Case Report

Pneumonectomy in an elderly patient with congenital unilateral lung hypoplasia and recurrent *Aspergillus*: a case report

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Background: Congenital lung disorders encompass a spectrum of several conditions, one of which is lung hypoplasia. While hypoplasia is typically identified and intervened upon in the neonatal period, some patients can remain asymptomatic or even be diagnosed as adults. Given the rarity of the condition persisting in adults though, it is not clear what medical or surgical interventions may be helpful if respiratory status declines.

Case Description: In this report, we describe an elderly man with a history of right lung hypoplasia, pulmonary artery agenesis, and bronchial atresia who developed progressive dyspnea and worsening cough. His condition was complicated by multiple *Aspergillus* infections for which he received prolonged courses of anti-fungal therapy. He was also treated for bacterial pneumonia many times over a 10-year period. However, as his symptoms remained refractory to medical management, he underwent pneumonectomy, which revealed diffuse cystic changes in the right lung. He is currently doing well post-operatively with resolution of his dyspnea.

Conclusions: Although hypoplastic lung disorders have been described in asymptomatic adults, this report highlights the successful utilization of pneumonectomy in an individual with refractory symptoms much later in adulthood. This case additionally describes possible complications of cystic lung disease in this patient population, serving as further rationale for aggressive intervention.

Keywords: Lung hypoplasia; pneumonectomy; *Aspergillus*; congenital; case report

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Introduction

Lung hypoplasia is a congenital disorder that is usually detected in childhood. Although the true incidence is not known, it is estimated to have a prevalence of 9% to 28% in cases of premature rupture of membranes at 15–18 weeks gestation (1). The majority of patients with lung hypoplasia have congenital abnormalities of other organs, which often leads to further complications. However, patients with various degrees of pulmonary underdevelopment occasionally remain asymptomatic and can be followed closely for symptom progression over time (2-5).

The development of diffuse cystic disease in the hypoplastic lung has not been previously described. In this report, we present an elderly patient with unilateral right lung hypoplasia which was complicated by recurrent *Aspergillus* infections. Given gradual progression of symptoms later in adulthood, he underwent pneumonectomy, which yielded significant improvement in his respiratory difficulties. This case serves to emphasize that age and other comorbidities should not be absolute

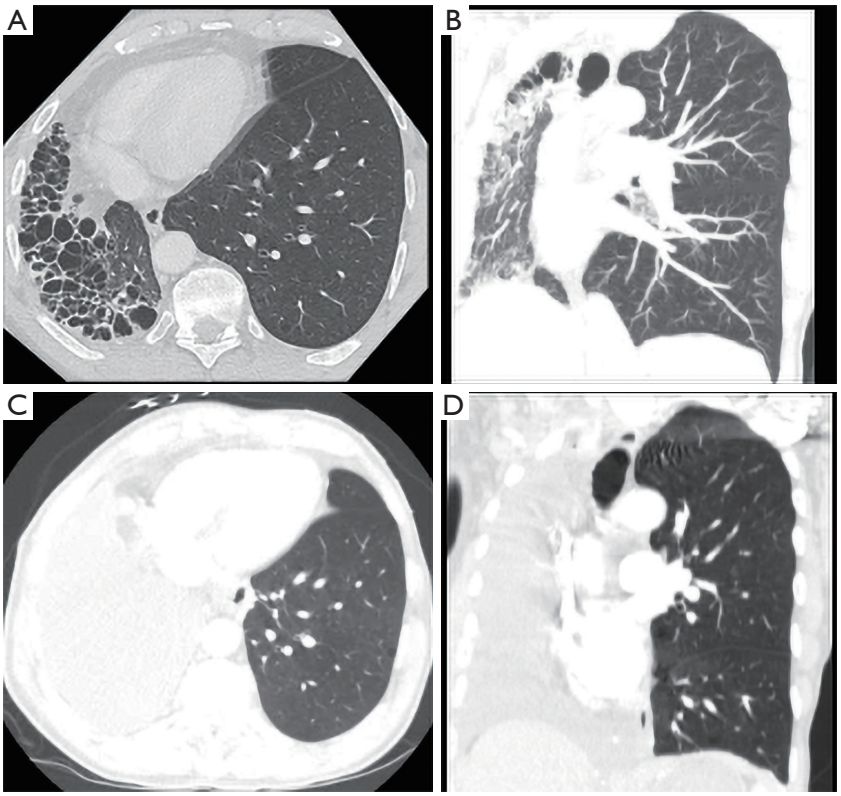

Figure 1 Patient's CT scans pre- and post-operatively. (A,B) Axial and coronal pre-operative CT images, respectively, showing hypoplasia and diffuse cystic changes in the right lung. (C,D) Post-operative CT images showing successful right sided pneumonectomy. CT, computed tomography.

contraindications for surgical intervention in these patients, particularly when optimal medication therapy is not sufficient for symptom control. We present the following case in accordance with the CARE reporting checklist (available at https://acr.amegroups.com/article/view/10.21037/acr-22-38/rc).

Case presentation

A 72-year-old man with a history of type 2 diabetes mellitus and chronic immune thrombocytopenic purpura presented with acute on chronic cough. He reported first receiving a diagnosis of right lung underdevelopment at the age of 6. Recent computed tomography (CT) scans confirmed congenital hypoplasia of his right lung, absence of his pulmonary artery with numerous bronchial artery collaterals, diffuse bronchial atresia, and peripherally distributed cystic disease. In addition to his diabetes medications, his current medication regimen consisted of tiotropium/olodaterol (Stiolto Respimat) 2.5-2.5 µg/actuation 2 inhalations per day, theophylline 200 mg daily, and as needed ipratropium/albuterol (Combivent). Several sputum cultures, the first of which was over 10 years from the current presentation, showed *Aspergillus fumigatas*, for which he was treated with extended courses of voriconazole, ranging from weeks to months. He experienced mild hemoptysis prior to starting his first course of voriconazole, though did not have any recurrence of this symptom later in life. However, voriconazole was often restarted when he experienced worsening cough or dyspnea and sputum cultures showed *Aspergillus*. In total, he received voriconazole for 46 months over this 10-year period. Recent serological studies for *Aspergillus* antibodies were negative. Radiographically, his repeat fungal infections correlated with multiple scattered mycetomas on serial CT scans (*Figure 1A,1B*). Additionally, he was empirically treated for bacterial pneumonia or acute bronchitis at least 16 times over a 15-year period in the setting of worsening cough and intermittent pleurisy.

Due to his favorable functional status and frequency

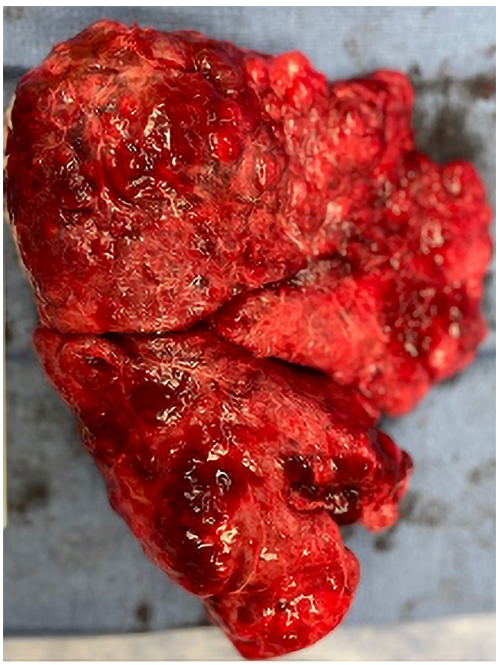

Figure 2 Intra-operative findings: diffuse cystic disease in right lung.

of *Aspergillus* infections, along with inadequate symptom control with his inhaler regimen to date, he was felt to be a suitable candidate for right-sided pneumonectomy. His pre-operative pulmonary function tests (PFTs) were compatible with long standing obstructive lung disease, with predicted forced expiratory volume in one second (FEV1) and diffusing lung capacity for carbon monoxide (DLCO) of 61% and 98% respectively, and preserved bronchodilator response. He underwent successful right pneumonectomy (*Figure 1C,1D*), with intraoperative findings revealing large cystic disease with purulence (*Figure 2*). Final pathology revealed prominent bronchiectasis with associated lymphocytic infiltrates and clusters of fungal microorganisms with hyphae-forms in the dilated airways of all three lobes, especially in the lower lobe. He had an uneventful postoperative course and is currently doing well, approaching 1 year from his surgery. He is using albuterol as needed for symptom control and is gradually increasing his exercise tolerance. Serial follow-up imaging has shown a well aerated left lung.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Unilateral pulmonary hypoplasia has been reported to have a mortality rate of up to 50% in the neonatal period (6). Most often, the underdevelopment of the lung is secondary to other congenital abnormalities, including but not limited to congenital diaphragmatic hernia, congenital heart diseases, and genitourinary malformations (7). The primary form of hypoplasia is rarer and thought to be idiopathic; as in this patient, it is not associated with other congenital disorders. While the majority of individuals with unilateral hypoplasia are diagnosed at a young age, a minority have none to minimal symptoms and are diagnosed in adulthood, often incidentally on chest imaging. In patients with agenesis, which refers to complete absence of pulmonary parenchyma, bronchi, and vessels, there is some evidence that right-sided agenesis is associated with decreased survival than left-sided agenesis possibly due to higher incidence of heart failure in the neonatal period, as well as increased incidence of multiorgan congenital underdevelopment (8). However, this mortality difference has not been reported in those with unilateral hypoplasia.

This patient's obstructive pattern on PFTs with a response to bronchodilators was suggestive of an asthma physiology, for which he was trialed on several inhalers and oral medications with moderate benefit. However, the gradual worsening of his respiratory status over a long period of time may have been due to his recurrent pulmonary infections. Specifically, the diffuse cystic disease in his hypoplastic lung likely served as a nidus for *Aspergillus* infections. An aspergilloma has been previously reported in a patient with congenital cystic adenomatoid malformation (CCAM), a rare congenital cystic lung disease (9). However, cystic disease in CCAM is often localized to one lobe and is not associated with pulmonary artery agenesis and bronchial atresia, suggesting that this was not the underlying etiology for this patient.

The role of pneumonectomy in this patient population is unclear. One retrospective study of various surgical interventions in adult patients with congenital lung diseases showed similar rates of complications and length of hospitalization with video-assisted thoracoscopic surgery (VATS) interventions versus thoracotomy approaches (10). The extensive adhesions in this patient would have made VATS challenging in this case, however the aforementioned

study indicates that there may be a role for minimally invasive interventions in this population. Moreover, the patient in this case is one of the oldest reported to have undergone definitive surgical intervention for his hypoplastic lung. We anticipate that evaluation for surgical intervention may have been delayed initially due to this patient's age. While older age is generally thought to be a risk factor for post-operative morbidity and mortality, studies have suggested that age itself should not be a contraindication for pneumonectomy and instead should be one component of a comprehensive risk-assessment approach (11,12). Additionally, there may be higher rates of perioperative complications for right-sided versus left-sided pneumonectomies, including but not limited to bronchopleural fistulas and pulmonary edema, though further investigation is needed to clarify true differences in outcomes between the two procedures (13).

Conclusions

This case report serves to highlight the potential beneficial role of pneumonectomy for progressive obstructive lung physiology in a rare patient presentation. Regular evaluation of this patient's dyspnea and other respiratory symptoms was key to his eventual successful outcome. Ultimately, the role of surgical interventions remains to be further defined for this patient population, and reports such as this will add to existing, limited data on adults with congenital lung conditions. It is crucial to determine both optimal medical management as well as timing for surgical intervention in adult patients with congenital lung disease.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://acr.amegroups.com/article/view/10.21037/acr-22-38/rc

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Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at https://acr.amegroups.com/article/view/10.21037/acr-22-38/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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