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CASE REPORT

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Endovascular embolization of a congenital inferior phrenic artery-to-pulmonary arteriovenous malformation: a rare case report

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Abstract

Background Pulmonary arteriovenous malformation (PAVM) is abnormal arteriovenous shunts between pulmonary artery (PA) and pulmonary vein, and rarely has congenital direct communications with systemic arteries.

Case Presentation A 33-year-old male presented to our hospital with intermittent bloody sputum with no evidence of pulmonary infection, trauma or surgery. Chest computed tomography angiography (CTA) indicated the congenital inferior phrenic artery (IPA)-to-PAVM surrounded by diffuse alveolar hemorrhage located in the lower lobe of right lung. Both the afferent PA and IPA were successfully embolized with coils. Recurrent hemoptysis did not occur during one-year follow up.

Conclusions The congenital communication between IPA and PAVM is rare, and the abnormal direct shunt would induce hemodynamically unstable condition within PAVM. Endovascular embolization of the afferent PA and IPA is a safe and effective method for this abnormal congenital shunt in lung.

Keywords Anomalous systemic artery, Pulmonary arteriovenous malformation, Hemoptysis, Embolization, Case report

Introduction

The capillary network between bronchial arteries and pulmonary artery (PA) is the only congenital communications between systemic arteries and PAs in the normal lung [1]. The abnormal congenital communications between systemic arteries and PAs are mentioned in some previous cases, such as pulmonary sequestration, anomalous systemic arterial supply to normal basal segment, and the direct congenital fistula between systemic and pulmonary arterial collaterals [2, 3]. Pulmonary arteriovenous malformation (PAVM) is the direct communication between PA and pulmonary vein and does not have congenital communications with systemic arteries [4]. According to the number of the afferent artery and whether the feeding arteries branch from the same segment of PAVM, PAVM is classified as the simple type and the complex type [5, 6]. Previous studies have reported that systemic artery-to-PAVM was the acquired response of lung or pleural infection, or the recurrence of PAVM after embolization [7–10]. In the present report, we presented a case of a congenital direct communications between inferior phrenic artery (IPA) and PAVM. Both

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the afferent PA and IPA were successfully embolized with coils.

Case report

A 33-years-old man presented with intermittent bloody sputum over a week (≤20 ml per day), and vessel murmur was auscultated in the right lower lung lobe. Results of arterial blood gas analysis did not indicate hypoxemia, and the oxygen saturation of the patient is 99%. Chest computed tomography angiography (CTA) scan showed one enlarged PAVM located in the right lower lobe surrounded by the gland glass opacities suggesting alveolar hemorrhage. Three-dimensional volume-rendered (3D-VR) images disclosed that the enlarged PAVM had two feeding vessels: one afferent PA (Largest diameter: 7.5 mm; Originating from PA of S7 in right lung), and one right IPA (Largest diameter: 5.5 mm; Originating from the celiac trunk) (Fig. 1). Graded transthoracic contrast echocardiography (TTCE) was performed according to established guidelines, and TTCE was graded as one (9 microbubbles in left ventricle) [11]. The clinical diagnosis of hereditary hemorrhagic telangiectasia (HHT) was confirmed basing on the Curaçao criteria (clear family history of epistaxis, PAVM, and multiple telangiectasias in tongue) [12]. This patient did not have the history of chest trauma, surgery, or lung infection.

Then the endovascular treatment was performed. After local anesthesia and right femoral artery puncture, selective angiography was performed to confirm the location and measurement of the right IPA (Fig. 2-A). Superselective catheterization was implemented to anchor at IPA 1 cm off the detected PAVM by 4 F cobra-2 catheter (Cook Medical, Bloomington, IN, USA) and one 1.98 F microcatheter (Asahi Intec, Seto, Aichi, Japan) (Fig. 2-B).

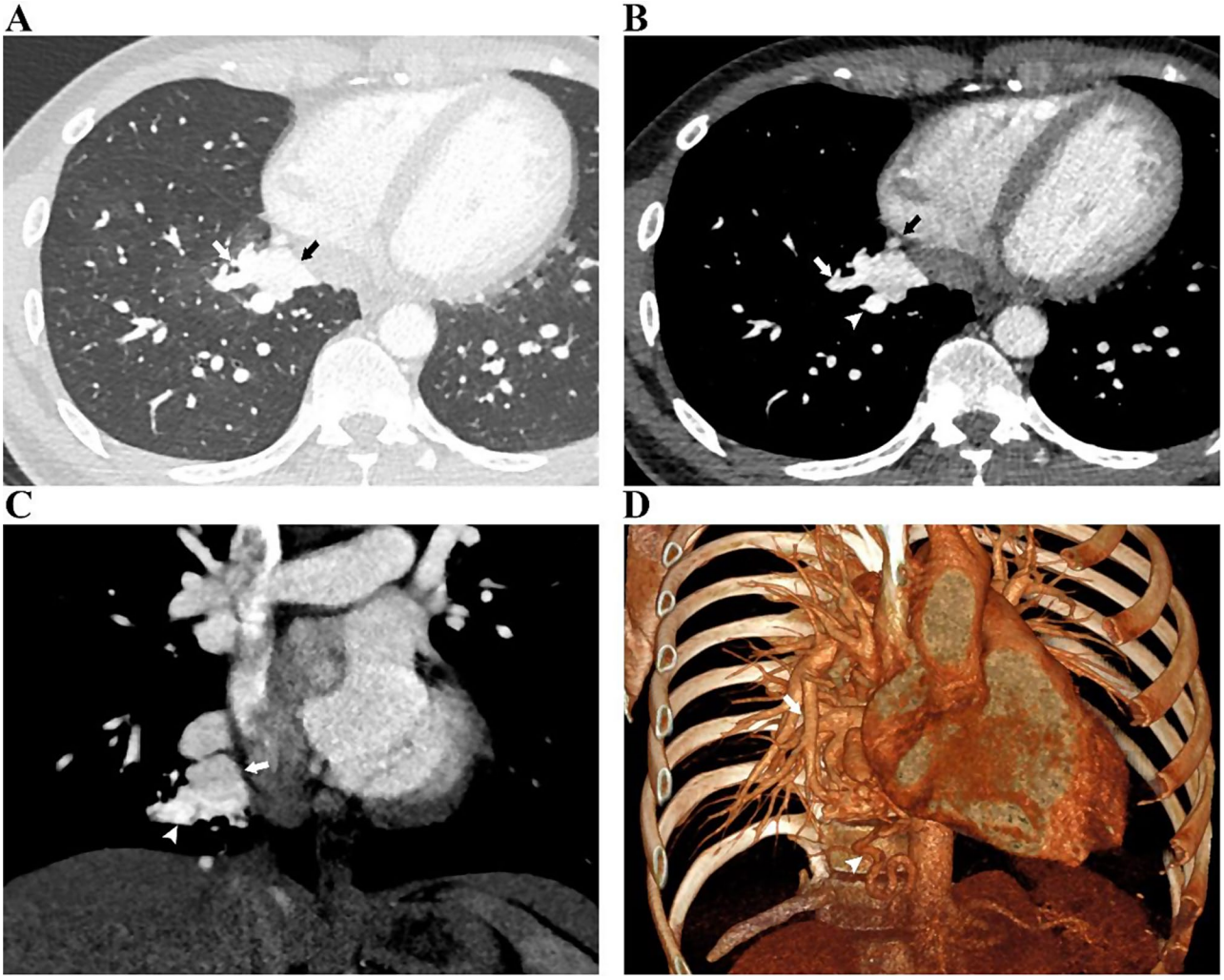

Fig. 1 **(A)** One enlarged PAVM (black arrow) is detected at right lower lobe, and is surrounded by ground-glass opacity (white arrow); **(B)** CTA clearly shows the afferent PA (white arrow), the right IPA (black arrow), and the draining pulmonary vein (white arrowhead); **(C)** Density stratification in the coronal chest CTA indicates the blood flow from systemic artery (arrowhead) has offset the blood flow of PA (arrow) because of pressure gradient between systemic artery and PA. **(D)** The 3D-VR image clearly shows the afferent PA (white arrow) and the abnormally enlarged right IPA (white arrowhead)

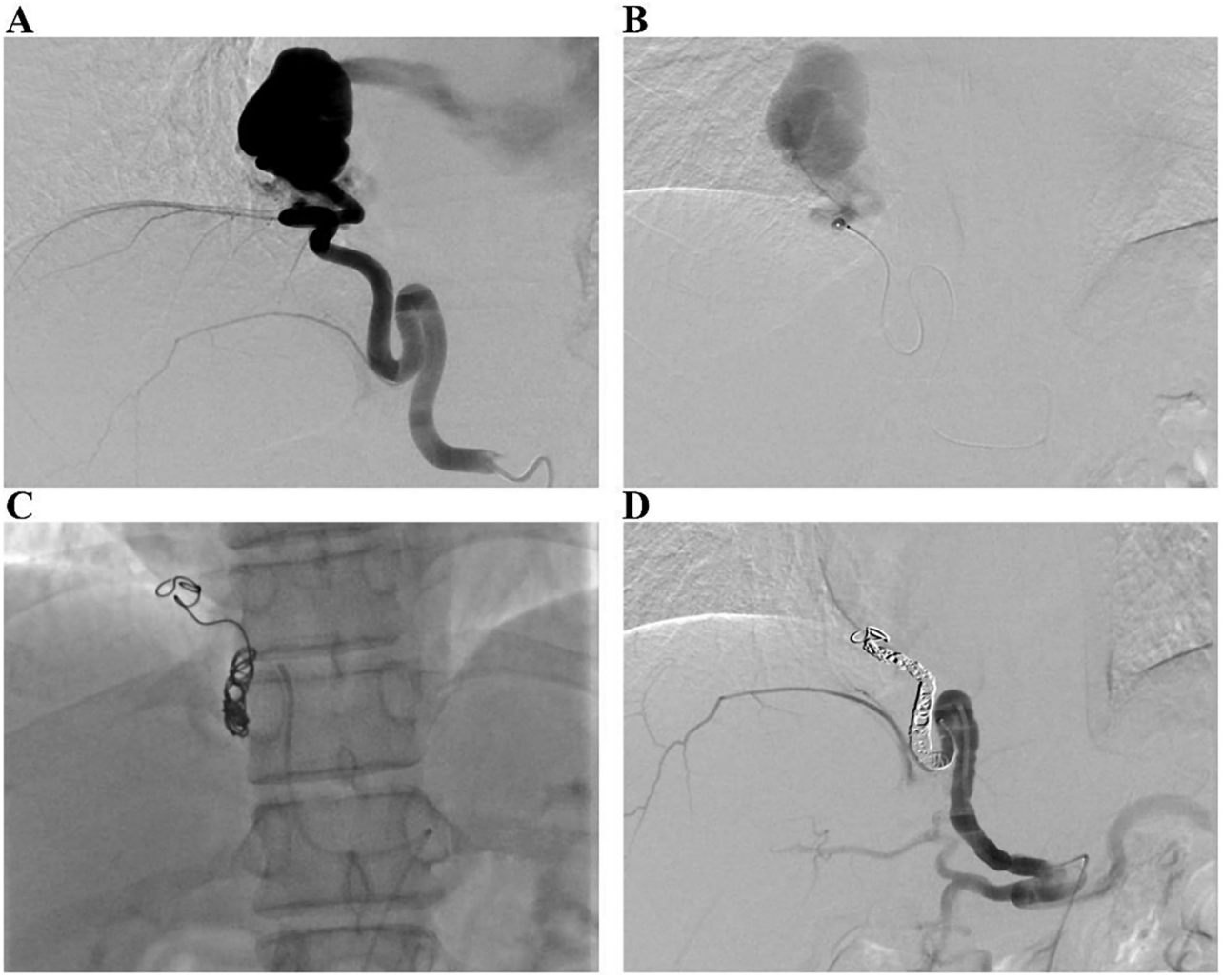

Fig. 2 Super-selective embolization of the abnormal supply of enlarged right IPA

Totally three coils (one 0.035 inch-14 cm-10 mm-NESTER and two 0.035 inch-14 cm-8 mm-NESTER) and seven micro-coils (one 0.018 inch-14 cm-8 mm-NESTER, two 0.018 inch-14 cm-6 mm-NESTER, two 0.018 inch-8/4 mm-TORNADO, and two 0.018 inch-7/3 mm-TORNADO) (Cook Medical) were consecutively deployed for IPA embolization (Fig. 2-C). The postoperative angiogram of IPA showed the PAVM was not visualized (Fig. 2-D).

After right femoral vein puncture, pulmonary angiography showed one obvious enlarged sac between one afferent PA and one pulmonary vein (Fig. 3-A). One 4 F cobra-2 catheter was selective catheterized to anchor at the afferent PA 1 cm off the detected PAVM with assistance of 7–70 cm introducer (Cook Medical) (Fig. 3-AB). Then the afferent PA was embolized by various kinds of coils (five 0.035 inch-14 cm-10 mm-NESTER, two 0.035 inch-14 cm-8 mm-NESTER, and two 0.035 inch-10/5 mm-TORNADO) (Fig. 3-C). After embolization, pulmonary angiography showed no sign of the sac of PAVM (Fig. 3-D).

The patient received three days preventive anti-inflammation and oxygen therapy after treatment. The patient was followed up for one year, and recurrent hemoptysis did not occur. TTCE and CTA were performed at one-year follow up. The CTA showed no recanalization of the embolized IPA and PA, and the sac of PAVM was significantly minimized (Fig. 4). TTCE was negative for pulmonary right-to-left shunt.

Discussion

Hypoxemia and dyspnea are the most common clinical symptoms, and they are both closely associated with the number and diameter of afferent PAs [13]. In the presented case, the diameter of afferent PA was 7.5 mm, but the patient did not suffer from hypoxemia. We considered that the enlarged IPA created a left-to-right shunt within the sac of PAVM, which would relieve the

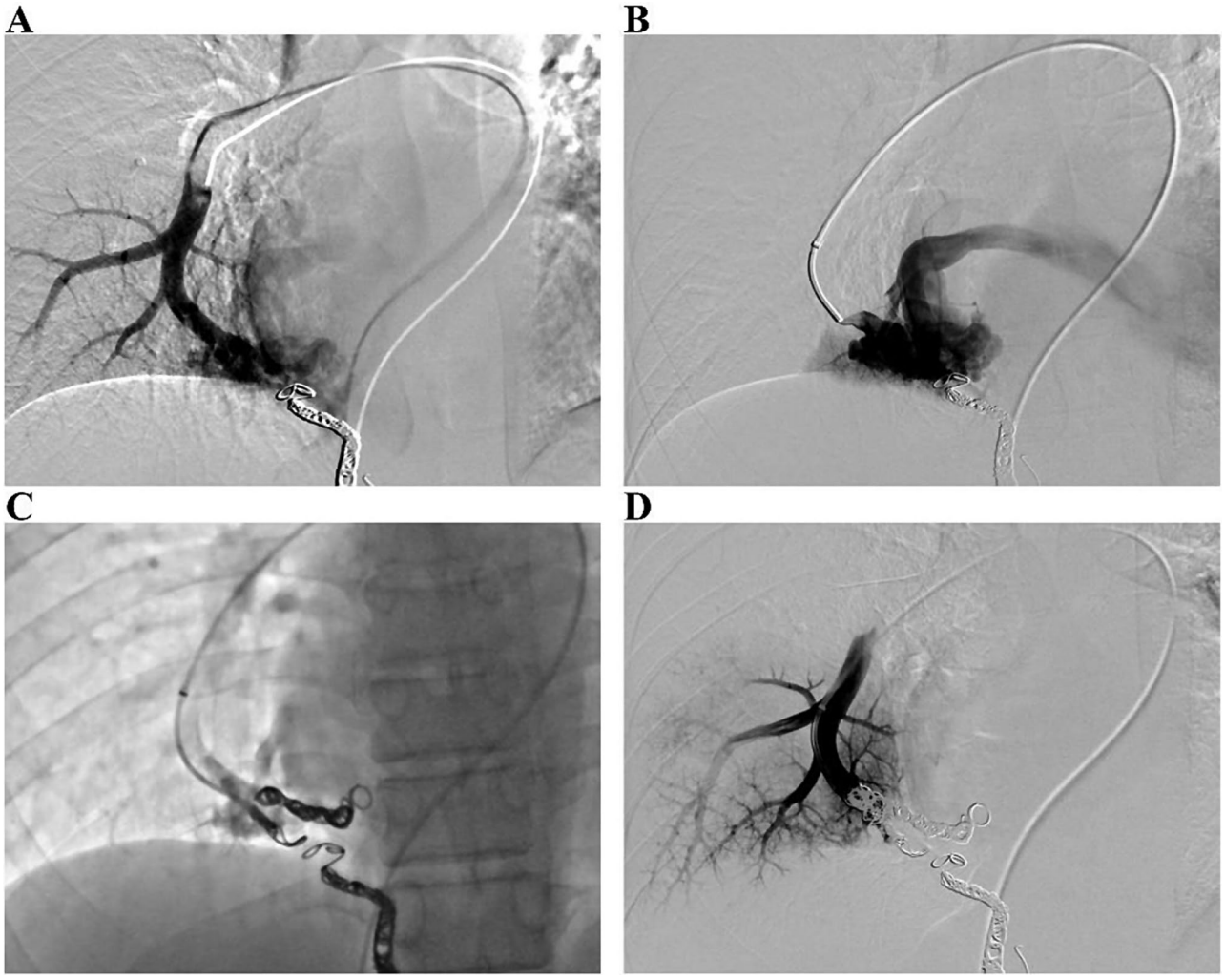

Fig. 3 Super-selective embolization of the afferent PA of the detected PAVM

right-to-left shunt in lung. The low-grade TTCE verified our assumption. Massive hemoptysis is a rare clinical symptom in PAVMs. The previous studies have described ruptured PAVM was responsible for this hemorrhage event, and the diameter of afferent PA is related to the severity of hemoptysis [14, 15]. In our report, intermittent bloody sputum was the initial symptom, and ground-glass opacities around PAVM on CTA indicated diffuse alveolar hemorrhage. Given the large diameter of the detected afferent PA did not correspond to mild hemoptysis, rupture of PAVM would not be the reason of hemoptysis in our case. We considered that high blood flow from the enlarged IPA increased the pressure within the sac of PAVM and then induced alveolar hemorrhage, which was in line with the cause of hemoptysis in anomalous systemic arterial supply to normal basal segments of the lung [16].

Systemic arteries-to-PAVM was rare. Some studies have reported that systemic arteries-to-PAVM was presented after embolization, and was the potential explanation for recurrence of PAVMs [7, 8, 17]. In addition, previous reports have reported chest trauma, surgery, or lung infection would also induce anomalous shunts between systemic arteries and PAs [18, 19]. It was believed chronic inflammatory stimulation and postprocedural local ischemia were responsible for this abnormal left-to-right shunt in lung. However, the reported patient had no definite history of chest trauma, surgery, or lung infection. The abnormal IPA-to-PAVM should be a rare congenital left-to-right shunt in the presented case.

Once confirming the afferent arteries of PAVMs, endovascular embolization was the primary treatment for all targeted arteries [17, 20]. Many embolization materials could be the option, including coils, detachable balloons, and amplatzer vascular plugs [21–23]. Distal embolization was recommended to prevent recanalization of afferent PAs, and the anchor technique and the scaffold technique were mostly widely used to prevent paradoxical embolization, vessel rupture or coil migration [24, 25]. In the present report, we also used these two

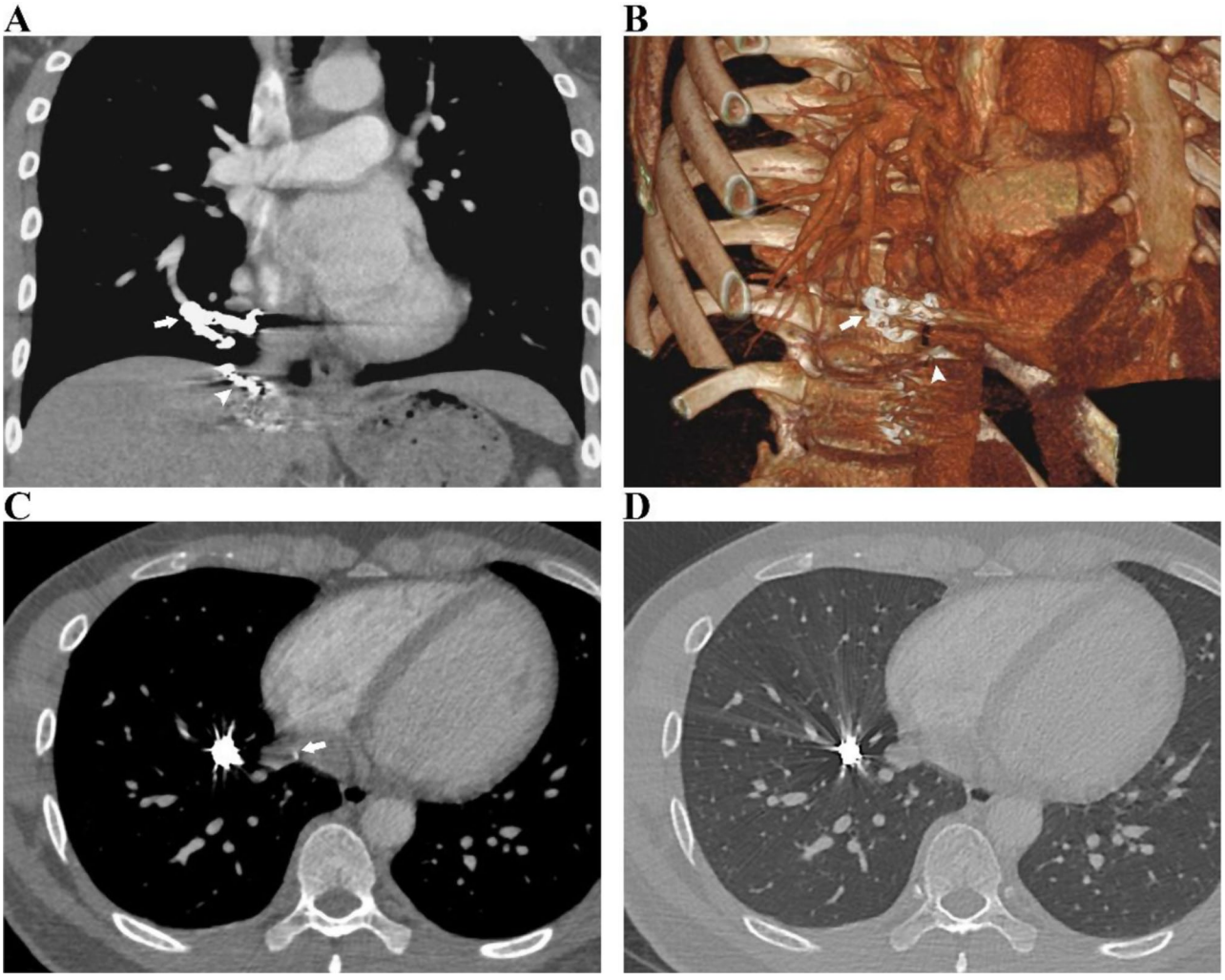

Fig. 4 **(A-B)** The coronal chest CTA image and 3D-VR image both shows no recanalization of the embolized IPA and PA; **(C)** The sac of PAVM shrinks obviously one year after embolization; **(D)** The previous ground-glass opacity around the PAVM is not detected at one-year follow up

techniques to achieve distal embolization in the detected PA and IPA. But when we deployed the first coil, we used one oversized coil to prevent coil migration in consideration of the high blood flow velocity of enlarged PA and IPA. To prevent coil migration, flow control using by a balloon catheter may also be effective. We also realized that detachable balloons, detachable coils or amplatzer vascular plugs was much safer in terms of paradoxical embolization, or coil migration. But we considered larger-size coils could also be a good option in consideration of medical cost.

Conclusions

The congenital IPA-to-PAVM was rare. Despite the supply of IPA relieved the right-to-left shunt of PAVM, the high blood flow from the enlarged IPA could also induce abnormal alveolar hemorrhage. Endovascular embolization was the safe and effective treatment for this abnormal congenital systemic artery-to-PAVM.

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Author contributions
BS and JWX conceptualized and designed case report with manuscript preparation. BS, XM, SJ were involved in the patient's care. All authors read and approved the final manuscript.

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Data availability
The data used during the current study are available from the corresponding author on reasonable request.

Declarations

Ethical approval
This study was approved by the ethics committee of Shanghai Pulmonary Hospital. Written informed consent to participate was provided by the patient.

Consent for publication
Written informed consent for publication was obtained from the patient and is available for review upon request.

Competing interests
The authors declare no competing interests.

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