A Case of Pulmonary Varix Associated with Superior Pulmonary Vein Occlusion

Sakurako Tajiri, MD,1 Jun Koizumi, MD,2 Takuya Hara, MD,2 Masahiro Kamomo, MD,3 Naoki Hayama, MD,1 Ichiro Kobayashi, MD,1 Yusuke Kondo, MD,1 Tetsuri Kondo, MD,4 Koichiro Asano, MD,1 and Tadashi Abe, MD1

A pulmonary varix is a localized dilatation of a pulmonary vein, which is usually asymptomatic presented as a mass on a chest roentgenogram, and diagnosed with pulmonary angiography. We encountered a case of 55 year-old man, in whom incidentally identified was a dilated blood vessel that passed through the minor fissure and returned to the inferior pulmonary vein, which we diagnosed as pulmonary varix. This vascular anomaly was accompanied by the occluded superior pulmonary vein, highly suggestive of the developmental mechanism of this disease.

Keywords: pulmonary varix, multislice CT, pulmonary vascular occlusion

INTRODUCTION

The first reported case of pulmonary varix was a child who died of gastrointestinal hemorrhage in whom varices were observed on autopsy in many organs including the lungs.1 Although it is rare, the disease has recently been occasionally reported with the recent dissemination of imaging. We encountered a patient with pulmonary varix, in whom the blood vessels passed through the minor fissure and returned to the inferior pulmonary vein, due to occlusion of the superior pulmonary vein. Herein, we report the case because it is highly suggestive of the developmental mechanism of this disease.

CASE REPORT

A 55 year-old man visited a physician due to fever, cough, and sputum. An abnormal shadow, a 26-mm mass, overlapping the right hilum in the frontal view, and an arch-shaped shadow in the retrosternal space, in the lateral view, was pointed out on plain chest X-ray radiography. The symptoms remitted after treatment with antibiotics, but the abnormal shadow did not shrink. His chest CT demonstrated a node in the right S3 region (Fig. 1A) and a blood vessel that was continuous from this node to the superior lobe and hilum of the lung (Fig. 1B). Granular shadows of micro blood vessels were also noted in the surrounding region. Based on these findings, the patient was diagnosed with pulmonary arteriovenous malformation (AVM) of A3b and V3/V5 as the afferent and efferent blood vessels and referred to our hospital for intravascular treatment.

The patient had no particular past medical history including bloody phlegm or hemoptysis. With regards to the family medical history, his mother and brother had diabetes, but no disease of vascular aberration. There were no abnormal findings in the physical examination or laboratory tests. Blood gas analysis under room air breathing shows PaO2, 84.2 Torr; PaCO2, 37 Torr; pH 7.41.

The patient was admitted for angiography, which shows no AVM in the pulmonary arterial phase (Fig. 2A)
on contrast injection through the root of the pulmonary artery. Two blood vessels extending up- and downward from the right pulmonary hilum overlapping the ascending aorta were visualized in the pulmonary venous phase. When a catheter was advanced to the right pulmonary artery and a right anterior oblique image was acquired, $A^3_b$ was bent and distributed forward, but no abnormal blood vessel was visualized in the pulmonary arterial phase, and it connected to a nidus-like abnormal blood vessel and flowed into $V^3_b$ and $V^5_b$. In the lateral view, on contrast imaging of $A^{1+3}$, no inflow of the pulmonary artery into a nidus was noted, and a nidus-like blood vessel appeared from $V^3_b$ in the phase visualizing the pulmonary vein following the lung parenchyma. The vessel passed through the minor fissure and then flowed into the left atrium through $V^5_b$ (Fig. 2B). On multiplanar reformation using the volume data of multislice CT, the pulmonary varix became like a nidus from the right $V^3_b$ and then became a single dilated vein again, penetrated the minor fissure, and reached the right $V^5_b$, distributing

**Fig. 1** CT before pulmonary angiography. A: axial reconstruction. B: surface rendering (right lateral view). A nidus-like lesion (A: arrow) is seen in segment 3 on axial reconstruction CT. Surface rendering demonstrates the nidus-like (B: arrow) lesion between the upper lobe vessel and middle lobe vessel.

**Fig. 2** Pulmonary angiography. A: The right pulmonary arteriography (RAO). B: The selective $A^{1+3}$ arteriography (lateral direction). Any arterio-venous malformation is not seen in arterial phase of the right pulmonary arteriography (A). The venous phase of the selective $A^{1+3}$ arteriography reveals a large curved vessel (B: arrowheads) between the upper and middle lobe.
Pulmonary Varix Associated with PV Occlusion

over 2 lobes (Fig. 3A, arrowheads). In addition, occlusion or stenosis of a tapering pulmonary vascular branch was noted in the right superior lobe (Fig. 3B, arrow).

Further examination showed no apparent cerebral AVM in the brain or old cerebral infarction on cephalic MRI, no evidence of a right-left shunt on pulmonary blood perfusion scan. Echocardiography showed favorable wall motion with no cardiac anomaly. The ejection fraction was 68.4%, and no right heart overload was noted. No treatment was performed, and the patient is now under the course observation.

**DISCUSSION**

The patient was diagnosed with pulmonary varix because the angiographic findings met all conditions reported by Bartram et al.2): 1) the pulmonary artery was normal in the arterial phase, 2) no pulmonary arteriovenous shunt was present, 3) a blood vessel flowed directly into the left atria from the pulmonary varix, 4) contrast medium remained in the varix, compared to retention in the normal pulmonary vein, and 5) the peripheral region of the dilated tortuous vein was normal. Pulmonary varix is divided into congenital and acquired cases. It is considered that congenital cases develop during embryonic development,2) whereas acquired cases mainly accompany mitral valve disease and are caused by blood retention due to pulmonary venous pressure elevation and reflux into the pulmonary vein.3)

In this patient, congenital heart disease was ruled out on cardiac Doppler echography, and no left atrial dilation or valve disease was noted. In addition, the morphology showing perforation of the interlobular fissure by the blood vessel cannot be explained by the acquired pulmonary varix. The occlusion of the right superior lobe branch of the pulmonary vein, clarified by multiplanar reformation of multislice CT, suggested that it was due to hypoplasia or occlusion of the branch, which caused vascular advancement as a collateral pathway simultaneously with the development of the interlobular region and secondarily formed the pulmonary varix, rather than due to abnormal distribution in the embryonic development of the pulmonary vein. Only a few cases of the pulmonary vein passing through the interlobular fissure have been reported by Kumazoe et al.4) and Hanson et al.5) The diagnosis of an abnormal blood vessel passing through the lung parenchyma and then thoracic cavity is important because it may cause hemorhax.6,7) Only one case of varix accompanied by pulmonary venous occlusion diagnosed by angiography has previously been reported,8) and no case directly imaged by multislice CT has been reported.

There is no sex difference in the incidence of pulmonary varix, and it can be observed at any age, but acquired cases accompanying heart disease increases with aging. It is asymptomatic and incidentally discovered on chest plain X-ray radiography in many cases, as a round-to-oval, homogeneous shadow, and benign or malignant lung tumor, mediastinal tumor, bronchial cyst, lymph node swelling, and pulmonary arteriovenous

---

**Fig. 3** Reconstructed CT after pulmonary angiography. A: Multi planer reformation (oblique). B: Partial maximum intensity projection (MIP). The curved vessel traverses the minor fissure (A: arrows). The partial MIP image reveals the occluded upper pulmonary vein (B: arrow).
fistula are included in diseases to be differentiated.\(^9\) CT is low-invasive, and the location and distribution of the lesion can be accurately identified. Particularly, the collection of volumetric data has recently become possible employing multislice CT, which has facilitated making the diagnosis without invasive angiography. However, temporary resolution is insufficient to image timecourse hemodynamics, and this may have been one reason that the lesion was diagnosed as pulmonary arteriovenous malformation on the first CT in our case. The tissue constitution of the varix wall is normal in surgical cases, and acquired pulmonary varix disappears after vulvular surgery in a previous report,\(^10\) suggesting that varix may be masked on CT because the volume of the venous lumen freely changes. Varices diagnosed on MRI and transesophageal echography have recently been reported.\(^\)\(^11\)\(^12\) To discover a hidden pulmonary varix and reliably rule out pulmonary arterial abnormality and right-left shunt, angiography is still important to make the definite diagnosis.

Since the varix was asymptomatic in the presented case, course observation was selected. In the literature, middle lobe syndrome due to hemoptysis and compression of the bronchus,\(^13\) swallowing disorder due to compression of the esophagus,\(^14\) and cerebral infarction assumed to be caused by parietal thrombus\(^7\) have been reported. Hemoptysis can occur due to complications such as bronchiectasis, active pulmonary tuberculosis, and pulmonary congestion due to venous retention, rather than varix rupture, in many cases, but the development of hemothorax has been reported, as described above.\(^7\) Thus, we will continue strict course observation.

We encountered a patient with a rare pulmonary varix in whom the blood vessel penetrated the interlobular pleura and flowed into the inferior pulmonary vein due to occlusion of the superior pulmonary vein, and these could be visualized employing multislice CT. Careful image reconstruction from the volume data is important to differentiate pulmonary varix from pulmonary arteriovenous malformation.

**Disclosure Statement**

The authors have no conflict of interest to declare.

**References**

1. Arnett JC, Patton RM. Pulmonary varix. Thorax 1976; **31**: 107-12. [Medline] [CrossRef]
11. Wildenhain PM, Bourekas EC. Pulmonary varix: magnetic resonance findings. Cathet Cardiovasc Diagn 1991; **24**: 268-70. [Medline] [CrossRef]