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CASE REPORT

### Pulmonary varix: A case report



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#### **KEYWORDS**

Pulmonary veins; Varices; Aneurysm; Pulmonary arterial hypertension Abstract The authors report a case of multiple pulmonary varices, a rare disease characterized by aneurysmatic venous dilatations, which can be present at any age and without gender predominance, occurring in isolation or associated with obstruction of the pulmonary veins. This condition usually manifests as a lung mass with variable clinical consequences. © 2016 Sociedade Portuguesa de Cardiologia. Published by Elsevier España, S.L.U. All rights reserved.

#### PALAVRAS-CHAVE

Veias Pulmonares; Varizes; Aneurisma; Hipertensão pulmonar

#### Variz pulmonar: caso clínico

Resumo Relata-se um caso de varizes pulmonares, na sequência diagnóstica realmente ocorrida. É patologia rara, em que a dilatação venosa aneurismática pode ser isolada ou associar-se a obstruções venosas, ocorrendo em qualquer idade, e sem predomínio de gênero. Frequentemente manifesta-se como massa pulmonar a esclarecer e apresenta repercussão clínica variável. © 2016 Sociedade Portuguesa de Cardiologia. Publicado por Elsevier España, S.L.U. Todos os direitos reservados.

#### Case report

We present the case of a 34-year-old female patient with six pregnancies, four deliveries and two miscarriages (G6P2A4),

with a history of slowly progressive dyspnea over the previous six years, associated with chest oppression and occasional syncope. In the last two pregnancies she noticed worsening of dyspnea during minimum exertion (New York Heart Association class IV) and generalized postpartum edema. On physical examination her heart rate was 90 bpm and blood pressure was 100/60 mmHg and she had normal pulses in all four limbs, precordial thrust in the lower left

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sternal border and a loud second heart sound without murmurs. Lung auscultation revealed no adventitious bruits, and there was no edema, ascites or hepatic splenomegaly.

Laboratory tests showed hematocrit 35%, hemoglobin 11.7 g/dl, GOT 316 U/l, GPT 143 U/l, and TSH 147  $\mu$ IU/l. Arterial blood gas testing on room air revealed pH 7.42, pO<sub>2</sub> 76.8 mmHg, pCO<sub>2</sub> 33.8 mmHg, O<sub>2</sub> sat 94.9%; serological tests for Chagas disease, HIV and hepatitis were negative, as were anti-NF, rheumatoid factor, anti-SCL-70, anti-HIV, anti-HBsAg and anti-HCV tests.

Functional pulmonary testing showed forced expiratory volume in 1 s (FEV1) 2.06 l (75% predicted); forced vital capacity (FVC) 2.39 l (68% predicted); VEF1/FVC 0.86 (109% predicted); total pulmonary capacity 4.65 l (94% predicted); residual volume 2.06 l (predicted 143); diffusing capacity: 1.616 ml/mmHg (75% predicted).

Cardiopulmonary testing revealed peak oxygen uptake 10.8 ml/kg/min; respiratory exchange ratio 1.28; VE/VCO<sub>2</sub> slope 56; and exercise oscillatory ventilation.

The electrocardiogram showed sinus rhythm with right atrial and right ventricular dilatation and hypertrophy and nonspecific repolarization abnormalities.

The chest X-ray demonstrated cardiomegaly with dilation of the right chambers and enlarged pulmonary vessels, more evident in the lung bases.

On echocardiography, there was moderate dilation of the right cavities with diffuse right ventricular hypokinesia, paradoxical septal motion, moderate tricuspid regurgitation, and dilatation of the pulmonary trunk and main branches. Pulmonary hypertension was detected with peak systolic pressure of 87 mmHg.

Following diagnosis of pulmonary hypertension by echocardiography, the patient was referred to our catheterization laboratory for characterization of pulmonary hypertension and study of pulmonary vascular reactivity.

Cardiac catheterization and pulmonary angiography revealed pulmonary artery pressure of 70/40/50 mmHg; right ventricular pressure 70/08/15 mmHg; mean right atrial pressure 15 mmHg with increased V wave (20 mmHg); mean pulmonary capillary wedge pressure in both lungs 10 mmHg; left ventricular pressures 100/02/10 mmHg; and aortic gradient: 100/60/73 mmHg.

Cardiac output was 2.1  $l/m/m^2$  and pulmonary vascular resistance index was 9 Wood units/ $m^2$ ; the patient was unresponsive to vascular reactivity testing administering 100%  $O_2$  and nitric oxide up to 80 parts per million.

A transseptal puncture was then performed followed by selective contrast injection in the pulmonary veins, and a left atrial and venous pressure gradient of 10 mmHg was measured. The pulmonary angiogram (Figures 1 and 2)

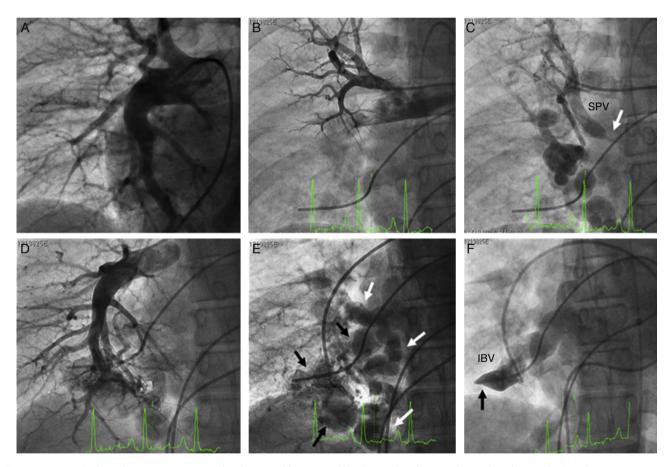


Figure 1 (A) Right pulmonary arteriography showing dilatation of this branch with normal peripheral vessels; (B) selective injection in the superior right pulmonary artery; (C) pulmonary venous return with significant obstruction of the superior vein (arrow) and collateral vessels to the inferior vein with presence of multiple varices; (D) selective injection in the right inferior pulmonary artery; (E) pulmonary venous return showing dilatation and twisting of the central veins of the middle and inferior lobes; (F) complete obstruction of the inferior basilar vein (arrow). IBV: inferior basilar vein; SPV: superior pulmonary vein.

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