

Pulmonary Varix Mimicking Pulmonary Arteriovenous Malformation in a Patient with Turner Syndrome

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Key Words

Pulmonary varix · Arteriovenous malformation · Turner syndrome

Abstract

A 36-year-old asymptomatic female with Turner syndrome was referred for a 3-cm opacity of the left lung detected by routine chest X-ray. A computed tomography scan of the chest suggested a vascular lesion such as pulmonary arteriovenous malformation, and transcatheter embolotherapy was considered. The lack of a right-to-left shunt on contrast echocardiography led to suspect an alternate diagnosis. Magnetic resonance imaging and pulmonary angiography eventually demonstrated a pulmonary varix associated with a partial anomalous pulmonary venous return. Contrast echocardiography may help to distinguish between pulmonary varix and arteriovenous malformation.

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Introduction

Pulmonary vein varix is a rare and benign disorder which usually presents as a parenchymal or mediastinal mass on chest X-ray and may mimic other vascular ab-

normalities or lung cancer [1]. We report the case of a young woman with Turner syndrome, in whom pulmonary arteriovenous malformation (PAVM) was suspected. The lack of a right-to-left shunt on contrast echocardiography led to suspect an alternate diagnosis. Magnetic resonance imaging and pulmonary angiography eventually demonstrated a pulmonary varix associated with a partial anomalous pulmonary venous return (PAPVR).

Case Report

A 36-year-old female with Turner syndrome without known cardiac malformation underwent a routine chest X-ray which disclosed a well-delineated left midlung field opacity, 3 cm in diameter, which was further interpreted on a computed tomography scan of the chest as PAVM (fig. 1). She was thus referred to our center for transcatheter embolotherapy.

The patient was asymptomatic, with no telangiectasia, clubbing or cyanosis. Heart and breath sounds were normal. Lung function tests including measurement of carbon monoxide diffusion capacity were normal. Arterial blood gas studies on room air were normal with PaO₂ in the supine position of 13.6 kPa. There was no orthodeoxia, and the alveolar-arterial PO₂ (AaPO₂) gradient under 100% oxygen was normal (2 kPa). In the context of Turner syndrome, a cardiovascular malformation was suspected. Echocardiography showed neither cardiovascular abnormality nor pulmonary hypertension. Transthoracic contrast echocardiography was performed by injecting 4.5 ml of agitated modified fluid gelatin solution (Plasmin, Fresenius Kabi, France) with 0.5 ml room air into a periph-

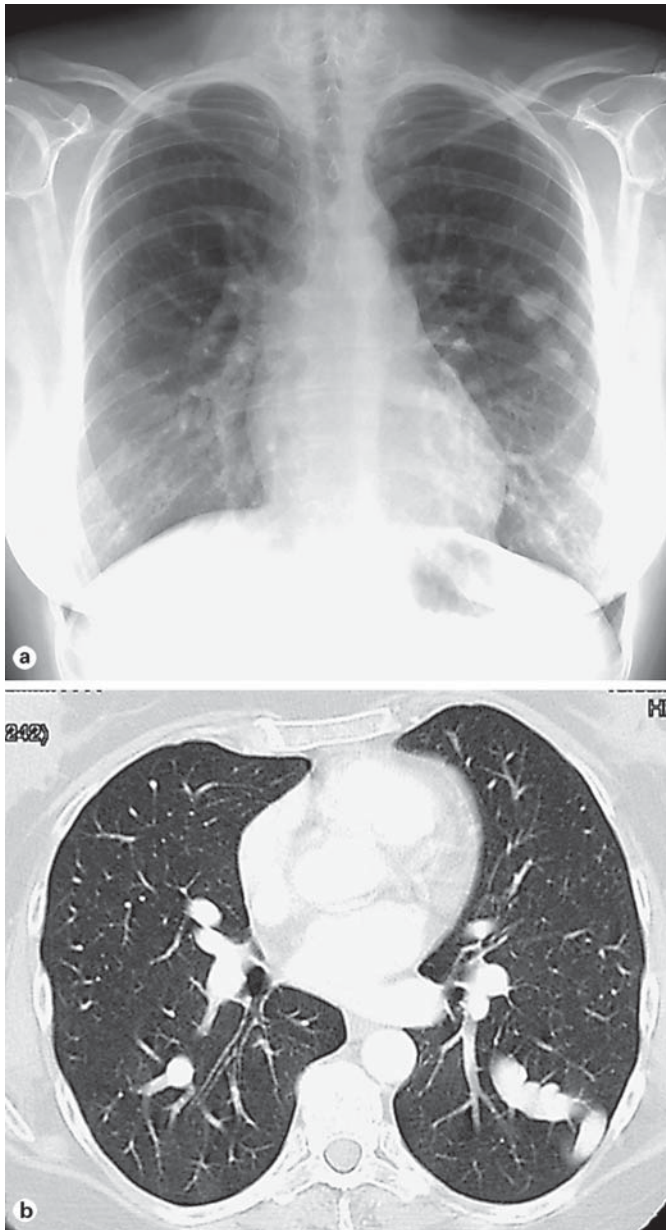


Fig. 1. Chest radiograph showed a well-delineated left lung opacity, with a contiguous smaller opacity (a). Computerized tomography of the chest showed a rounded opacity suggestive of pulmonary arteriovenous malformation (b).

eral vein while simultaneously imaging the left and right atria with 2-dimensional echocardiography. No contrast was visualized in the left atrium, so that right-to-left shunting (and thus PAVM) was considered unlikely. Chest computed tomography showed serpiginous opacities of vascular nature in the peripheral and central left lung, as well as PAPVR with the right upper pulmonary vein draining directly into the superior vena cava. The arterial phase of the left pulmonary angiography was normal; the venous phase identi-

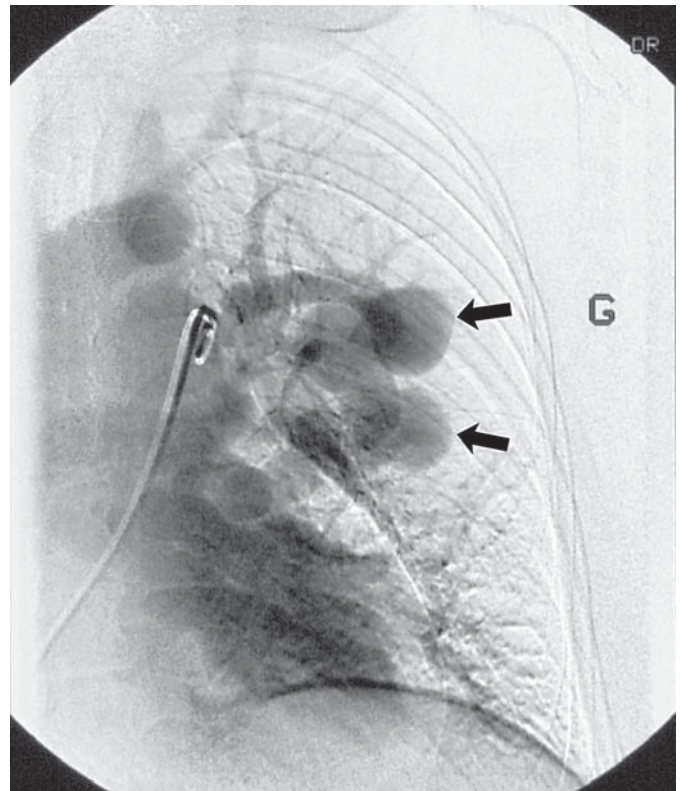


Fig. 2. The venous phase of digital subtraction left pulmonary angiography demonstrated pulmonary varices (arrows) of the left upper and lower pulmonary veins.

fied a delayed pooling of contrast in dilated pulmonary veins leading to the final diagnosis of pulmonary varices (fig. 2). The varicose dilatations involved peripheral as well as central segments of inferior and superior left pulmonary veins. Right pulmonary angiography confirmed the PAPVR detected on computed tomography (fig. 3). The arterial pulmonary pressure was normal. Pulmonary angiography and magnetic resonance imaging both excluded proximal pulmonary vein stenosis. No treatment was performed and a radiological follow-up was recommended.

Discussion

Pulmonary varix, arterial aneurysms, PAPVR and the more common PAVM (especially encountered in patients with hereditary hemorrhagic telangiectasia) may all present with similar radiological findings. However, only PAVM may be responsible for right-to-left shunting, since the other three lesions do not comprise vascular communications bypassing the pulmonary capillary bed. Measurements of AaPO₂ under 100% oxygen and con-

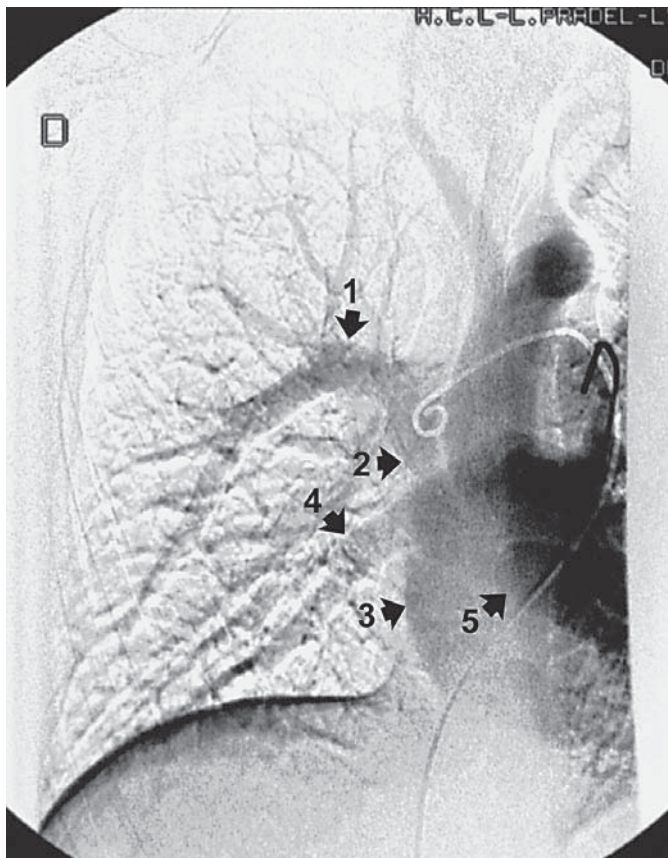


Fig. 3. The venous phase of digital subtraction right pulmonary angiography showing the right upper pulmonary vein draining directly into the superior vena cava. 1 = Right upper pulmonary vein; 2 = superior vena cava; 3 = right atrium; 4 = right lower pulmonary vein; 5 = left atrium.

trast echocardiography are two accepted methods used to detect right-to-left shunting. We previously reported that contrast echocardiography was the most useful noninvasive test for the screening of PAVM in patients with hereditary hemorrhagic telangiectasia, with a sensitivity of 92% and a predictive value for a negative test of 97% [2]. Therefore, we reasoned that a negative contrast echocardiography in this patient presenting with a pulmonary vascular abnormality should suggest another diagnosis than permeable PAVM.

Pulmonary varix is a rare abnormality defined as a localized dilatation of a pulmonary vein [1]. It has been reported as a consequence of longstanding pulmonary venous hypertension due to mitral regurgitation [3] and in a patient with end-stage liver disease and portal hypertension [1]. Isolated and congenital pulmonary varix may result from a developmental aberration with dilatation of

a persistent embryologic venous drainage channel and may be associated with various other congenital malformations.

Isolated pulmonary varices are usually asymptomatic. Multislice helical computed tomography, magnetic resonance imaging and transesophageal echocardiography may in some cases obviate the need of pulmonary angiography, which shows the characteristic opacification of a dilated pulmonary vessel only at the venous phase [4]. Pulmonary varices may progressively enlarge over time, especially in patients with mitral regurgitation, thereby requiring surgery valvular replacement, which may result in regression of the varix. Rare but potentially severe complications of pulmonary varix include rupture into the bronchial tree or pleural space and systemic emboli secondary to thrombosis within the varix. Surgery is usually not warranted for uncomplicated cases.

PAPVR is a congenital vascular malformation found in 0.4–0.7% of autopsies, corresponding to a persistent embryonic anastomosis between the systemic and pulmonary vein plexus. Depending on the number and site of anomalous pulmonary vein connections and the presence of frequently associated congenital heart disease (most commonly atrial septal defect), patients may remain asymptomatic or present with clinical manifestations of right ventricle volume overload and pulmonary hypertension secondary to left-to-right shunting [5].

Congenital heart diseases, mostly bicuspid aortic valve and coarctation of the aorta, are highly prevalent in Turner syndrome (35–50%). PAPVR has recently been recognized as one of the most common congenital vascular malformation with this syndrome [6]. Pulmonary varix has been exceptionally reported in Turner syndrome, together with PAPVR [7] (as in this patient) or aortic coarctation [8], or as a consequence of a proximal pulmonary vein obstruction following surgical correction of PAPVR 6 years before [9], suggesting that X-linked factors may be involved in determining vascular defects in Turner syndrome [6].

In conclusion, we report two rare venous malformations (pulmonary varix and PAPVR) that may not be coincidental in a patient with Turner syndrome and emphasize the value of contrast echocardiography to distinguish pulmonary varix from PAVM.

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