Radiology Case Reports 17 (2022) 4183–4187

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Pulmonary varix clearly demonstrated by 3D-CT before pulmonary angiography ☆

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ARTICLE INFO

Article history:
Received 12 June 2022
Accepted 8 August 2022

Keywords:
Pulmonary varix
Pulmonary arteriovenous malformations
Pulmonary vein
Pulmonary angiography

ABSTRACT

Pulmonary varix is a rare and usually asymptomatic localized dilation of a pulmonary vein. This disease should be distinguished from other pulmonary and mediastinal diseases, particularly pulmonary arteriovenous malformations. Herein, we encountered a case of pulmonary varix clearly demonstrated by 3-dimensional reconstructed computed tomography (3D-CT) which proved useful in its diagnosis. The 3D-CT enabled easy understanding of the vascular connections and confirmation of the absence of an inflow pulmonary artery. We also performed angiography which showed findings consistent with those obtained by the 3D-CT, thus confirming the diagnosis of pulmonary varix. After the diagnosis, the patient was followed up for several years without any treatment and she remained asymptomatic. On follow-up CT, the lesion remained unchanged.

Introduction

Pulmonary varix is a rare and usually asymptomatic localized dilation of a pulmonary vein [1]. This disease was first reported by Heidinger et al. in 1907, and earlier cases were only diagnosed at autopsy. Since 1951, pulmonary varix has been diagnosed by angiography [1].

Pulmonary varix should be clearly distinguished from other pulmonary and mediastinal diseases, particularly pulmonary arteriovenous malformations. There are presently no treatment guidelines for pulmonary varix [2]. In cases that are asymptomatic, course observation may be selected. However, in cases that are symptomatic or have a high risk such as pulmonary varix rupture or cerebral infarction due to a thrombus in the aneurysm, surgical or transcatheter treatment should

☆ Competing Interests: There are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

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https://doi.org/10.1016/j.radcr.2022.08.023

be considered [2]. In this article, we report a case of pulmonary varix that had been incidentally identified by CT imaging for pneumonia.

Case report

The case involves a 44-year-old woman. She had a history of bronchial asthma from the age of 28 years, and also received treatment for atopic dermatitis. She was treated for pneumonia in another medical institution. We performed non-contrast chest computed tomography (CT) for pneumonia. Incidentally, the CT demonstrated an abnormal shadow in the right segment 6 region (S6). She had no symptoms after recovering from pneumonia, but was referred to our hospital for the diagnosis and treatment of the abnormal shadow.

Computed tomography

We performed contrast CT using a 64-row MDCT (Discovery 750HD; GE Healthcare; Chicago, USA) in 2 phases with the following imaging conditions: tube voltage 120 keV, helical pitch 0.984, auto exposure control, and axial images output at 1.25 mm slice thickness. A contrast material (iopamidol 370 mgI) was administrated. We obtained the pulmonary artery phase (7.1 seconds after bolus tracking of the pulmonary artery) and the late phase (90 seconds after injection) (Figs. 1A and B). Abnormal dilations of the branches of the right inferior and superior pulmonary veins were demonstrated. Both the right inferior and superior pulmonary veins continued to and connected at S6. There was no pulmonary artery that inflowed to the right inferior and superior pulmonary veins. On the axial image, it was not easy to demonstrate correctly the 3-dimensional (3D) vascular connections. Thus, we transferred the data to a workstation (ziostation2) to generate 3D-reconstructed images (Figs. 1C and D). These 3D-reconstructed images enabled easy understanding of the vascular connections and confirmed the absence of an inflow pulmonary artery. As there was no apparent pulmonary arteriovenous malformation, we diagnosed the disease as a tortuous type of pulmonary varix.

Angiography

We also performed angiography to confirm our pulmonary varix diagnosis. After right femoral artery puncture, we inserted a guiding sheath (Parent Plus 45^{TM}; Medikit, Tokyo) and performed pulmonary angiography using a 4F catheter. The catheter tip was located in the right inferior pulmonary artery, but the right superior pulmonary artery was also contrasted by the backflow (Fig. 2A). Abnormal vessels were not demonstrated in the phase of pulmonary artery dominant (Fig. 2B), but were depicted in the phase of pulmonary vein dominant (Fig. 2C). In addition, the abnormal vessel staining persisted even after the pulmonary veins had been washed out (Fig. 2D). These findings were consistent with the findings obtained on the 3D-reconstructed images. Based on these findings, the patient was definitively diagnosed with a tortuous type of pulmonary varix.

After the diagnosis, the patient was followed up for several years without any treatment and she remained asymptomatic. On follow-up CT, the lesion remained unchanged.

Discussion

Pulmonary varix is a localized dilation of a segment of a pulmonary vein which normally enters the left atrium [1]. It is a very rare disease with only 71 cases reported as of 2011 [3]. Its incidence remains unknown and it has been reported infrequently [4]. There is also no gender or age difference [3,4].

The localization of pulmonary varix was reported to be predominantly in the right lower lobe: 43 cases (60%) in the right lower lobe, 12 cases (17%) in the left upper lobe, 6 cases (8%) in the right upper lobe, 3 cases (4%) in the right middle lobe, and 3 cases (4%) in the left lower lobe [3]. In present case, the pulmonary varix was located in the right lower lobe.

There are 2 groups of causative factors: *congenital* and *acquired*. For the first group which is due to congenital fragility of the pulmonary vein wall, histologic examination of the pulmonary vein indicated no intrinsic wall defects [4]. Other congenital cardiovascular malformations can be associated with pulmonary varix, such as patent ducts, double outlet ventricles, coarctation of the aorta, pulmonary vein stenosis, ventricular septal defects and Klippel-Trenaunay-Weber syndrome [4]. The second group is acquired, mainly due to pulmonary venous hypertension after trauma or mitral valvular disease [2]. In the case of mitral valvular disease, all cases have been reported to be in the right lobe [5]. In the case of mitral regurgitation, the anatomical location of the mitral valve may be a contributing factor. In this condition, the regurgitated blood is directed from the left ventricle to the right pulmonary venous orifice, causing increased pressure in the right pulmonary venous locally, which leads to the formation of a varix at this site [2]. Pulmonary varices have been reported to disappear 9 months to 2 years after mitral valve replacement, or to remain unchanged in 5 years [2]. Also, no correlation has been established between the severity of pulmonary venous hypertension and the development of a pulmonary venous varix [4]. This suggests that additional factors other than regurgitation may contribute to the onset of the disease [2]. It has been reported that congenital cases often occur on the right side, the cause of which is unclear. In the present case, there were no secondary factors, and the patient was diagnosed with a congenital pulmonary varix.

Pulmonary varix is morphologically classified into 3 types: saccular type (localized, ovular form), tortuous type (extensive, irregular margins), and confluent type (localized at the confluence of pulmonary veins [3,4]. The saccular type is predominantly observed in the left upper lobe, and the tortuous and confluent types are primarily seen in the right lower lobe [4]. The confluent type is the most common type, and it usually occurs at the confluences of pulmonary veins adjacent to the left atrium [4]. Berecova et al. reported that most pulmonary varices in patients with valvular disease were of the confluent type (62%), with a few tortuous types (19%) [3]. The saccular type has not been associated with mitral valvular disease [4]. The present case had tortuous dilated vasculatures in

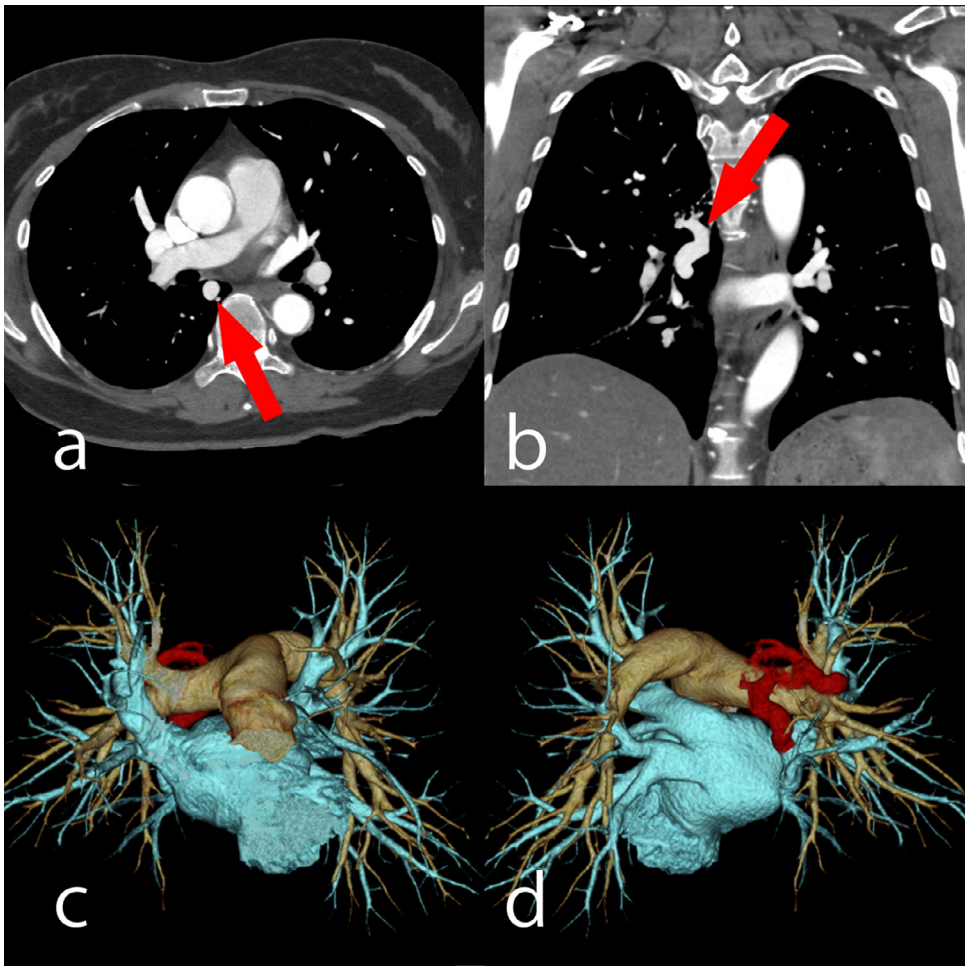

Fig. 1 – Contrast-enhanced CT. (A) Axial image; (B) Coronal reconstruction. Abnormal dilation of the right inferior pulmonary vein (arrow). On the axial and coronal images, the continuity of the vessels is not clear. (C, D) Three-dimensional reconstructed CT (3D-CT) with color indications as follows: pulmonary artery (dark yellow), pulmonary vein (light blue), and abnormal vessel (red). (C) Anterior-posterior view; (D) Posterior-anterior view. On 3D-CT, abnormal dilations of the right inferior and superior pulmonary veins were clearly demonstrated, and they were anastomosed in segment 6. This image clearly showed the absence of an inflow artery, thus pulmonary arteriovenous malformation was considered negative. The 3D-CT diagnosis was pulmonary varix.

the right lung, thus the patient was diagnosed with a tortuous type.

Pulmonary varices are usually asymptomatic and are incidentally detected during health check-ups [2]. They do not usually require any treatment unless complicated by cardiac disease [6]. The present case was also asymptomatic and was identified incidentally. With less frequency, the symptoms reported include cough, dyspnea, palpitations, chest oppression, orthopnea, hemoptysis, and cerebral infarction [7]. Middle lobe syndrome due to hemoptysis, compression of the bronchus, and swallowing disorder due to esophageal compression have been reported [4,8]. Death caused by rupture or thrombosis has also been reported [1]. Rupture can be caused by enlargement owing to cardiac disease or pulmonary tuberculosis infiltration, and infarction can be caused by a blood clot in the pulmonary vein [7]. Therefore, the course of pulmonary varix requires strict observation. If there is an increase in the size of the pulmonary varix or there is a risk of complications, surgical or vascular interventional treatment should be considered [4].

Pulmonary arteriovenous malformation is a disease with similar symptoms and imaging findings. Pulmonary varix should be accurately distinguished from pulmonary arteriovenous malformation because their treatment strategies are completely different. Pulmonary varix is almost asymptomatic and usually requires no treatment. In contrast, pulmonary arteriovenous malformation should be considered for treatment even in asymptomatic patients if the feeder diameter is over 3 mm. This is because of the risk of serious complications, such as cerebral infarction and cerebral abscess caused by right-left shunting [9]. The essential form of pulmonary arteriovenous malformation is a nodular area called a nidus, consisting of a collection of dilated thin-walled vascular sacs, a dilated pulmonary artery called a feeder that flows directly into the nidus, and a dilated pulmonary vein called a drainer that flows directly out of the nidus, forming a shunt in

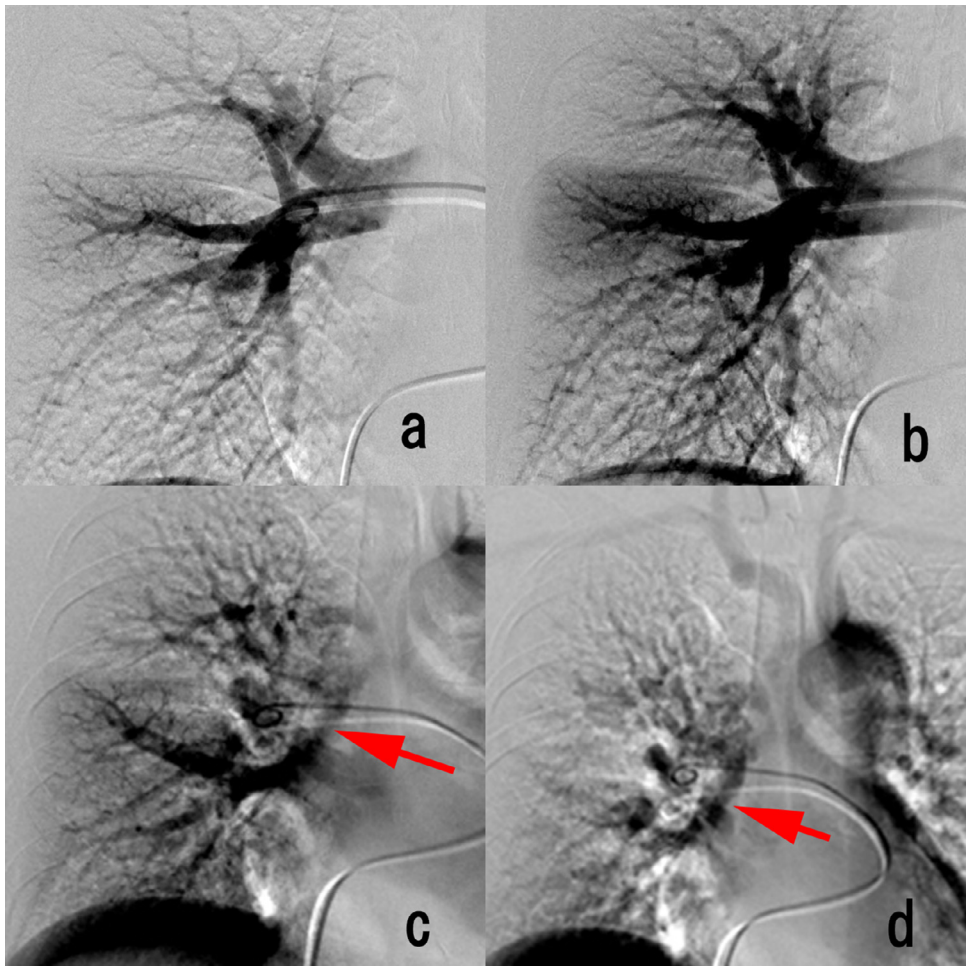

Fig. 2 – Pulmonary angiography. Pulmonary angiography with the catheter tip on the right inferior pulmonary artery. (A, B) The right inferior pulmonary artery was demonstrated, and the right superior pulmonary artery was also shown by the backflow. Arteriovenous shunt was not identified in this phase. (C) The right pulmonary vein was predominantly shown in this phase, and abnormal vessels (arrow) were identified. (D) The pulmonary vein was almost obscured in this phase, but the brachiocephalic artery, left internal carotid artery, and left subclavian artery were identified. The abnormal vessel (arrow) still remained. Based on these findings, the patient was definitively diagnosed with a tortuous type of pulmonary varix.

the pulmonary artery and pulmonary vein [7]. The presence of this shunt is the main feature that distinguishes both.

Pulmonary varix can be diagnosed by pulmonary angiography, with the following 5 diagnostic criteria set by Bartram and Strickland: 1) normal arterial phase, 2) filling of the varix in the venous phase, 3) direct drainage to the left atrium, 4) delayed emptying, and 5) localization to the proximal portion of the pulmonary vein [1,4]. Recently, with the spread of helical CT, it has become possible to reconstruct 3D images using 3D-CT. 3D-CT may demonstrate the above-mentioned criteria except criterion 4), and may be expected to distinguish pulmonary varix from pulmonary arteriovenous malformation. In the present case using 3D-CT (Fig. 1C and D), anastomosis with the pulmonary artery could not be identified, readily distinguishing pulmonary varix from pulmonary arteriovenous malformation.

Angiography remains the gold standard for identifying and diagnosing pulmonary aneurysms because of its superior temporal resolution and blood flow information. However, it has the disadvantage of being highly invasive. In recent years, the diagnosis of these diseases has been established using contrast-enhanced CT and magnetic resonance imaging (MRI) because of their noninvasiveness [9]. A case had been reported in which the pulmonary varix was diagnosed noninvasively by 4-dimensional (4D)-CT or 4D-MRI with not only anatomical information but also blood flow information [3]. However, there are several problems: the magnetic susceptibility is strongly influenced by the mixture of air and blood vessels which have very different magnetic susceptibilities; the left lower lung field is easily affected by motion artifacts from heartbeats; and a high spatial resolution signal-to-noise ratio is required to delineate the peripheral areas of the lesion. Delineation becomes difficult depending on the location [10].

Other cases have been reported in which transesophageal echocardiography was useful in evaluating blood flow [11]. In such cases, transesophageal echocardiography was used

to observe a pulmonary varix in the left inferior pulmonary vein, which is considered to occur infrequently. It can observe the direction of the mitral regurgitation signal due to mitral valve deviation, the direct flow into the pulmonary varix, and a turning flow in the pulmonary varix. Although it is not possible to make a diagnosis using transesophageal echocardiography alone, it may be useful as an adjunctive diagnostic tool for assessing the risk of augmentation in cases of mitral valve disease. A definitive diagnosis of pulmonary varix can be made by multidetector CT imaging with 3D and maximum intensity projection reconstruction, which can also provide a good visualization of the whole pulmonary vascular tree [4].

Conclusions

In this study, we encountered a case of pulmonary varix which was clearly demonstrated and diagnosed by 3D-CT. Although angiography was considered essential for diagnosis, 3D-CT provided comparable results. These results indicate that 3D-CT is useful in the diagnosis of pulmonary varix.

Patient consent

We fully explained to the patient that we would publish in a medical journal and that we would take all ethical considerations into account to protect her rights.

We have obtained the patient's consent by having her signature a document stating these. Thus, we obtained formal ethical approval for our paper. We thank the patient for her kindness.

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