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Pulmonary Vein Varices Are Syndromic Features in Turner Syndrome

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We report herein a case of pulmonary vein varix in a 23-year-old asymptomatic female patient with Turner syndrome (45, X), who was initially referred for transcatheter embolotherapy of suspected pulmonary arteriovenous malformations (PAVMs) found incidentally. Careful review of the computed tomography of the chest (Fig. 1) in fact did not show any pulmonary artery abnormality or aneurismal sac, ruling out PAVMs. The diagnosis of pulmonary varix associated with partial anomalous pulmonary venous return (PAPVR) was eventually established after thorough review of bizarre tortuous veins. Arterial blood gases were normal. Echocardiography did not demonstrate any other cardiovascular abnormality or pulmonary hypertension. Noninvasive follow-up was recommended.

Pulmonary vein varices are extremely rare, with fewer than 100 cases reported. They may be secondary to pulmonary venous hypertension caused by left heart disease [1] or to obstruction of pulmonary veins [2], or they may remain idiopathic, e.g., isolated or associated with other vascular malformations, such as PAPVR, coarctation of the aorta, or congenital heart disease.

Interestingly, we have previously reported in *Respiration* [3] a very similar case of pulmonary vein varix associated with PAPVR in a patient with Turner syndrome. At that time, the diagnosis of pulmonary varix was not raised until agitated saline echocardiography and 100% oxygen test failed to detect right-to-left shunt in the anomalous vessels spuriously suspected to correspond to PAVMs. Although such an association was considered possibly coincidental at that time despite a previous case [4], the current identical observation raises the hypothesis that the association of pulmonary varix with Turner syndrome may indeed represent a rare syndromic association related to genetic defects in vascular development.

Financial Disclosure and Conflicts of Interest

The authors declare that they have no conflicts of interest related to this article.

Fig. 1. Unenhanced computed tomography of the chest showing dilated pulmonary veins (pulmonary varices) in the lingula (**a**, arrow) and in the left upper lobe (**b**, arrow) crossing the fissure.

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