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Treatment for hemoptysis after percutaneous closure of a large intralobar pulmonary sequestration in elderly patient

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Pulmonary sequestration is a rare congenital cardiovascular malformation.[1,2] Open surgical excision is the usual treatment, although thorascopic intervention and transcatheter embolization have been reported. However, only a limited number of articles involving percutaneous closure the rare pulmonary malformations have been published in the literature.[3] What's more, previous studies involving percutaneous pulmonary sequestration closure with repeated respiratory symptoms at the very young age suggested that the optimal therapeutic strategy has yet to be determined.[4] We hereby describe a particular case of percutaneous closure of a large intralobar pulmonary sequestration at elderly age.

A 62-year-old man who complained of recurrent hemoptysis after activity for 20 years. On examination, there was a normal dynamic pulse at the rate of 88/min and blood pressure measured with appropriate size cuff of 153/94 mmHg. There was no cyanosis. Also there were no signs of heart failure. Cardiovascular examination revealed no abnormal palpable heart sounds. The complete blood profile was normal. The ECG showed left ventricular hypertrophy without left atrial enlargement. Echocardiography revealed that intra-cardiac anatomy was normal without any shunt lesions. But there was left ventricular and ventricular septum hypertrophy for his age but a normal ejection fraction. Chest CT revealed that the veins of the inferior lobe of the left lung increased, and flake ground glass density was seen. Abnormal vessels and ring calcification were found from descending aorta under mediastinal window. The diagnosis of the case was not clear hence CT angiography were done, and 3D volume rendering images were reconstructed revealed that congenital pulmonary malformations, the left inferior pulmonary artery originated from the descending aorta in morphology. The local lung tissue is separated from the left lung and supplied by anomalous systemic artery. There was normal pulmonary venous supply to both the lungs without dilated and hypertrophied. Therefore, the diagnosis of a large left intralobar pulmonary sequestration was made.

The patient was subjected to cardiac catheterization and angiography for further confirmation of such a rare diagnosis. After local anesthesia, the right femoral artery and vein were punctured, then the internal and external sheath were placed. The coronary angiography was performed through the right femoral artery and no stenosis was found in the left main trunk, the anterior descending branch, the circumflex branch and the right coronary artery. The pressure of the left/right pulmonary artery and right ventricle were measured through the femoral vein by end-hole catheter (43/22 mmHg, 40/18 mmHg and 42/0 mmHg respectively). Pulmonary angiography showed that the left inferior pulmonary artery was relatively low profile and dysplasia. Aortography was performed at the descending aorta, and abnormal vessels of the left inferior pulmonary artery were seen from the aorta at about one rib distance above the diaphragm. Hand injection of the contrast medium showed that the abnormal vessels were seriously tortuous and anatomically deformed without drainage into pulmonary veins and left atrium. The main extra- and intra-orifice of the abnormal vessels was 16.8 mm and 10 mm respectively, and the length was 15 mm. Many attempts using end-hole catheter, 5 or 6 Fr. Judkin's right guiding catheter, J-guide wire and loach guide wire failed to establish an arteriovenous orbit through this segment of the vessel.

According to the special shape of the abnormal vessels, an 8F EBU (Medtronic, Medical, Minneapolis, Minnesota, USA) guiding catheter along the guide wire was sent into abnormal vessels by retrograde approach via femoral artery. The abnormal vessels were closed with embolization of a 14 mm AVP Ⅱ (Amplatzer vascular plug Ⅱ, AGA Medical,
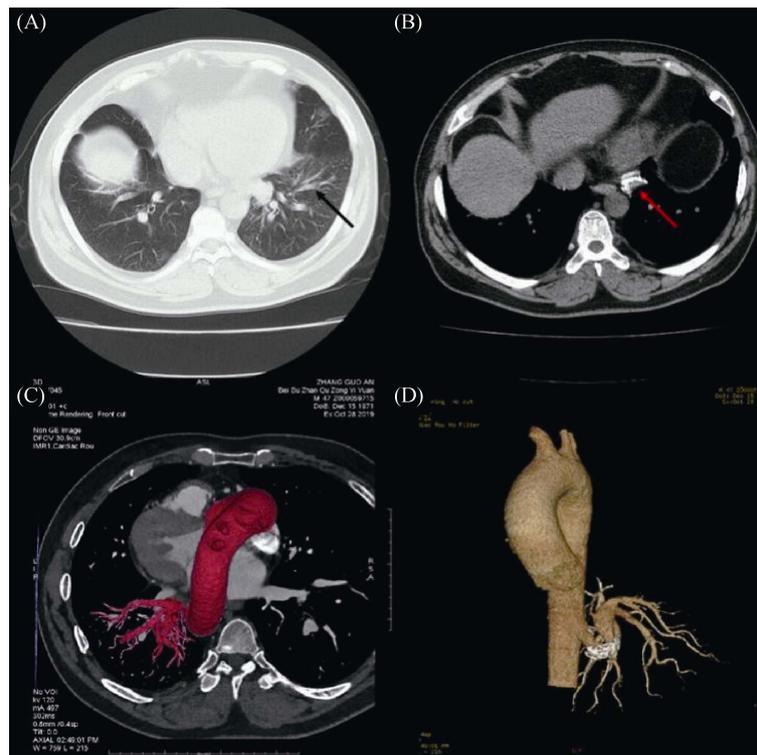
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Figure 1. Chest CT and CTA preoperative evaluation. (A): The veins of the inferior lobe of the left lung increased, and the density of flake ground glass (black arrow) was observed; (B): abnormal vessels and ring calcification (red arrow) were found from descending aorta under mediastinal window; (C): CTA revealed congenital pulmonary malformations supplied by anomalous systemic artery at coronal section view; and (D): the left inferior pulmonary artery originated from the descending aorta in morphology at sagittal CTA view. CT: computed tomography; CTA: computed tomography angiography.
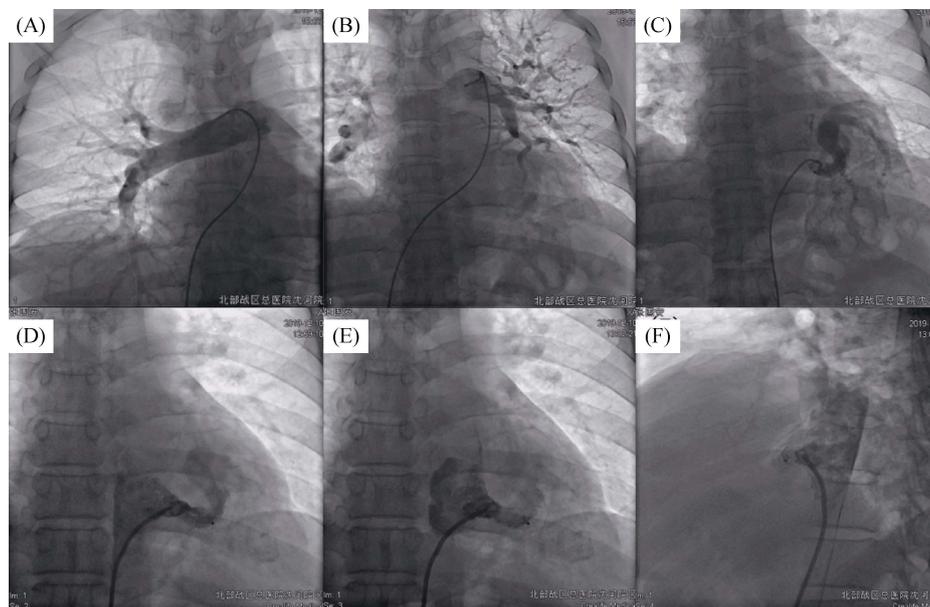

Figure 2. The angiography video review. (A): No obvious abnormality was found in right pulmonary arteriography; (B): the left inferior pulmonary artery was relatively low profile and dysplasia in left pulmonary arteriography; (C): the large abnormal vessels of the left inferior pulmonary artery in morphology were found from the descending aortography; (D): the ACP Ⅱ was successfully deployed and moderate residue shunts was found after contrast injections immediately; (E): adaptable shape of discs of the occluder with mild residual leak was confirmed angiographically at 10 min later; and (F): a stable position and adaptable shape of discs of the occluder without residual leak was confirmed angiographically at 20 min later. ACP Ⅱ: Amplatzer vascular plug Ⅱ.
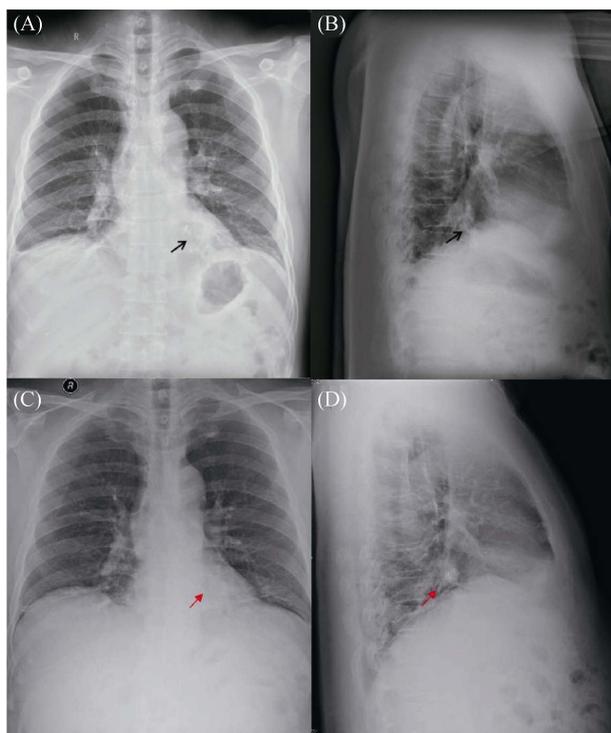
Figure 3.   X-ray evaluation during follow-up. (A & B): a stable position and adaptable shape of discs of the occluder (black arrow) was confirmed under postoperative eutopic and lateral X-ray examination; and (C & D): no displacement of the occluder (red arrow) and other complication was observed at 6-month eutopic and lateral X-ray follow-up.

Golden Valley, MN, USA). Before the occluder released, a stable position and adaptable shape of discs of the occluder without obvious residual leak was confirmed angiographically at 20 min later. The device was successfully deployed. No significant pressure difference was found in the continuous pressure curve monitoring of descending aorta along transcatheter closure device. Access site was closed using manual compression without complication. The optimal position of the occluder without dislodgement was confirmed by first postoperative 24 h X-ray examination. The patient was discharged in stable condition on the third day after closure uneventfully. The 6-month follow-up completed and there was no displacement of the occluder. The patients had good cardiopulmonary condition without hemoptysis symptoms during the follow-up.

As hypoplasia of the lung, a part of the lung tissue is separated from the normal lung and supplied by anomalous systemic artery without gas exchange function. Pulmonary sequestration can be divided into two types: intralober type and extralobar type. The intralober type and the normal lung tissue are wrapped under the same pleura, which is more common clinically. If the pulmonary parenchyma lacks any

communication with the airways, the patients might remain asymptomatic for years. Once communicating with bronchus, symptoms are often non-specific, including chest pain, dyspneoa, and recurrent pulmonary infections symptomatic. The extralobar type is wrapped under its own abnormal pleura, which is relatively independent from the normal lung and asymptomatic, but 50% of them can be combined with other congenital malformations. The venous drainage usually is normal, but anomalies can sometimes occur, with drainage either directly to the right atrium, or to one of the major systemic veins of the thorax or abdomen. This latter feature being seen in extralobar pulmonary sequestrations and the scimitar syndrome.[5]

In this case, the patient with pulmonary sequestration was easy to be misdiagnosed. It may be asymptomatic for years, albeit that signs of recurrent infection, such as fever and productive cough, are more common as reported. Pulmonary sequestration can also be a rare cause of significant left to right shunting, with the subsequent left ventricular volume overload producing signs of congestive cardiac failure.[6] However, left ventricular hypertrophy was detected by EGG and TTE due to hypertension history in our case. We guess the reason of this patient presented the only recurrent hemoptysis rather than the respiratory infections symptoms was the hemodynamic mechanism of immune inflammatory factors in local unique anatomy in elderly age, which needs further investigation. We confirmed this was an intralober type pulmonary sequestration as the cardiac catheterization and angiography results and venous drainage direction. Clinically, pulmonary sequestration should be differentiated from anomalous origin of pulmonary artery and pulmonary arteriovenous fistula. The anomalous origin of pulmonary artery often leads to pulmonary vascular obstructive disease and severe pulmonary hypertension caused by shunt at very young age. The pulmonary arterio-venous fistula is that the lung has normal bronchial connections and pulmonary arteries which cause cyanosis as venous blood is shunted into the systemic circulation and distinguish it from pulmonary sequestration.[7]

To our knowledge, this is the first case of such elderly patient with large intralobar pulmonary sequestration, who received percutaneous closure using AVP Ⅱ occulder. Correction by surgery remains the gold standard therapy.[8] Most of the previous literature reports of percutaneous closure were more younger patients. The contrast medium dose and the size of delivery sheath were the main factors restricting the success of interventional therapy. However, vascular tortuous and calcification in elderly patients was obvious, in order to improve the success rate of interventional therapy, we need to change a variety of interventional

appliances according to local anatomy conditions. Flexibility of the articulations and lack of patches inside the AVP Ⅱ occluder allow this relatively soft device to adapt to the patient's anatomy rather than distorting the anatomy to its shape.[9] AVP II has been used in various low- and high-flow lesions including venous collaterals, modified Blalock Taussig shunts, porto-systemic connections, aorto-pulmonary collaterals.[10] The AVPs family offers different sizes to fit a wide range of vascular diameters.[11] There were indications of interventional therapy. Due to the tortuous route of branch vessels, it is difficult to get the guide wire and catheter in destination place. In this case, the proximal part of the abnormal vessel opened upward and the EBU guiding catheter was inserted through the femoral artery, which the coaxiality was good, and the occluder was easily to be precise placed and released. For this kind of single large branch of arterial blood supplied to the lesion of pulmonary sequestration, the choice of vascular plug was ideal. The proximal pressure of the occluder from the aorta after occlusion was much higher than that of the distal pressure, the possibility of occluder falling into the systemic circulation was very low. The diameter of target vessel measured by angiography plus 2–6 mm was recommended for the size of the plug selection. In our case, we successfully used the 14 mm AVP II through an 8Fr EBU guide catheter, which did not cause any complication.

In conclusion, percutaneous large intralobar pulmonary sequestration closure is a viable option for elderly patients with hemoptysis but there are technically difficult considerations to vessels tortuous and calcification. The differenttial diagnosis can be made by CT angiography, cardiac catheterization and cardiovascular angiography. In the case of this elderly patient, it was suggested that carefully observing the condition of abnormal vessels, and proper interventional appliances selection might contribute to procedural success.

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