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Case Report

Giant branching aneurysmal aberrant systemic artery for intralobar pulmonary sequestration: Computed tomographic depiction of arterial and bronchial anomaly

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Abstract

Aberrant systemic arteries supplying the intralobar pulmonary sequestration can become dilated and have atherosclerotic change. Computed tomography is very useful in demonstrating the aberrant artery. We report a case of intralobar pulmonary sequestration with giant branching aneurysmal aberrant artery, and demonstrated the discontinuity of the bronchus by 64-slice computed tomography, which has not previously been described.
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1. Introduction

Pulmonary sequestration, which is defined as nonfunctioning lung tissue supplied by anomalous systemic artery and lacks normal communication to the tracheobronchial tree, is the most common congenital abnormality of lung. Anomalous systemic arteries supplying low-resistance pulmonary parenchyma often become dilated and tortuous. We report a case of intralobar pulmonary sequestration having a giant aneurysmally dilated aberrant systemic artery that was larger than aorta and that had marked dilated branches diagnosed by using 64-slice computed tomography (CT). We also demonstrated the discontinuity of bronchus, which is essential in differentiating intralobar pulmonary sequestration to "anomalous systemic artery to normal left basal lung". To our knowledge, none of the three aforementioned features have been reported.

2. Case report

A 46-year-old woman who had been told that she had a mass lesion in the left lung, 10 years before, and suffered coughing for 1 month. The patient had no prior history of significant medical disease and no habit of smoking. Physical examination showed mild rhonchi in the left lung. Laboratory examination for carcinoembryonic antigen was within the normal limits. Chest radiograph revealed a mass in the retrocardiac area. CT was performed by using a 64-slice CT scanner (Aquilion 64; Toshiba Medical Systems, Tokyo, Japan) with 100 mL of iodinated contrast injection of a rate of 3.0 mL/s; the image demonstrated that the "mass" was a markedly dilated anomalous artery having branching distal runoffs, which originated from the descending aorta. Its

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maximal diameter was larger than the aorta in the same axial plane, measuring about 3.3 cm in diameter (Fig. 1). Volume-rendered image clearly showed the aberrant artery that supplied the left lower lobe (Fig. 2). The dilated bronchus in the left lower basal lung did not communicate with normal left lower lobe bronchus and was impacted with low-density material (Fig. 3). The venous drainage of the sequestrated lung leads to the left atrium by means of left inferior pulmonary veins (Fig. 4). CT angiography showed the atherosclerotic change of the aberrant artery (Fig. 5). Having anomalous arterial supply and discontinuity to the normal tracheobronchial tree while having normal venous drainage of the left lower basal lung by means of left inferior pulmonary vein to the left atrium, the patient was diagnosed as "intralobar pulmonary sequestration".

The patient was treated successfully by left thoracotomy, followed by repair of the descending aortic aneurysm and resection of the left lower lobe. The patient made an uneventful recovery and was discharged from the hospital 10 days after the operation. Pathologic finding was consistent with intraloblar pulmonary sequestration, whereas examination of the dilated anomalous artery revealed that the atherosclerotic change with mural thrombi and the lung parenchyma had chronic inflammatory cell infiltration and bronchiectasis.

3. Discussion

Pulmonary sequestration is the most common congenital abnormality of lung, accounting for 0.2–6.4% of all congenital pulmonary malformation.[1] It is defined as nonfunctioning lung tissue supplied by anomalous systemic artery, which most commonly arises from the thoracic descending aorta at the inferior pulmonary ligament and lacks normal communication to the normal tracheobronchial tree. Pryce[2] first described and classified the pulmonary sequestration into intralobar and extralobar types, the major differences being location and venous drainage of sequestrated lung; typically intralobar sequestration is located within the normal visceral pleura with

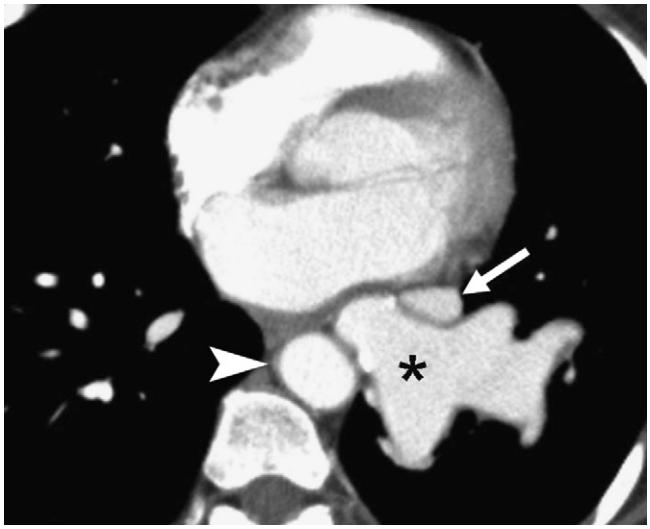

Fig. 1. Axial computed tomography scan showing branching aneurysmal dilatation of the aberrant systemic artery (asterisk) posterior to the left inferior pulmonary vein (arrow). The aorta (arrowhead) was in the medial aspect of the aberrant artery.

Fig. 2. Volume-rendered image showing the aberrant artery that supplied the left lower lobe.

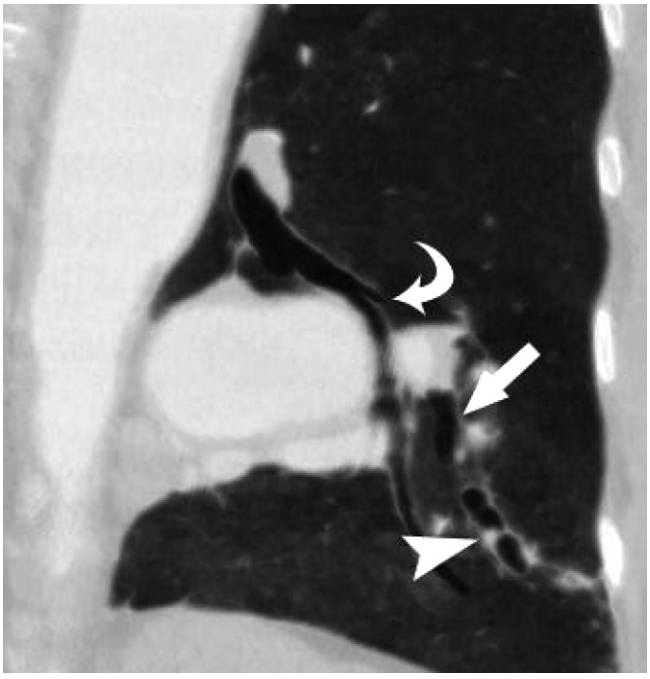

Fig. 3. Minimal intensity projection image showing that the dilated bronchus (white arrow) had no communication to the blind-ended proximal bronchus (curved arrow) and was impacted with mucus (white arrowhead).

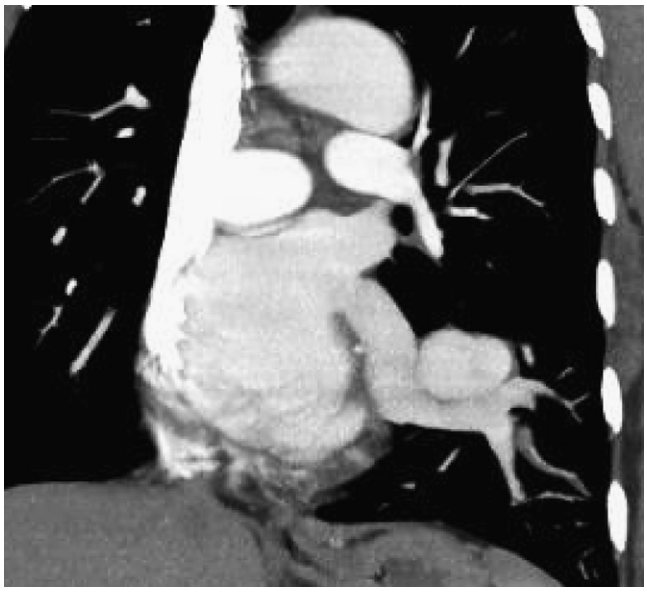

Fig. 4. Oblique coronal thin-slab maximal intensity images depicted that the drainage vein of the left basal lung was to the left atrium by means of the left inferior pulmonary vein.

normal venous drainage to left atrium, whereas extralobar sequestration has its own visceral pleura and anomalous venous drainage into systemic veins routing to the right atrium. Since then, reports and studies about pulmonary sequestration has increased by time. The most frequently involved lung tissue in pulmonary sequestration is the left basal segment with left superior segment spared.

Diagnosing pulmonary sequestration heavily depends on imaging. Chest radiography of intralobar sequestration reveals an irregular-margined consolidation; sometimes tortuous vessel-like structure can be visualized. Contrast-enhanced CT or CT angiography can clearly delineate the origin and course of the anomalous systemic artery. Sequestrated lung in pulmonary intralobar sequestration typically contains multiple cysts filled with fluid or thick mucus, and persistent mucus secretion into ectatic bronchi may also evolve to cystic change.[3]

In our patient, the sequestrated pulmonary parenchyma had relatively normal appearance on imaging, and the patient was initially diagnosed as "anomalous systemic arterial supply to the normal lung", which was first described as Type I pulmonary sequestration by Pryce.[2] It is considered that a persistent primitive aortic branch that originally supplied the developing lung bud leads to systemic arterial supply to lung tissue. It shares many common features with intralobar pulmonary sequestration except having normal bronchi communication.[4] We meticulously examined the images by multiplanar reconstruction and found the bronchial discontinuity for diagnosis of intralobar pulmonary sequestration and the minimal intensity projection reconstruction CT image gave us an overview for the feature in a single image.

Anomalous systemic arteries supplying the low-resistance pulmonary parenchyma often become dilated and tortuous, with mean diameter of aberrant feeding artery of

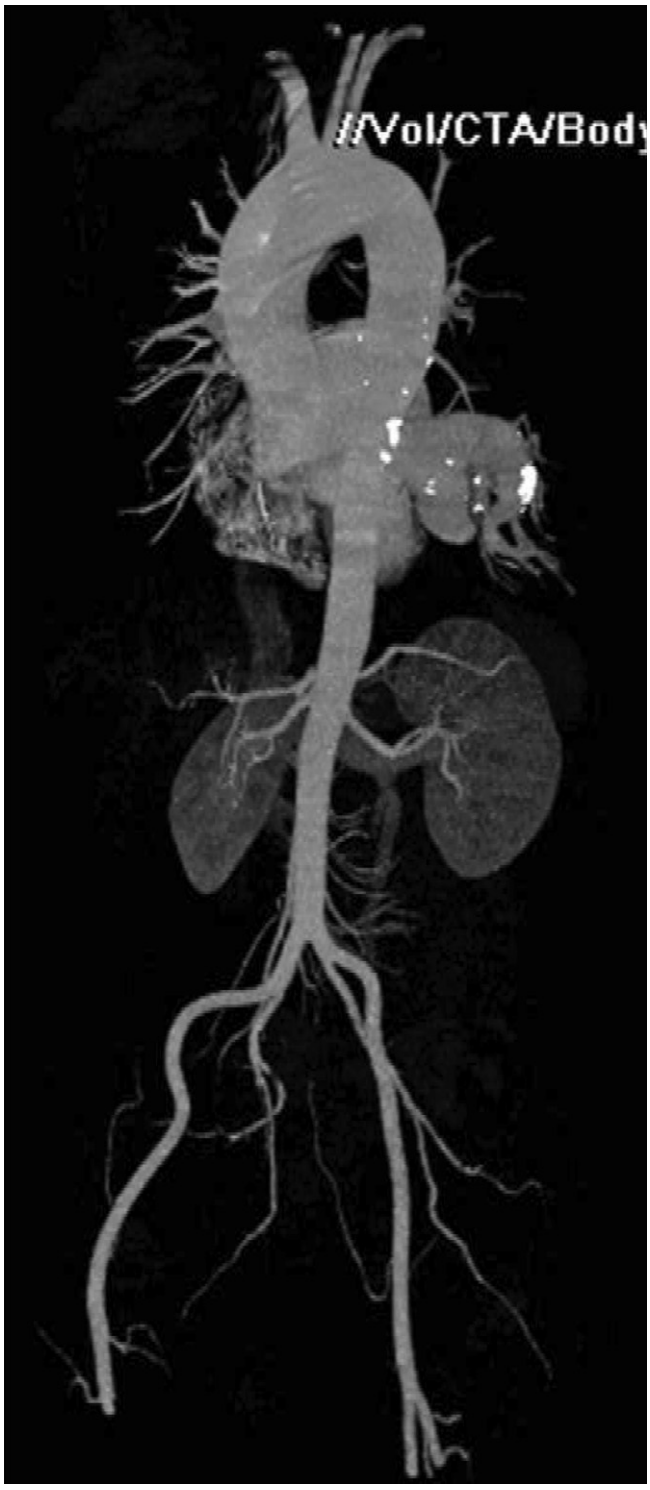

Fig. 5. Thick-slab maximal intensity images of the aorta showing premature atherosclerosis of the anomalous artery and its orifice on the aorta whereas the other part of aorta and its major branches were spared.

6.3–6.6 mm.[3] However, only a few cases have been reported of intralobar sequestrations associated with an aneurysm of an aberrant artery, and the aberrant artery having a diameter greater than that of the descending aorta has not been reported.[5–8] Aberrant artery can show atherosclerotic change

due to long-standing exposure to high systemic arterial pressure and repeated inflammatory insults. In our patient, of the entire course of aorta and its major branches, only the orifice and the largest anomalous arterial branch presented intramural thrombosis and premature atherosclerosis. This increased systemic arterial inflow may also leads to dilatation of the left inferior pulmonary vein. The patient had neither prior history of hypertension nor cardiovascular disease, which may support the pathogenesis of premature atherosclerotic change of aberrant vessels.

Surgical resection with lobectomy of the lung is the treatment of choice for systemic arterial supply to normal basal pulmonary segments in patients with symptoms such as hemoptysis, congestive heart failure, or heart murmur. Other operative procedures include segmentectomy, anastomosis between the anomalous artery and pulmonary artery and ligation of the anomalous artery. Aneurysmal dilatation of the anomalous artery carries potential risk of rupture, therefore our patient received lobectomy even though there was no symptoms. CT angiography with multiplanar reformations have replaced invasive angiography in accurate diagnosis to depict the bronchial discontinuity and provided information for preoperative planning in this patient. Furthermore, in asymptomatic patients, CT angiography serves as regular imaging follow-up modality to monitor the morphological change of the anomalous artery.

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