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Endovascular Therapy for Hemoptysis Caused by a Specific Pulmonary Sequestration Spectrum: Dual Arterial Supply to Normal Lung

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Abstract

The spectrum of pulmonary sequestration represents a wide variety of morphogenetic defects in the embryonic thorax. With a combination of various anatomical anomalies, different treatment options are provided for each form of the disease spectrum. A lobectomy is often applied in symptomatic classic pulmonary sequestration since major complications are derived from the sequestered lung. For rare forms in the disease spectrum without a bronchial tree or venous anomaly, endovascular therapy is the better choice to avoid lung function deterioration. We report a rare case in the disease spectrum with a dual arterial supply (coexistent anomalous systemic artery and pulmonary artery) to a normal (non-sequestered) lung and serial images before and after successful endovascular treatment.

Keywords: Adolescent; Adult; Hemoptysis; Pulmonary sequestration; Computed tomography angiography; Digital subtraction angiography; Endovascular procedures

Introduction

In 1946, Pryce defined pulmonary sequestration as "a disconnected bronchopulmonary mass or cyst with an anomalous systemic artery supply [1]." Because researchers began to describe an increasing number of cases of similar, but not identical, variations, Sade et al. proposed the concept of the sequestration spectrum in 1974 to incorporate more variations that were not included in the initial definition [2]. In 1987, Clements and Warner proposed wheel theory and used a diagram to illustrate their concept of the "pulmonary Mali osculation spectrum", which they described as the abnormal connection between the respiratory primordium and the enteric primordium [3]. Their study expanded the range of the spectrum to include all congenital pulmonary anomalies that exhibited an abnormal connection with one or more of the four major components of lung tissue: tracheobronchial airway, lung parenchyma, arterial supply, and venous drainage.

A detailed image survey for anatomical anomalies in the spectrum of pulmonary sequestration is crucial since there are various treatment options for each combination. An anomalous systemic artery (ASA) supply to a normal (non-sequestrated) lung with normal venous drainage is a rare subtype [4,5]. Furthermore, a dual arterial supply (coexistent ASA and pulmonary artery) is even rarer than a sole supply (ASA only), and there are only 13 proved and published cases [6-14] and our case to our knowledge. Our article presents a case of a dual arterial supply to a normal lung with typical imaging and successful endovascular treatment.

Case Report

A 35-year-old female presented with increasingly frequent blood-tinged sputum in the last 3 months. No fever, sore throat, dyspnea, chest pain, trauma, significant weight loss, or night sweating was noted. She denied contact with any tuberculosis patients. Physical examination at our outpatient department (OPD) revealed normal vital signs; no other apparent abnormality was found. Blood hemoglobin and platelet levels were normal.

A chest X-ray showed a subtle tubular structure behind the left heart and relatively increased vascular opacity in the left lower lung field. A subsequent chest computed tomography angiography (CTA) revealed that the structure was an ASA supplying the basal segments of the left lower lobe (LLL) (Figures 1A-1D). The ASA arose from the left anterolateral aspect of the descending thoracic aorta (DTA) at the T8-T9 level. The CTA also revealed the presence of the left pulmonary artery and its branches in the superior and basal segments. Compared with those on the contralateral side, the branches of the left pulmonary artery were smaller. Increased vascularity in LLL was noted. Computed tomography (CT) and three-dimensional (3D) reconstructive imaging proved that the trachea, bronchi, and small airway were normal and that they showed no narrowing or sequestration (Figure 1E). Radiographic features of ground-glass opacity and consolidation of the pulmonary parenchyma were not observed.

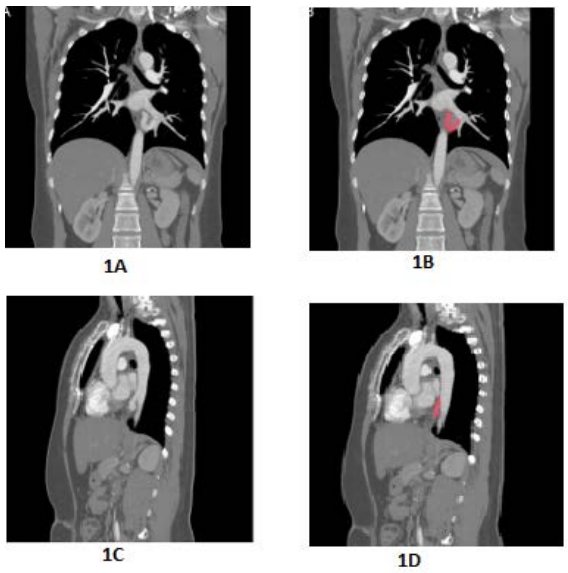

Figure 1(A-D): (A, B) Coronal and (C, D) Sagittal CTA imaging demonstrate an ASA arising from the left anterolateral aspect of DTA.

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Figure 1E: 3D reconstructive airway imaging also confirms a normal airway distribution.

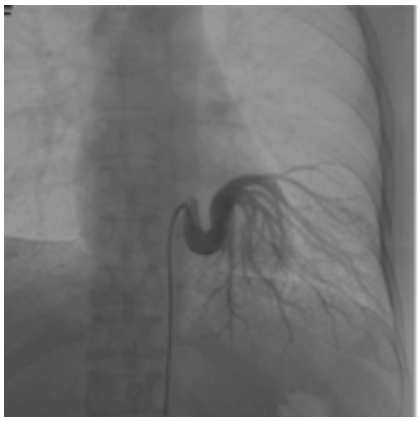


Figure 1F: Selective DSA of the ASA depicts that the artery supplied the basal segments of the LLL.

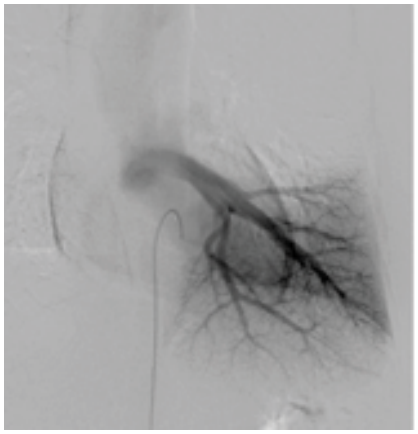


Figure 1G: Subsequent venous phase imaging shows normal blood drainage into the left pulmonary vein.

Selective digital subtraction angiography (DSA) of the ASA confirmed that the artery supplied the basal segments of the LLL (Figure 1F). Venous phase imaging proved normal blood drainage into the left pulmonary vein (Figure 1G). Moreover, a perfusion isotope scan showed no significant defect (Figure 1H). This result and the CTA outcome both revealed normal distribution of the left pulmonary

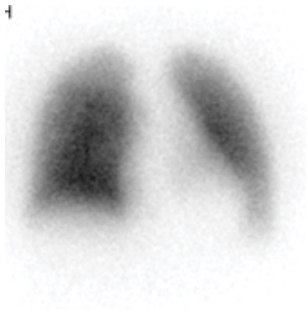
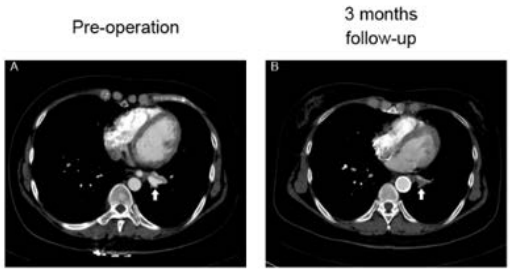
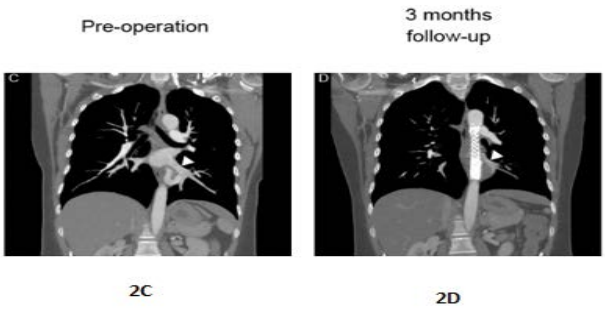


Figure 1H: Perfusion isotope scan shows no significant defect.



Figures 2A and 2B: Preoperative and follow-up CTA that are performed at 3 months, focusing on the interval changes of (A, B) ASA (arrow).

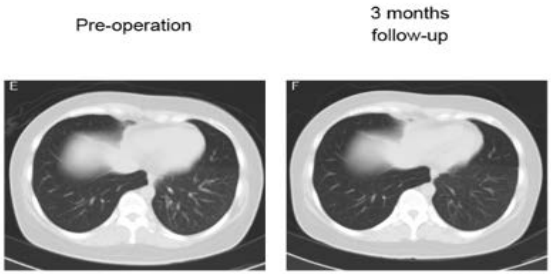


Figures 2C and 2D: Left inferior pulmonary vein (arrow head).

artery. Echocardiography showed no abnormality, except for trivial mitral valve regurgitation. These findings were consistent with a dual arterial supply to a normal lung with normal venous drainage.

Subsequently, she received thoracic endovascular aortic repair (TEVAR) with GORE TAG stent-graft coverage of the anomalous systemic artery. She was discharged uneventfully because of her stable condition, and there was no immediate complication.

She visited the OPD for regular follow-up, and her hemoptysis was completely resolved. The follow-up CTAs were performed at 3 months after the procedure and showed regression of the ASA and no intravascular enhancement (Figures 2A and 2B). The interval diameter of the left lower pulmonary vein was 10.5 mm and 7.6 mm, respectively (Figures 2C and 2D). Preoperative hypervascularity in the LLL disappeared, and the follow-up images did not indicate any



Figures 2E and 2F: (E, F) Vascularity, and pulmonary parenchyma. Regression of the ASA and no intravascular enhancement are found at 3 months after operation. The preoperative and postoperative diameter values of the left lower pulmonary vein are 10.5 and 7.6, respectively. Preoperative hyper vascularity in the LLL disappears, and the postoperative images do not indicate any other anomaly of the pulmonary parenchyma.

other anomaly of the pulmonary parenchyma (Figures 2E and 2F). The second perfusion isotope scan performed 8 months after surgery did not reveal any difference from the preoperative data.

Discussion

We have reviewed all the case reports published in English in the last 20 years by using search engines (e.g., PubMed and Google). The studies that we found employ confusing or unclear names to refer to rare forms of pulmonary sequestration spectrum, and studies that do not clearly identify each anatomical anomaly are excluded. The cases with pulmonary arteriovenous malformation or fistula are also excluded. As of May 2018, 13 case reports have described a dual arterial supply to normal lungs with normal venous drainage [6-14], including the present case.

A review of the 13 cases revealed no obvious gender predilection (male: 7, female: 6, n=13). The age at diagnosis ranged from 17 years to 58 years (average: 35 years, median: 35 years). Specifically, in contrast to cases of classic pulmonary sequestration, which causes airway obstruction, these cases occasionally had recurrent infection as the clinical manifestation but mostly displayed the symptom of hemoptysis (84.6%, 11 of 13). This symptom of hemorrhage may be attributed to the high blood pressure caused by a systemic or dual supply; in addition, these cases did not exhibit airway obstruction and therefore mainly presented hemoptysis. Among the cases, ASA orifices mainly appeared in the thoracic descending aorta (69.2%, 9 of 13), followed by the celiac trunk or the very proximal abdominal aorta (30.8%, 4 of 13). The ASA and involved pulmonary lobe are single in all cases. Almost all of the cases involved basal segments of the lung (92.3%, 12 of 13). Among these cases, the number of cases with an anomalous supply to the LLL (n=9) was 3 times the number of cases with an anomalous supply to the right lower lobe (n=3). In four cases (30.8%), focal ground-glass opacity was detected; the radiographic features were related to acute hemorrhage [8-14]. Two case reports were published in 2006 and 2015; observation about surgery was noted, specifically that no gross lung parenchymal ischemia was observed after transient ASA ligation for a while [9,14]. Four cases received endovascular therapy with coils or glue [6-12]; all the treated cases become symptomless.

Coils, vascular plugs, tissue glue, stent, a combination of multiple materials, or added temporary embolic particles for demising the blood flow of ASA were adopted in some case reports and resulted in effective treatment for hemoptysis. There is no obvious suggestion for material choice because of the limited cases currently.

During the 5-year follow-up period, the CTA images demonstrate complete regression of the ASA. Notably, the diameter of the left inferior pulmonary vein significantly decreased from 10.5 mm to 7.6 mm because the ASA perfusion disappeared, i.e., cardiac preload decreased. The lung parenchyma remained normal on CT imaging, and it is also proved by the constant perfusion isotope scan.

Conclusion

A detailed image survey for anatomical anomalies in the pulmonary sequestration spectrum is crucial since there are various treatment options for each combination. Endovascular treatment ensures effectiveness for the rare forms in the disease spectrum without a bronchial tree anomaly. Our case provides an experience that using stent-graft can also reach the therapeutic effect. The benefit of treatment in dual arterial supply cases are reducing hemoptysis and preserving normal lung function.

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