pulse oximetry during exercise, dropped from

94% to 83%. In turn, baseline saturation of oxygen reached 100% with high flow concentrations of oxygen (FiO2 of 40%).

Given that our patient had a Qp:Qs less than 2 and the complexity of the surgical procedure, due to his scoliosis and the portal hypertension, a conservative approach was initially decided.

Discussion

Mild hypoxemia is multifactorial and occurs in approximately one third of all patients with chronic liver disease and portal hypertension[4] being different pulmonary vascular mechanisms involved in its occurrence.

The normal route to decompress the hypertensive portal vein is diverting of the portal flow, up to 90%, through portasystemic collaterals, such as the periesophageal, coronary, gastric, azygous or hemiazygous veins, into the superior vena cava. These new routes may favor the development of shunts from the portal to the pulmonary veins and the subsequent arterial hypoxemia because the deoxygenated portal venous blood mixes with the oxygenated pulmonary venous blood. However, portopulmonary shunts do not seem to contribute greatly to the systemic arterial oxygen desaturation due to two reasons: firstly because the oxygen content of the portal vein is high, normally about 50 mm Hg, and secondly because the flow through the portopulmonary anastomosis is low[5].

Another cause that may favor the development of arterial hypoxemia, in patients with liver disease, is the appearance of pulmonary arteriovenous communications. Although these lesions are quite uncommon, they must be taken into account in the differential diagnosis of hypoxemia[6]. In this context, contrast echocardiography has become an excellent tool for evaluation of cardiac and intrapulmonary shunts because microbubble opacification of the left atrium, within three to six cardiac cycles after right-atrial opacification, indicates microbubble passage through an abnormally pulmonary vascular bed[7]. However, this method may fail when there is a patent foramen ovale and may not discern between discrete arteriovenous communications and diffuse precapillary and capillary dilatations[8].

Also, the hepatopulmonary syndrome may favor arterial hypoxemia. Expanded vascular channels between arteries and veins within the lung may allow red cells to move much more quickly through the lung parenchyma, decreasing the time available to load oxygen[9] resulting in hypoxemia which normalizes after receiving 100% oxygen[10] as occurred in our patient.

However, other mechanisms may also influence the degree of arterial hypoxemia during exercise such as the portopulmonary hypertension as also seen in our patient (although pulmonary hypertension may also be due to the volume overload produced by the left-right shunt), the presence of an Eisenmenger syndrome, an expiratory flow limitation, a restrictive lung disease, hyperinflation which may divert an increased percentage of the cardiac output to the respiratory muscles, hypoventilation during exercise, or a limitation of the cardiac output to supply the required flow and oxygen content to the systemic circulation due to a left-to-right shunt.

In conclusion, we present a rare instance of multiple vascular malformations which included a left persistent superior vena cava, an anomalous drainage of the right superior pulmonary vein into the azygos vein and a portal vein cavernomatosis, the latter being able to explain, at least in part, the appearance of arterial oxygen desaturation during exercise in a patient with a left-to-right shunt congenital heart disease. ■

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