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## Surgical management of pulmonary arteriovenous fistula in a female patient

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## ABSTRACT

**INTRODUCTION:** We herein describe a rare case of a pulmonary arteriovenous fistula (PAVF).**PRESENTATION OF CASE:** The patient was a 20-year-old asymptomatic female, admitted to our hospital because of an abnormal shadow in the right lung field on chest X-rays. Chest computed tomography (CT) revealed two nodules with well-defined margins in the right upper and lower lobes. Contrast-enhanced three-dimensional CT (3D-CT) revealed two enhanced solitary lung nodules which were connected with linear structures suggestive of feeding arteries and drainage veins, respectively. Based on these findings, we made a preoperative diagnosis of PAVF. We performed partial pulmonary resection of the right upper and lower lobes by video-assisted thoracoscopic surgery (VATS). The histopathological findings revealed small and medium-sized vascular channels composed of arteries with mild and irregularly thickened muscle walls and juxtaposed or seemingly anastomosing dilated veins. Based on these findings, a diagnosis of PAVF was confirmed. The patient had an uneventful postoperative course.**DISCUSSION:** A PAVF is often associated with various complications, and pregnancy could be a risk factor for these complications because of the increase in the shunt fraction. Females with known PAVF should be maximally treated prior to becoming pregnant as complications of PAVF during pregnancy can have devastating consequences. Therefore, we thought that treatment should be recommended in this case in the event she might later choose to become pregnant.**CONCLUSION:** Surgical resection using VATS for a limited number of ipsilateral isolated pulmonary arteriovenous fistulae is recommended due to its safety, low recurrence and low mortality rate.© 2015 The Authors. Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Most pulmonary arteriovenous fistulae have no symptoms and are detected as an abnormal shadow on the chest radiograph. A definitive diagnosis is made by means of pulmonary arteriography or three-dimensional computed tomography (3D-CT) angiography,

although hypervascular lesions can mimic pulmonary arteriovenous fistulae.

A pulmonary arteriovenous fistula (PAVF) is often associated with various complications, and pregnancy could be a risk factor for these complications because of the increase in the shunt fraction. We describe a rare case of a young female with a PAVF treated with video-assisted thoracic surgery (VATS).

## 2. Case report

The patient was a 20-year-old asymptomatic female. During a physical examination, no cyanosis, clubbing of the fingers nor skin telangiectasia was detected. Her pulse oximetry oxygen saturation (SpO<sub>2</sub>) was 98% on room air. No significant murmur was audible in the right mammary area. Abnormal shadows in the right upper

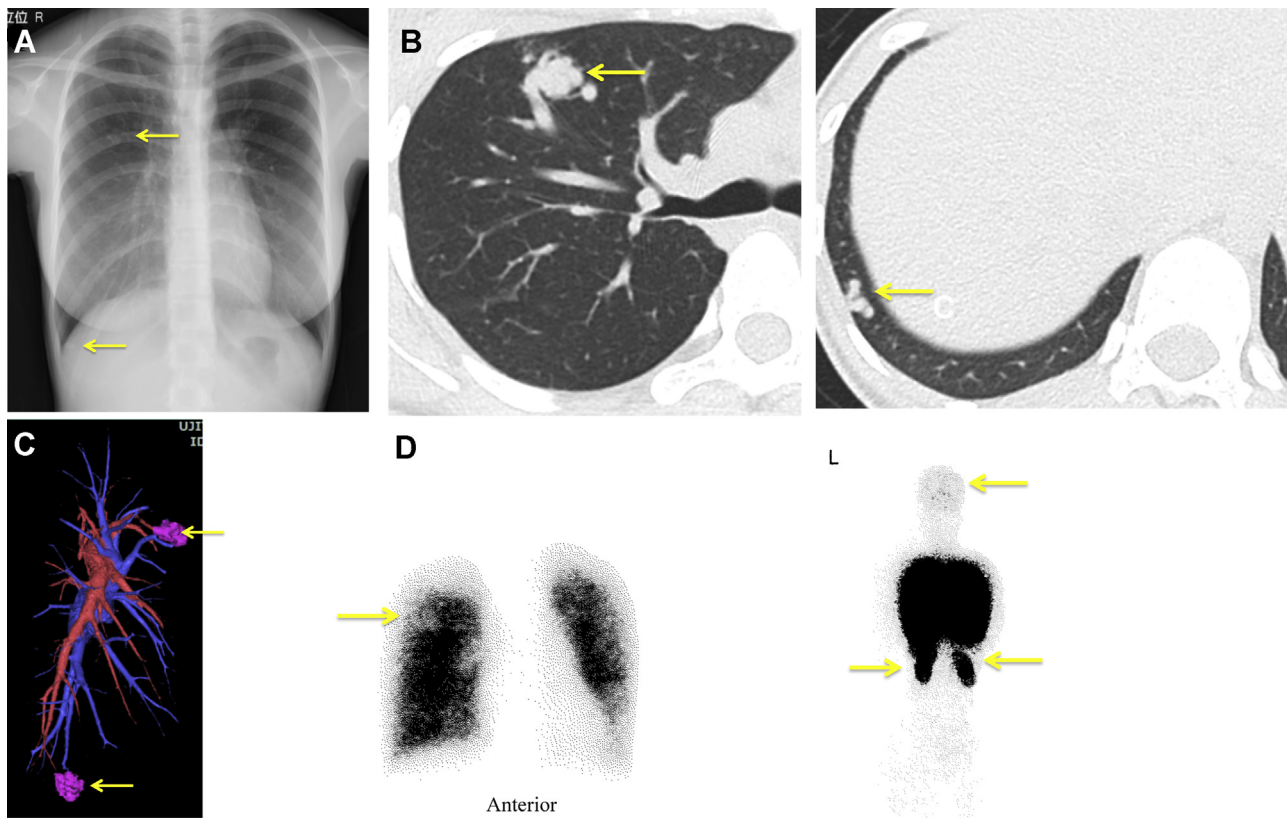
**Abbreviations:** 3D-CT, three-dimensional computed tomography; PAVF, pulmonary arteriovenous fistula; VATS, video-assisted thoracoscopic surgery; CT, computed tomography.

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**Fig. 1.** (A) Chest X-rays showed abnormal shadows in the right upper and lower lung field. (B) Chest computed tomography revealed a  $20 \times 14$  mm nodule with well-defined margins and smooth contours in the right upper lobe, and a  $15 \times 10$  mm nodule with a similar form in the right lower lobe. (C) Contrast-enhanced three-dimensional CT revealed two enhanced lung nodules which were connected with linear structures suggestive of feeding arteries and drainage veins, respectively. (D) A right-to-left shunt fraction was 15.3% and abnormal uptake was detected in the brain and bilateral kidneys by lung perfusion scintigraphy.

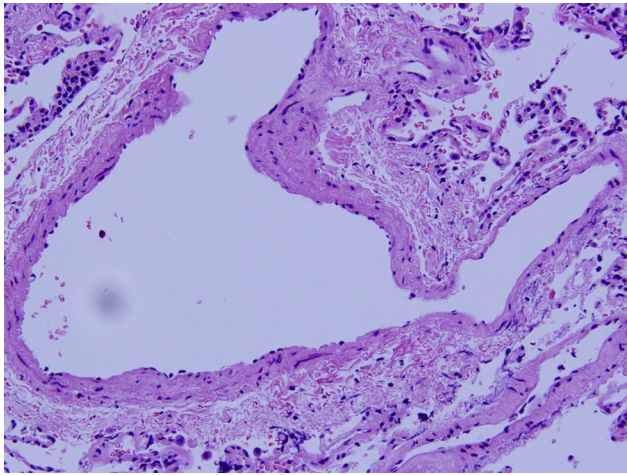
and lower lung fields were detected on chest X-rays (Fig. 1A). Chest computed tomography (CT) revealed a  $20 \times 14$  mm nodule with well-defined margins and smooth contours in the right upper lobe, and a  $15 \times 10$  mm nodule with a similar form in the right lower lobe (Fig. 1B). Contrast-enhanced 3D-CT revealed two enhanced lung nodules which were connected with linear structures suggestive of feeding arteries and drainage veins, respectively (Fig. 1C). Although an arterial blood gas analysis showed no hypoxemia, with an arterial oxygen pressure (PaO<sub>2</sub>) of 80 mm Hg on room air, the right-to-left shunt fraction was 15.3% and abnormal uptake was detected in the brain and bilateral kidneys by the lung perfusion scintigraphy (Fig. 1D). We thus made a preoperative diagnosis of PAVF. We performed partial pulmonary resection by VATS.

During the procedure, the patient was placed in the left lateral decubitus position at the first time, and the right lung was deflated. A videothoracoscope was inserted through the seventh intercostal space. Intraoperatively, pulmonary nodules were not palpable in the right upper and lower lobes, and there was no significant bruit. A nodule, which was non-tortuous in shape and covered with the visceral pleura, was detected in the right lower lobe (Fig. 2), although neither a feeding artery nor drainage vein was obvious intraoperatively. Mild telangiectasis on the visceral pleura was also observed (Fig. 2). The location of the other nodule in the right upper lobe was predicted based on the blood vessel. The histopathological findings of hematoxylin–eosin stained sections revealed small and medium-sized vascular channels composed of arteries with mild and irregularly thickened muscle walls, and juxtaposed or seemingly anastomosing dilated veins (Fig. 3). Based on these findings, a diagnosis of pulmonary arteriovenous fistula was confirmed. It was

difficult to preoperatively diagnose the PAVF, because hypervascular lesions such as those due to inflammatory changes, can also present as strongly enhanced nodules after injection of contrast material. Abnormal uptake in the brain and bilateral kidneys by the lung perfusion scintigraphy suggested a right-to-left shunt. The patient had an uneventful postoperative course and was discharged seven days after the operation.



**Fig. 2.** A nodule, which was non-tortuous in shape and covered with the visceral pleura, was detected in the right lower lobe. Mild telangiectasis on the visceral pleura was also observed.



**Fig. 3.** The histopathological findings of hematoxylin-eosin stained sections revealed small and medium-sized vascular channels composed of arteries with mild and irregularly thickened muscle walls, and juxtaposed or seemingly anastomosing dilated veins ( $\times 100$ ).

### 3. Discussion

PAVF is a relatively rare disease, which is considered to occur at a frequency of 0.02% [1]. Most of pulmonary arteriovenous fistulae are congenital and considered to be abnormal developments of the capillaries, and cases associated with Rendu-Osler-Weber have also been reported [1,2]. Our case had neither a family history of Rendu-Osler-Weber nor any symptoms. The causes of acquired or secondary pulmonary arteriovenous fistulae include chest trauma, thoracic surgery, hepatic cirrhosis, infections (actinomycosis, schistosomiasis), metastatic carcinoma and systemic disease [3,4]. This case had none of these causes, so we thought that her PAVF might have been congenital. Twenty-eight percent of cases are considered to have no symptoms [1]. It was previously reported that when a single isolated pulmonary arteriovenous fistula is 2 cm or smaller, there are no significant symptoms appear [5]. Generally, when a PAVF is 2 cm or larger, symptoms such as breathing difficulty, cyanosis, hypoxemia, finger clubbing and polycythemia, occur. Therefore, treatment is considered to be required in most cases with a PAVF larger than 2 cm in size. When the right-left shunt is 20–30% or greater, the rate of occurrence of severe complications such as rupture of the fistula, hemoptysis, cerebral infarction and cerebral abscess, is considered to be approximately 30% [1]. Although the right-left shunt of this case was 15.3%, abnormal uptake was detected in the brain and bilateral kidneys by the lung perfusion scintigraphy. The existence of the right-left shunt was obvious, so it was thought that the present case was at high risk of the complications associated with PAVF. Moreover, PAVF expand during pregnancy because of increases in the blood volume, cardiac output, and venous distensibility [6]. Females with known PAVF should be maximally treated prior to becoming pregnant, and should be alert to the potential for complications due to the PAVF during pregnancy. Therefore, because our present patient was a young female, we thought that treatment should be recommended in this case in the event she might later choose to become pregnant.

The abnormal uptake of the brain and bilateral kidneys in the lung perfusion scintigraphy disappeared after the surgical treatment. The excision was highly successful for the two ipsilateral isolated fistulae, and mortality rate is low in this surgery. Embolization is an appropriate treatment modality in cases with multiple pulmonary arteriovenous fistulae which are not suitable for surgery. The complications of embolization include pleuritic chest pain, pulmonary infection, air embolism, the migration of

coils and paradoxical embolism [1,3]. Therefore, surgical excision was preferable in the present case given her prognosis.

Pulmonary angiography or contrast-enhanced 3D-CT has been the standard diagnostic tool for pulmonary arteriovenous fistulae [7]. However, it has limitations in detecting pulmonary arteriovenous fistulae presenting as small nodules and enhanced nodules [8]. In this case, contrast-enhanced 3D-CT was effective for the diagnosis and lung perfusion scintigraphy was also supportive to obtain evidence of the right-to-left shunt fraction.

### 4. Conclusion

In conclusion, surgical resection using VATS for cases with a limited number of ipsilateral isolated pulmonary arteriovenous fistulae is therefore recommended due to its safety and the low recurrence and mortality rates.

### Conflict of interest

Yoshinobu Ichiki and other co-authors have no conflict of interest.

### Funding

None.

### Author contribution

Yoshinobu Ichiki: study design, data collections, data analysis, writing.

Junji Kawasaki: data collections.

Takayuki Hamatsu: data collections.

Taketoshi Suehiro: data collections.

Makiko Koike: data collections.

Fumihiro Tanaka: study design, data collections, data analysis.

Keizo Sugimachi: data collections.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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