

Unusual Case of Pulmonary Hypertension

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Key Words

Angiography · A-V fistula · Laminectomy · Pulmonary hypertension

Abstract

Truly reversible pulmonary hypertension is rare. Acquired systemic arteriovenous (A-V) fistulas following spinal surgery (laminectomy) are a less recognized cause of secondary pulmonary hypertension. We describe a patient who presented with symptoms and clinical evidence of pulmonary hypertension and underwent endovascular correction of an acquired A-V fistula, which led to improvement according to clinical and noninvasive hemodynamic criteria.

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Introduction

A less recognized reversible cause of secondary pulmonary hypertension are acquired systemic arteriovenous (A-V) fistulas following spinal surgery (laminectomy).

We describe a patient who presented with symptoms and objective findings of pulmonary hypertension. Fur-

ther evaluation revealed the presence of peripheral A-V fistula. Endovascular correction led to resolution of symptoms and improvement in pulmonary pressures.

Case Report

A 47-year-old woman was referred to our clinic for evaluation of pulmonary hypertension. Nine months prior to her admission, she experienced gradually progressive dyspnea on exertion, recent onset of fatigue, and lower extremity edema, while 3 months later, paroxysmal nocturnal dyspnea, and two- to three-pillow orthopnea developed. The last 6 months, she had been hospitalized in various departments, and underwent diagnostic evaluation with the final diagnosis of primary pulmonary hypertension. Past surgeries include a lumbar spinal disk surgery 18 months before admission. A chest radiograph at that time was normal.

She works in a hospital as a cleaning lady, and does not use tobacco, alcohol or other drugs. Current medications included iloprost (nebulizer), diltiazem, furosemide and spironolactone, without improvement.

The physical examination revealed an alert woman with no distress. She was afebrile, with a heart rate of 78 beats/min, a respiratory rate of 18 breaths/min, blood pressure of 120/75 mm Hg, and room air O₂ saturation was 98%. Cardiac examination showed jugular venous distension to the mandible, an increased splitting of S₂ and a 2/6 blowing holosystolic murmur across the precordium, being accentuated during inspiration. The rhythm was regular. Examination of the lungs revealed minimal crackles in the bases with vesicular breath sounds. Abdominal examination demonstrated

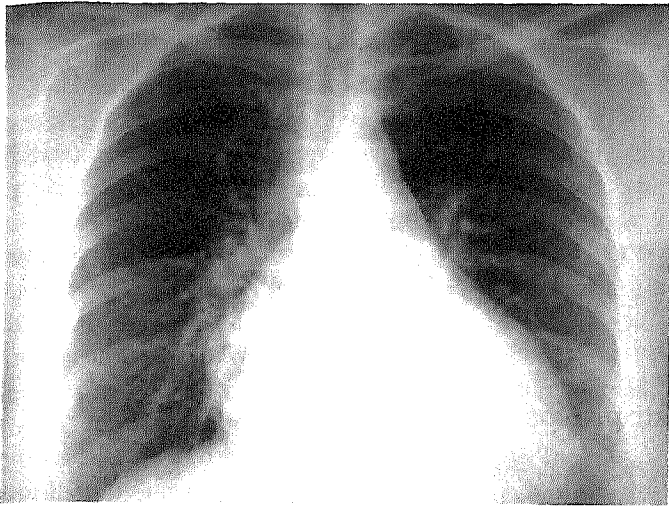


Fig. 1. Chest X-ray demonstrates moderate cardiomegaly with pulmonary vascular vein prominence, mild volume overload and enlargement of the pulmonary arteries.

hepatomegaly and the presence of a systolic abdominal bruit, which spread to the right inguinal region. There was 2+ pitting edema to the right, and 1+ pitting edema to the left knee, with chronic stasis bilaterally.

Laboratory evaluation was normal, and the following results of arterial blood gas tests performed at rest with room air were obtained: pH 7.48, PO₂ 97.2 mm Hg and PCO₂ 28.6 mm Hg. An electrocardiogram showed sinus rhythm, right axis deviation, R/S > 1 in V1, large P in lead II, and ST depression in lead III. A chest radiograph showed moderate cardiomegaly with pulmonary vascular vein prominence, mild volume overload and enlargement of the pulmonary arteries (fig. 1). An echocardiogram showed left atrial enlargement (41 mm), right atrial and right ventricular enlargement, an ejection fraction of 65%, moderate mitral and tricuspid valve regurgitation, diastolic dysfunction, and right ventricular systolic pressure of 48 mm Hg. Evaluation before referral included: pulmonary function studies showing normal findings, a ventilation-perfusion (V/Q) scan demonstrating a low probability of pulmonary embolism, spiral computed tomography of the chest showing cardiomegaly, and CT of the abdomen revealing inferior vena cava dilatation and moderate ascites. After diuresis, the patient underwent right heart catheterization: systolic and diastolic pulmonary artery pressure were 54 and 27 mm Hg, respectively (mean 39 mm Hg), cardiac output was 6.0 liters/min and the cardiac index 3.6 liters/min/m² with a body surface area of 1.68 m². Pulmonary vascular resistance was 240 dyn-sec/cm⁵, central venous pressure was 14 mm Hg, pulmonary capillary wedge pressure 21 mm Hg, blood pressure 147/56 mm Hg and heart rate 83 beats/min.

In our patient, considering the normal chest radiograph before spinal surgery, the onset of symptoms of pulmonary hypertension 9 months after the operation, the fact of an abdominal bruit and a lower extremity edema, the lack of improvement after treatment with diuretics, diltiazem and iloprost nebulizer, and the normal findings from our extensive evaluation, the differential diagnosis was primarily limited to systemic A-V shunts. Doppler examina-

tion of the abdominal and leg vessels revealed an A-V fistula between the right common iliac vein and the right common iliac artery being 9 mm in diameter, with dilatation of the common iliac vein, inferior caval vein and of the intrahepatic veins. A subsequent angiography confirmed the Doppler findings, and after intravascular ultrasound, the fistula was successfully occluded by two covered stents placed on the arterial side of the fistula.

The patient experienced rapid improvement of symptoms within the 1st week after angiography. Two weeks, and 3 and 6 months later, she underwent chest radiography, echocardiography and Doppler examination of the abdominal and leg vessels which showed no abnormal findings.

Discussion

Pulmonary hypertension is often enigmatic in presentation. This is because the symptoms and signs are vague and common to many diseases. Furthermore, symptoms of pulmonary hypertension may well be overshadowed by the symptoms of accompanying diseases and, in addition, because there is no sphygmomanometer for the pulmonary circulation, making a diagnosis requires cardiac catheterization [1].

To reach a diagnosis, the clinician must be aware of the possibilities. Once pulmonary hypertension is identified, a cause must be found. Investigations include a V-Q scan, pulmonary function tests, arterial blood gas measurements, CT scanning and echocardiography. Sometimes, these investigations do not lead to a clear diagnosis, and other clues must be sought [1].

When systemic A-V shunts are present, the clinical and hemodynamic aspects may vary depending on whether these shunts are congenital or acquired, and on their physical characteristics. Depending on the size of the fistula, and transfer of blood from a high- to a low-pressure system, this sequence of events may take years or may occur rapidly [2]. Although A-V shunts have long been known to potentially cause high output failure, marked pulmonary hypertension is rare [2]. Causes for acquired A-V communications include penetrating trauma, surgery, vascular procedures including central lines, Paget's disease, chronic liver disorders and myelofibrosis [2-6].

Our patient did not report onset of dyspnea until 9 months after the disk surgery, with onset of symptoms of right cardiac failure even later. The occurrence of an A-V shunt with pulmonary hypertension after spinal disk surgery has rarely been reported in the English literature, and in all of them high output cardiac failure was present [2, 3]. In our case, high output cardiac failure was absent as CO was on the superior normal value. This fact can lead

to an erroneous diagnosis of the etiology of pulmonary hypertension and can compromise the patient's outcome.

A systemic A-V shunt after laminectomy can rarely cause pulmonary hypertension, and this has to be considered as a rare cause of secondary pulmonary hypertension, which up to now has not been included in conventional differential diagnoses of pulmonary hypertension. Although treatment for most acquired A-V shunts is sur-

gical, recently endovascular techniques have been recommended, lowering the high morbidity and mortality related to conventional repair [5].

When pulmonary hypertension of an uncertain etiology occurs in patients previously submitted to disk surgery, a careful clinical examination can lead to a correct diagnosis of an iatrogenic A-V fistula, and surgical or endovascular correction usually results in the restoration of the normal cardiac function [7].

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