

Pulmonary arteriovenous fistula diagnosed by contrast echocardiography: a case report of a daughter and a mother

Gaoyun Pan (✉ 853701271@qq.com)

Wenzhou People's Hospital <https://orcid.org/0000-0002-7109-4548>

Xinxin Dong

Wenzhou People's Hospital

Jianguo Xu

Wenzhou People's Hospital

Case report

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Abstract

Background Pulmonary arteriovenous fistula is a rare disease with a direct connection between the pulmonary artery and the vein, and in most cases is congenital. In a proportion of patients, it can cause hypoxemia, cyanosis and dyspnea. The golden standard for the diagnosis of PAVF is pulmonary angiography. We experienced two cases of a daughter and a mother with PAVF diagnosed by contrast echocardiography, which is simple and sensitive for the detection of pulmonary arteriovenous fistula.

Case presentation Case 1: A 22-year-old female was admitted to hospital because of "unconsciousness for 3 hours after sudden seizures". CT showed left frontal cerebral arteriovenous malformation with hemorrhage, a nodule of upper lobe of left lung, arteriovenous malformation possible. Intracranial hematoma removal, arteriovenous malformation resection were performed urgently. Postoperatively, the patient presented severe hypoxemia. Contrast echocardiography showed continuous dense bubbles were visualized in the left heart from the third heart cycle following imaging in the right heart, suggesting pulmonary arteriovenous fistula. Case 2 : The mother of the first patient, 44-year-old female, with no history of dyspnea, cyanosis, and stroke, was medically screened for suspected pulmonary arteriovenous fistula due to her daughter's disease. Contrast echocardiography also indicated pulmonary arteriovenous fistula.

Conclusions Contrast echocardiography is an excellent tool for the detection of pulmonary arteriovenous fistula. Patients with suspected pulmonary arteriovenous fistula should be examined by chest radiography combined with contrast echocardiography as first line screening tests, especially in patients with severe condition.

Background

Pulmonary arteriovenous fistula (PAVF) is a rare disease with a direct connection between the pulmonary artery and the vein, and in most cases is congenital. In a proportion of patients, it can cause hypoxemia, cyanosis and dyspnea. The golden standard for the diagnosis of PAVF is pulmonary angiography. We experienced two cases of a daughter and a mother with PAVF diagnosed by contrast echocardiography, which is simple and sensitive for the detection of PAVF.

Case Presentation

Case 1

A 22-year-old female was admitted to hospital because of "unconsciousness for 3 hours after sudden seizures". Physical Examination: coma, T 37.2 °C BP 137 / 80mmHg, P 125bpm SpO2 90%, bilateral pupils of equal size with diameter of 3.5mm, fixed with light reflect test, auscultation of cardiopulmonary negative, pathological signs were not drawn, cluster of papular lesions on the left breast, slightly rounded, ruby-colored, sharply demarcated from surrounding skin.

She underwent brain surgery 16 years ago with a history of epilepsy. CT of the brain and chest, CTA of the brain: left frontal arteriovenous malformation with hemorrhage, nodule of upper lobe of left lung, arteriovenous malformation possible, ground glass nodule of lower lobe of right lung, and scattered inflammatory foci in two lungs(Figure 1,2). Laboratory tests: white blood cell count: $24.7 \times 10^9 / L$; neutrophil percentage: 95.7%; red blood cell count: $4.57 \times 10^{12} / L$; hemoglobin: 153g / L; platelet count: $194 \times 10^9 / L$; C-reactive protein: 1.3 mg / L. Intracranial hematoma removal, arteriovenous malformation resection, and bone plate decompression were performed urgently. Postoperatively, symptomatic supportive treatment such as boost-capacity, sedation, anti-epilepsy, and anti-infection were given. However, the patient's condition further deteriorated, with bilateral pupil dilated, dull light reflection, unstable circulation, hypoxemia, oxygen saturation between 35–50% under pure oxygen, and PaO₂ of 54mmHg. Repeated blood routine: white blood cell count: $9.8 \times 10^9 / L$; neutrophil percentage: 84.2%; red blood cell count: $3.12 \times 10^{12} / L$; hemoglobin: 103g / L; platelet count: $196 \times 10^9 / L$. C-reactive protein: 133.9 mg / L. Repeated chest CT revealed worsening of both lung infections. Contrast echocardiography was performed with 10ml agitated saline injected into left cubital vein, while the apical four chamber view was acquired simultaneously. Contrast echocardiography showed continuous dense bubbles were visualized in the left heart from the third heart cycle following imaging in the right heart, suggesting pulmonary arteriovenous fistula(Figure 3). There were no obvious abnormalities in conventional echocardiography, no obvious abnormality on the chest radiography, and no obvious abnormality in the electrocardiogram. Because the patient cannot be moved, pulmonary angiography can't be performed to confirm the diagnosis. Despite symptomatic treatment, the patient died of respiratory failure ultimately.

Case 2

The mother of the first patient, 44-year-old female, with no history of dyspnea, cyanosis, and stroke, was medically screened for suspected PAVF due to her daughter's disease. Her arterial blood gas analysis showed PaO₂ of 38mmHg and SaO₂ of 73% under room air. Though conventional echocardiography showed no abnormal finding, contrast echocardiography revealed that continuous dense bubbles were visualized in the left heart from the third heart cycle following imaging in the right heart, which suggested PAVF(Figure 4). Accordingly, CT pulmonary angiography suggested left pulmonary arteriovenous fistula(Figure 5).

Discussion And Conclusions

PAVF refers to the direct connection of pulmonary arteries and pulmonary veins, forming a fistula or tumor-like lesion and bypassing capillaries. the disease is mostly congenital, with the incidence of 2–3 per 100,000 [1]. The clinical manifestation vary from being totally absent to severe cyanosis, congestive heart failure, and even fulminant respiratory failure[2,3]; 47–80% of patients with PAVF have hereditary hemorrhagic telangiectasia (HHT), on the contrary, almost 15 to 35% of patients with HHT have PAVF[4–6]. Also know as Rendu-Osler-Weber syndrome, HHT is a condition which is transmitted in autosomal dominant pattern, and manifested by capillary dilatation and bleeding on the surface of the skin and

mucosa and arteriovenous malformations(AVM) of the lung, brain and liver[7–9]. The first present patient, the daughter, most likely had HHT owing to pimples on the breast and cerebral AVM. Multiple complications of the central nervous system are common and may initially appear in PAVF, such as brain abscess, transient ischemic attack (TIA), migraine, and cerebral infarction, with a incidence of 10–19% [9]. The present first patient had a history of epilepsy, which could not be ruled out as a complication of central nervous system of PAVF.

The diagnostic method for PAVF includes shunt fraction measurement, chest radiography, contrast echocardiography, computed tomography, radionuclide perfusion lung scanning and pulmonary angiography. The golden standard for the diagnosis of PAVF is pulmonary angiography, while contrast echocardiography is a newer noninvasive method that appears to be very simple and sensitive for the detection of PAVF[10]. Contrast echocardiography is an excellent tool for evaluation of intracardiac and intrapulmonary right-to-left shunt(RLS), and is able to identify small shunts even when they are not confirm by other methods[11]. Because the air microbubble's diameter is greater than capillary vessel, the microbubbles can't pass through to left heart in patients without RLS. As long as the bubbles are visualized in the left heart, RLS could be verified, even though several bubbles. Barzilai et al[11] performed contrast echocardiography in nineteen patients with HHT (four with suspected PAVF). Fourteen of the patients with HHT had a positive result, of which eleven patients underwent pulmonary angiography and all had validated PAVF; only six of the 11 patient and eight were found to have abnormality with chest radiography and PaO₂ respectively. Contrast echocardiography probably has a sensitivity of nearly 100% for detecting PAVF, and has been shown to be more sensitive than plain film and CT [12]. This method needs to inject 10 ml of agitated saline (with 1ml of air) into a peripheral vein while simultaneously scanning the right and left heart with 2D echocardiography. Both solutions contain microbubbles which can yield strong reflection by ultrasound imaging compared with the normal blood flow [13].

We experienced two cases of a daughter and a mother with PAVF diagnosed by contrast echocardiography. The findings of contrast echocardiography of PAVF are as follows: delayed appearance of microbubbles in the left heart, mostly 3 cardiac cycles after right atrial bubbles is visualized; dense continuous microbubbles in the left heart, with less change with Valsalva manoeuvre; when the shunt is of large amount, the left heart contrast agent is still present when the right heart contrast agent subsides; visualization of bubbles in the pulmonary veins is direct evidence of pulmonary arteriovenous fistula. Typical intracardiac RLS (PFO or ASD) are usually seen in the first three cardiac cycles following the visualization of right atrial microbubbles, and intracardiac RLS amount depends on the pressure between the right and left atrium. Intracardiac RLS occurs only when the right atrial pressure surpass the left atrium, which is obvious with Valsalva manoeuvre[14]. Normally there is a tiny amount of intracardiac RLS by contrast echocardiography, which is easily distinguished from PAVF. Theoretically, it is supposed that the shunt through pulmonary circulation in PAVF occurs later than that in PFO, which takes more time for the contrast agent to pass from the pulmonary artery through PAVF to the pulmonary vein. Occasionally, the shunt in PAVF could occur within three cardiac cycles, and the shunt in PFO can occur later than three cardiac cycles. The shunt in the present patients with PAVF both happened at the third cardiac cycle. Therefore, PFO and PAVF cannot be reliably differentiated depending on the emerging

time of shunt alone, while the appearance of dense continuous bubbles is more reliable and specific for the diagnosis of PAVF.

The intrapulmonary shunt revealed by contrast echocardiography requires additional assessment of PAVF, which generally should be digital subtraction angiography. We recommend that patients with suspected PAVF should be examined by chest radiography combined with contrast echocardiography as first line screening tests, which can be performed bedside when necessary. If positively, advanced invasive examination such as digital subtraction angiography should be take into account to validate the diagnosis. Because of the severity condition, the daughter could not undergo pulmonary angiography and further lobectomy or embolotherapy. The severe hypoxemia of the first present patient may be caused by complication of PAVF and cerebral AVM such as PAVF rupture, Sequelae of hemorrhage of cerebral AVM, or deterioration of lung infection. Unfortunately the severe hypoxemia could not be reversed ultimately, which could have been avoided if the PAVF and cerebral AVM was recognized and treated earlier. For the mother, we advised that she take a pulmonary angiography, if necessarily, surgery or embolotherapy should be considered.

Conclusion

Contrast echocardiography is an excellent tool for the detection of pulmonary arteriovenous fistula. Patients with suspected PAVF should be examined by chest radiography combined with contrast echocardiography as first line screening tests, especially in patients with severe condition.

Declarations

Ethics approval and consent to participate

The case report was approved by the ethics committee of Wenzhou People's Hospital. Written informed consent was obtained.

Consent for publication

Written informed consent of publication was obtained.

Availability of data and materials

The data and images used and analysed during the case report are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests

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Authors' contributions

GP performed the contrast echocardiography and drafted the manuscript. XD performed the conventional echocardiography. JX collected the patients' clinical history.

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Figures

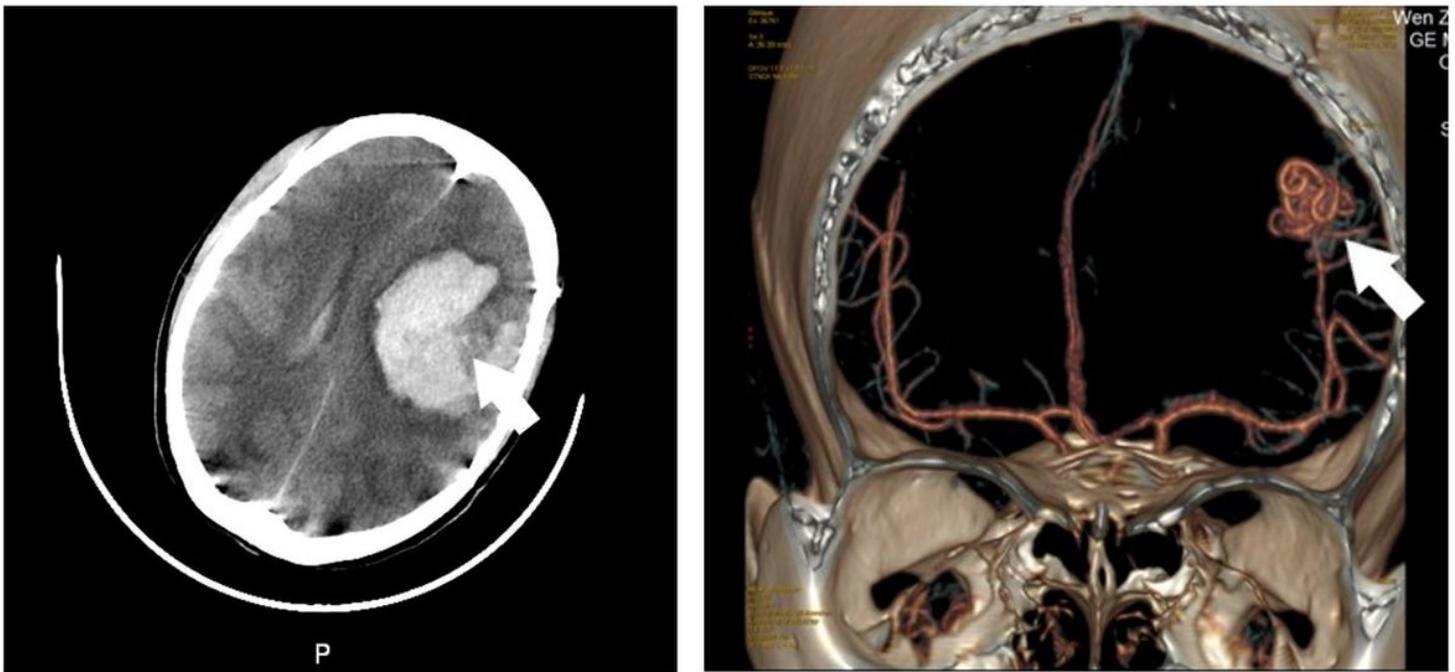


Figure 1

Left panel: cerebral CT showed left frontal high density shadow indicating hemorrhage(white arrow) ; right panel: 3D reconstruction of cerebral arteries indicating arteriovenous malformation(white arrow).

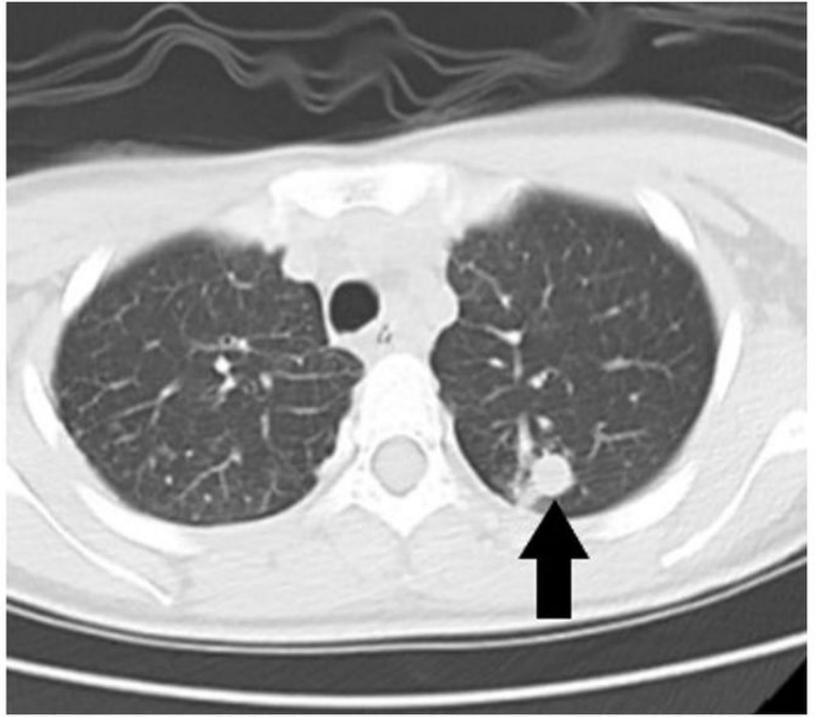


Figure 3

Left panel: No significant abnormal finding showed in chest radiography. Right panel: thoracic CT showed a nodule of the upper lobe of left lung, arteriovenous fistula possible (black arrow).

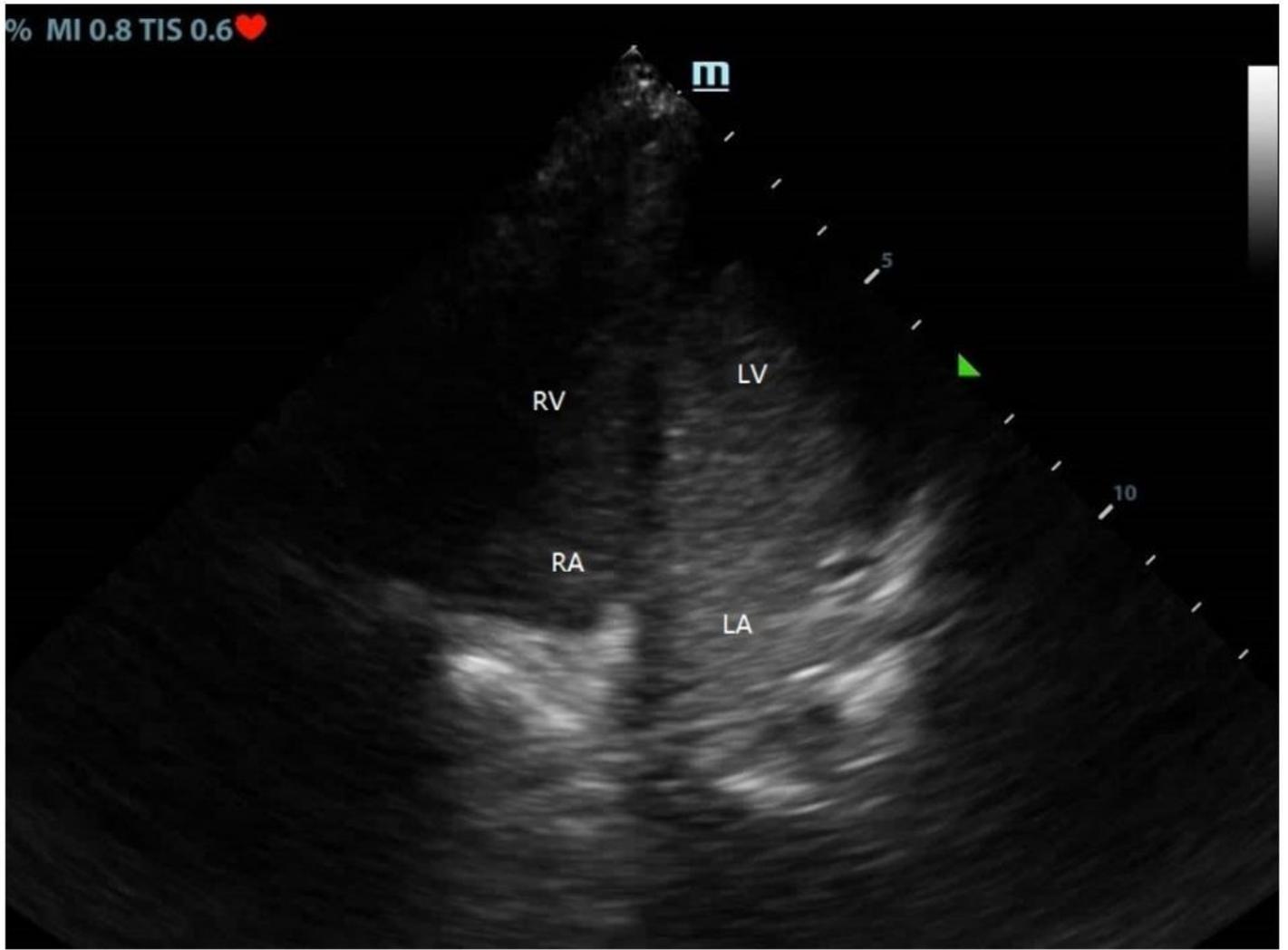


Figure 5

Contrast echocardiography showed dense microbubbles the left heart from the third cardiac cycle following the imaging of microbubbles of the right heart, suggesting pulmonary arteriovenous fistula.

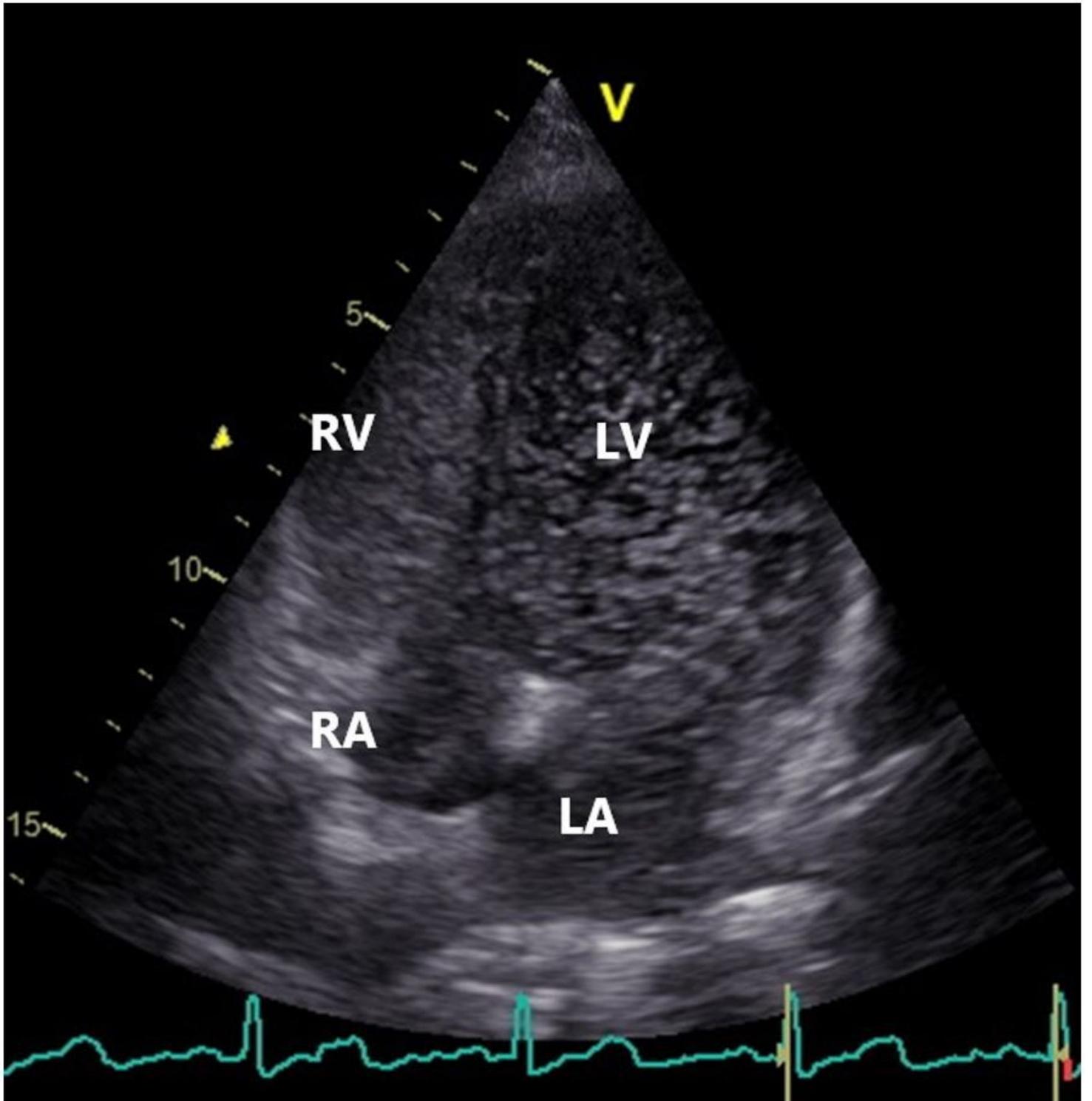


Figure 7

Contrast echocardiography showed dense microbubbles the left heart from the third cardiac cycle following the imaging of microbubbles of the right heart, suggesting pulmonary arteriovenous fistula.

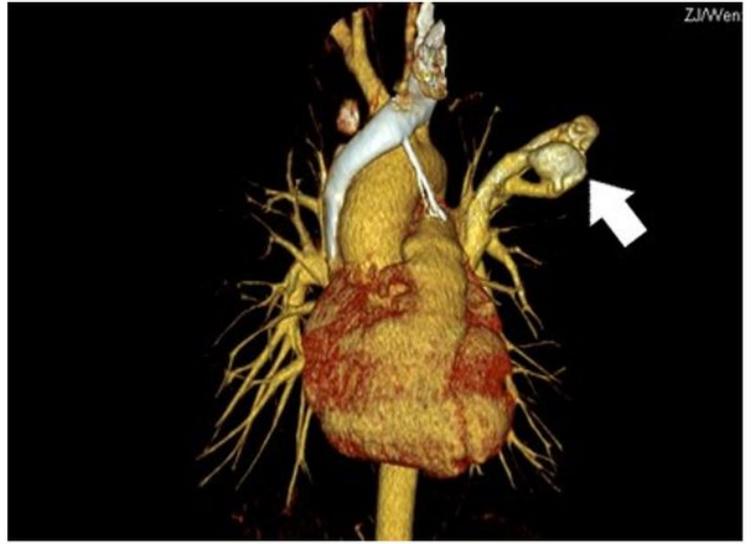
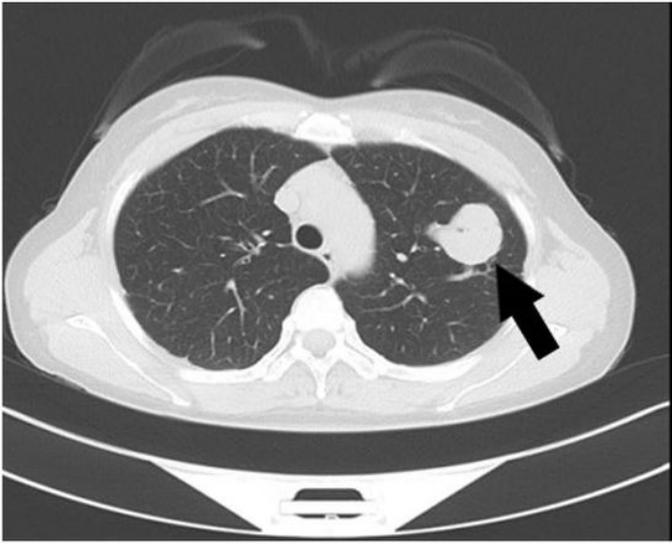


Figure 9

Left panel: thoracic CT indicated left pulmonary arteriovenous fistula [black arrow]; right panel: thoracic CT angiography indicated left pulmonary arteriovenous fistula (white arrow).