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CASE REPORT

Successful treatment of multiple pulmonary arteriovenous fistulae with thoracoscopy

Yufei Wang[1*], Ke Wang[2*], Chunyan Guo[3] & Zhanlin Guo[1]

1 Department of Thoracic Surgery, Affiliated Hospital of Inner Mongolia Medical College, Hohhot, Inner Mongolia, China
2 Department of Thoracic Surgery, 253 Hospital The People's Liberation Army, Hohhot, Inner Mongolia, China
3 Department of Anesthesiology, Affiliated Hospital of Inner Mongolia Medical College, Hohhot, Inner Mongolia, China

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Correspondence
Zhanlin Guo, Department of Thoracic Surgery, Affiliated Hospital of Inner Mongolia Medical College, 1 North Valley St, Hohhot, Inner Mongolia 010010, China.
Tel: +86 471 345 1489
Fax: +86 471 345 1489
Email: nmggzl@163.com

*These authors contributed equally to the paper.

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Abstract

Congenital pulmonary arteriovenous fistulae occur as a result of abnormal blood vessel development in the lungs. Blood takes a short pass from the pulmonary artery to veins. Multiple pulmonary arteriovenous fistulae are a rare occurrence, especially when involving both lungs. Fistulae located at the edge are prone to rupture and bleeding. We discuss a case of a 15-year-old overweight male with multiple pulmonary arteriovenous fistulae successfully treated with wedge-shape excision via video-assisted thoracoscopic surgery.

Introduction

Congenital pulmonary arteriovenous fistula is a disease present at birth and is usually associated with abnormal development of blood vessels in the lungs. Blood directly flows from the pulmonary artery to pulmonary veins, skipping the alveoli. It was first discovered by Churton in 1897, and was named multiple pulmonary artery aneurysm;[1] it was later proven by Smith via angiocardiography in 1939. In 1942, Hepburn and Dauphinee performed the first pulmonary arteriovenous fistula surgery.[2] The application of embolization to treat pulmonary arteriovenous fistula was reported for the first time in 1978.[3] Congenital pulmonary arteriovenous fistula is an autosomal dominant genetic disease and is passed down in the family. The Rendu–Osler–Weber variant, for example, is associated with a 9q3, 12q gene mutation on the chromosome.[4] Patients exhibiting symptoms with a limited pathogenic area could be treated with surgery or interventional therapy; however, patients without any symptoms require careful attention. Hemorrhaging caused by a sudden rupture of the veins could be fatal. Surgical approaches need to be adjusted according to the pathogenic area and type. Lung excision is the most common approach, such as wedge-shape excision of the lung, lobectomy, and pneumonectomy. Surgical options follow the principle of minimizing tissue removal and preserving maximum lung function.[5]

Case presentation

In September 2016, a 15-year-old overweight male attended the emergency room after experiencing difficulty breathing and chest pain for an hour. The patient came in a semi-reclined position and showed stable life signs. A routine blood examination indicated: hemoglobin 154.0 g (normal range: 120–172); hematocrit 45% (normal range: 36–50%); arterial blood gas PaO_2 58 mmHg (normal range: 83–108);

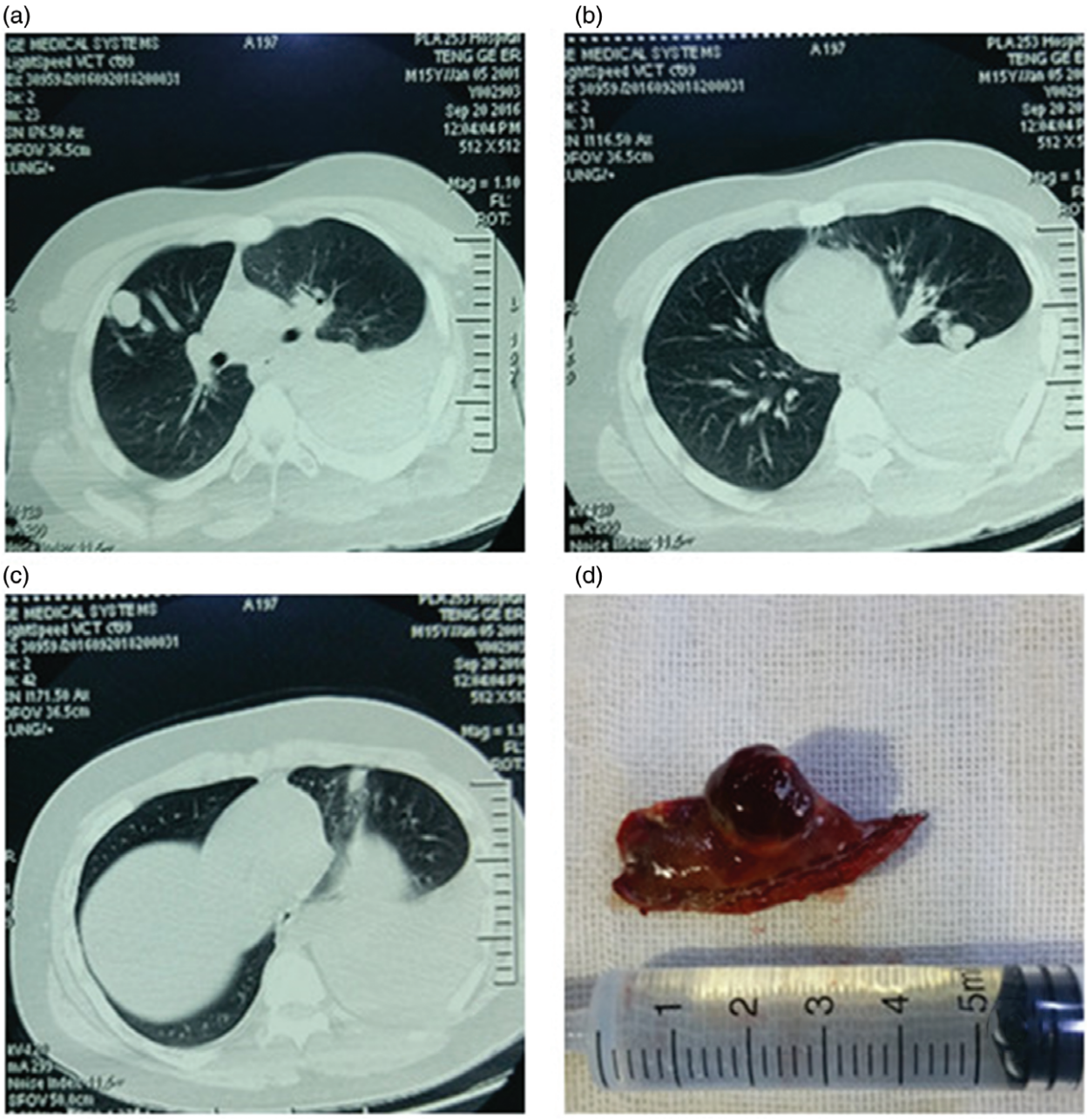

Figure 1 Chest computed tomography images taken before emergency surgery: (**a**) right upper lobe; (**b**) pleural effusion in the left chest; (**c**) lingual segment of the left upper lobe; and (**d**) sliced focal pathogenic lung.

and SO_2 90% (normal range: 95–98%). Chest computed tomography (CT) showed clots in both lungs and a great deal of pleural effusion in the left chest (Fig 1). Uncoagulated blood was removed via conventional thoracentesis. The patient had been diagnosed with multiple lung pulmonary arteriovenous fistulae a year ago but had chosen not to receive treatment. Fistulae were found in the right upper, left upper, and left lower lobes (Fig 2).

When the patient was hospitalized, a diagnosis of multiple lung pulmonary arteriovenous fistulae was confirmed. Thoracoscopy was performed in the emergency room. An observational incision was made in the seventh rib midaxillary line and a 3.5 cm surgical incision in the fourth rib of the left anterior axillary line. Video-assisted thoracoscopic surgery (VATS) revealed approximately 2000 mL of blood and blood clotting around the left chest. A fistula was

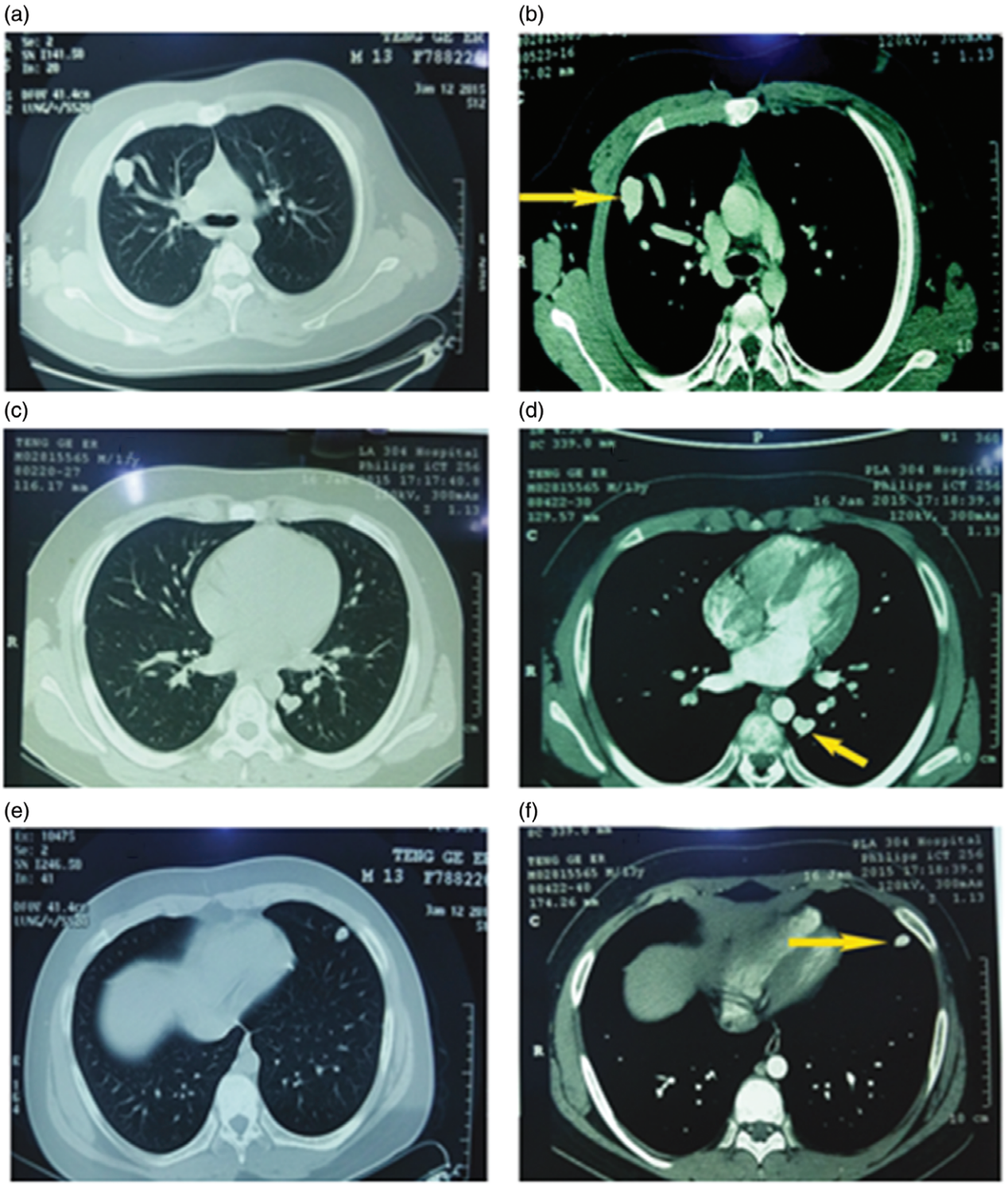

Figure 2 Enhanced computed tomography images: (**a**,**b**) right upper lobe; (**c**,**d**) left lower lobe and posterior segment; (**e**,**f**) lingual segment of the left upper lobe and anterior basal segment.

found on the lingual segment of the upper lobe located at the edge of the lung just below the visceral pleura. A purple fistula approximately 1 cm in diameter was jutting out of the surface with tension, and had ruptured 0.2 cm. Straight-line cutting with a suture instrument was used to remove the lingual segment of the upper lobe of the left lung via a wedge-shaped incision. A red fistula was also detected on the left lower lobe of the lung located at the posterior segment interface of the lower lobe and segment in lung parenchyma, but had not ruptured or bled.

Discussion

Fistulas may exhibit diverse symptoms, including repeated hemoptysis, nose bleeds, difficulty catching breath, an increase in hemoglobin level[6,7] or cerebral infarction or abscess. In this case, the patient was hospitalized for left pyohemothorax, an increased level of hemoglobin, drumstick finger, fatigue, and hypomnesia.

Enhanced CT is a noninvasive approach with high accuracy and has gradually replaced pulmonary angiography as the primary technique for treating fistula.[8] As the patient in our case was diagnosed via enhanced CT a year ago with multiple pulmonary arteriovenous fistulae, a regular chest CT scan was taken before surgery.

Pulmonary arteriovenous fistula is the result of blood flow from the pulmonary artery to the pulmonary veins, leading to a higher level of unsaturated oxygen in the veins; however ventilation is not interrupted. Pco_2 usually remains normal at a normal level. Most cases have increased red cells induced by the abnormally low level of oxygen. Complications such as bacterial infections and brain abscess may occur because of direct intercommunication between systemic and lung circulation.[9] Although our patient had around 2000 mL of chest bleeding, the hemoglobin and red cell hematocrit were normal. This may be attributed to the increase in hemoglobin level.

Although fistulae are benign, they develop over time, particularly in teenagers. Fistulae, particularly pulmonary arteriovenous fistulae, do not diminish and require treatment.[10] Currently, primary treatment methods include surgery and transcatheter embolotherapy. Surgery is helpful to completely remove fistulae, including partial or local excision of the lungs, lobectomy, and pneumonectomy. The surgical technique used depends upon the location of the pathogenic area. If a fistula is located on the surface and below the pleura, wedge-shaped excision of the lung is recommended. If fistulae are located deeply or in multiple locations, lobectomy is the best option. The decision to perform pneumonectomy should be made with extreme caution. A doctor must ensure that the other lung is fully functional. A lung transplant is recommended if a patient has a diffusive and extensive pathogenic area. There are greater disadvantages of conducting a lung transplant (such as the large wound required), particularly in children as their lungs are still developing. Currently, angiocardiography is the first option.[11] It is effective for single pulmonary arteriovenous fistula or partially complicated pulmonary arteriovenous fistula. Recurrence as a result of recanalization of fistulae occurs in 5–10% of patients; thus, patients should be followed-up periodically.[12–15]

The patient in our case was aware of his condition but left it untreated for a year, which can lead to the dangerous situation of ruptured fistula. Multiple pulmonary arteriovenous fistulae rarely occur, particularly in both lungs. Fistulae located at the side of lung are prone to rupture and bleeding. Patients usually seek medical attention for chest pain and shortness of breath, hydropneumothorax, and pleural effusion. In our case, pulmonary arteriovenous fistulae developed in three locations. Because the focal lingual segment was located underneath visceral pleura, there was a greater likelihood of rupture as the fistula grew outwards. Wedge-shaped excision of the lung was performed via VATS to treat the rupture.

A single focal fistula located next to the pleura should be removed via VATS without delay. Pathogens near this location may transform into hemopneumothorax, increasing the fatality rate. In our patient, a focal fistula was also located on the right lung next the to pleura. Performing simultaneous surgery increases risk; therefore embolization is recommended to remove other focal fistulae separately. Embolization blocks the fistula and the flow of blood. It requires a smaller wound than surgery, preserves normal lung tissue,[16] can be performed multiple times, and is an easier procedure. Complications associated with embolization include pulmonary infarction, lesion reperfusion, embolism transfer, or drop out and leads to distal body circulation of embolism displacement or embolism of the pulmonary artery at other locations.[17] Generally, these complications occur at focal fistula with a larger blood flow volume or in shorter veins with larger diameters.[18] Long-term follow-up should be conducted in patients treated via embolization. The relapse rate is reported as 5–15%,[19] especially in children. Relapse occurs after vascular recanalization of the embolism, if the blood-supply vein was not found before surgery, or after the formation of vascular branch circulation near an embolism.[20]

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