Echocardiographic Diagnosis of Pulmonary Vein Varix

Jun Shiraishi, MD; Tetsuya Tatsumi, MD; Masaki Kimata, MD; Daisuke Kambayashi, MD; Akiko Mano, MD; Satoshi Yamanaka, MD; Miyuki Kobara, MD; Akihiro Azuma, MD; Masao Nakagawa, MD

Pulmonary vein varix is a rare abnormality, often resembling a pulmonary or mediastinal mass on chest roentgenograms, and pulmonary angiography has been the mainstay of diagnosis. An unusual case of pulmonary vein varix was clearly defined by echocardiography performed in a 47-year-old woman with chest discomfort who had been found to have an abnormal structure behind the cardiac silhouette on a chest radiograph. The diagnosis of left lower pulmonary vein varix was noninvasively confirmed by transthoracic echocardiography and transesophageal echocardiography. (Circ J 2003; 67: 796–798)

Key Words: Echocardiography; Magnetic resonance imaging; Pulmonary vein varix

Case Report

A 47-year-old woman was admitted because of chest discomfort. She did not have a history of cardiovascular disease and the results of physical examination, routine blood tests and electrocardiography were unremarkable. Chest radiography suggested a mediastinal mass inside the left cardiac border in the posteroanterior view (Fig 1A) and behind the cardiac silhouette in the lateral view (Fig 1B). Two-dimensional transthoracic echocardiography (TTE) was performed. On the parasternal long-axis view, the motion and chamber size of the left ventricle were normal, and there was no enlargement of the left atrial or right ventricular chambers. On the short-axis view and apical 4-chamber view, an echo-free mass was observed outside the posterolateral wall of the left atrium (Fig 2A, B). There was no indentation of the left ventricular wall, suggesting that the mass was an extrapericardial structure. Color Doppler echocardiography detected blood flow from the mass into the left atrium (Fig 2C). Mitral regurgitation was not evident. Transesophageal echocardiography (TEE) more clearly demonstrated dilatation of the pulmonary vein in the proximal left lower lobe with normal intrapulmonary venous connections (Fig 3A, B). The varix communicated with the left atrium. The right pulmonary veins and left upper pulmonary vein were normal. The wall of the varix was

Fig 1. Postero-anterior (A) and lateral (B) chest X-ray showing the mediastinal mass (black arrowhead and white arrowhead, respectively).

Fig 2. Two-dimensional (A, short-axis view; B, apical 4-chamber view; C, apical 4-chamber color Doppler image) echocardiography showing echo-free space (white arrowheads) outside the posterolateral wall of the left atrium and the blood flow from the space into the left atrium. LV, left ventricle; LA, left atrium; Ao, aorta.

Circulation Journal Vol.67, September 2003
akinetic, and blood flow from the peripheral pulmonary vein was observed in the varix (Fig 3B). There were no echoes suggestive of a thrombus. On the basis of the echocardiographic findings, a left lower pulmonary vein varix was diagnosed and we used cardiac MRI to confirm the diagnosis. T1-weighted images showed marked enlargement of the proximal portion of the left lower pulmonary vein, which was not covered with a hyperintense zone suggestive of pericardial fat, a finding considered consistent with a pulmonary vein varix (Fig 4). The maximum diameter of the pulmonary varix was \(4 \times 3\) cm on MRI. Cardiac catheterization showed normal hemodynamics and coronary arteries. Left ventriculography showed normal contraction without distortion. Pulmonary angiography was performed. The arterial phase was normal. Delayed images of the pulmonary circulation confirmed dilatation of the proximal left lower pulmonary vein, which was akinetic and communicated with normal peripheral pulmonary veins (Fig 5).

These findings confirmed the diagnosis of a pulmonary vein varix and the patient was given anticoagulant therapy to decrease the risk of systemic embolization. Follow-up chest radiography and echocardiography have revealed no complications.

**Discussion**

Pulmonary vein varices are rare vascular abnormalities of the lung that are mostly asymptomatic and detected incidentally as an abnormal structure on chest radiographs. The complaint of the present patient was not related to a pulmonary vein varix, because her symptoms disappeared soon after admission.

Pulmonary vein varices can be classified into 3 types based on the morphological findings on chest roentgenograms: \(^1\) the saccular type is defined as a localized, oval, or saccular dilation of the pulmonary vein; the tortuous type as a twisted, elongated dilation of the pulmonary vein; and the confluent type as a dilation in the confluence of the pulmonary vein. The saccular type involves the left upper lobe as frequently as the right lower lobe! According to this classification, the present patient had a saccular type at an uncommon site.

The etiology of pulmonary vein varices is uncertain, but the rare presence of these vascular lesions in neonates suggests that some pulmonary vein varices are congenital anomalies, probably resulting from dilatation of persistent embryological venous drainage channels. \(^2\) Histological examination of a pulmonary vein varix suggests no intrinsic defect in the structure of the wall! The confluent and tortuous types of pulmonary vein varices are often associated with mitral valvar disease and regression of the varices after valvular replacement has been reported, suggesting that pulmonary venous hypertension plays an important role in the development of aneurysmal dilatation of the pulmonary veins. \(^3\) However, localized saccular dilatation of the pulmonary vein has not been reported in any patient with mitral valvar disease. Pulmonary venous hypertension thus appears to be unrelated to the saccular type of pulmonary vein varix and we consider that the present case of the saccular type of pulmonary vein varix is congenital.

Several congenital or acquired morphologic abnormalities can develop within or around the left atrium. Differential diagnosis of the accessory cavity includes cor triatriatum, left atrial aneurysm, diverticulum, and pulmonary varix. \(^4\) A prominent confluence of the left atrial appendage and left pulmonary vein is easily distinguished from the partitioning membrane of the cor triatriatum by TEE. \(^5\) Echocardiography demonstrated a connection between the cavity and the peripheral pulmonary veins, suggesting that in this case it was not a left atrial aneurysm. In addition, MRI showed that the structure was not covered with pericardial fat, a finding considered inconsistent with a left atrial aneurysm. The cavity was also not an atrial diverticulum, because that contracts in synchrony with other parts of the heart!...
To our knowledge, this is the first report of a pulmonary vein varix was safely detected by TTE and diagnosed by TEE, disproving the contention that angiography is essential for diagnosis. Recently, enhanced CT, MRI, and TEE have been reported as important in the assessment and diagnosis of pulmonary vein varix.\(^9\)–\(^{12}\) For abnormalities of the pulmonary veins, MRI is as accurate as angiography and 2-dimensional echocardiography.\(^{13}\) However, TTE and TEE can detect and evaluate the mitral valvular disease that is often associated with pulmonary vein varix. Moreover, TEE can define the anatomic characteristics of a pulmonary vein varix and its direct connection to the left atrium without being affected by body fat or lung tissue. As in this patient, TTE with color Doppler imaging might also be useful in detecting and evaluating pulmonary vein varices.

Although usually benign, pulmonary vein varices have 3 major complications: systemic embolization secondary to thrombosis, rupture into the pleural space or a bronchus, and unnecessary diagnostic thoracotomy. A pulmonary vein varix associated with mitral valvular disease is treated by repair or replacement of the mitral valve, not by surgical intervention of the varix. Pulmonary vein varices without valvular disease can be followed by chest roentgenography, echocardiography, CT or MRI at regular intervals. Varices that increase in size or show a risk of complications are candidates for surgical intervention.

References


*Circulation Journal Vol. 67, September 2003*