performed. The manubrium was particularly thick and posteriorly deviated; therefore, the posterior half was shaved off with a saw before the distal sternum was closed with wire. The proximal sternum was widened with Actifuse ABX bone graft substitute (ApaTech, Elstree, UK).

The patient was extubated immediately after the operation. After resolution of mild pericardial and pleural effusions, respiratory function improved significantly. Exertional dyspnea resolved, and FEV_1 increased from 1.08 L (37% predicted) preoperatively to 1.35 L (46%). On examination, the chest was clear, ventilation was bilateral, and there were no extraneous tracheal sounds. The patient felt much better.

Comment

Despite the high prevalence of respiratory dysfunction associated with HS, with nearly half of patients dying of related problems [2], the heterogeneity of airway involvement complicates management. The most commonly understood mechanisms of airway obstruction in HS are GAG deposition within the tracheal wall [4, 5] and external compression because of mediastinal soft tissue GAG accumulation [6]. Neither pathology was noted in our patient. Furthermore, the nature of tracheal problems described here contrasted with the tracheal softening proposed to cause airway collapse in HS [4, 5], as our patient's trachea was rigid, thickened, and abnormally shaped. Therefore, it was particularly challenging to determine the most appropriate treatment strategy or predict its efficacy.

Current treatments for respiratory compromise in HS, as in other circumstances of tracheomalacia, include continuous positive airway pressure, tracheostomy, laser excision of obstructive lesions, and stenting [8]. The methods were considered, but rejected, and we decided to explore the mediastinum, remodel the manubrium, and perform aortopexy with possible external splinting to the trachea.

The tracheal rigidity was not only unanticipated, but also limited the effect of aortopexy alone. Only after tracheopexy was an improvement in tracheal diameter observed. Nonetheless, the use of tracheopexy for tracheobronchomalacia has been limited, and it has not been described previously in the setting of HS. Although the preliminary results are encouraging, in hindsight and given the tracheal inflexibility, we have considered that greater enlargement might have occurred if partial thickness incisions were made in the affected tracheal cartilages (Fig 2B).

With the emergence of enzyme replacement therapy and its effect on increasing life expectancy in HS patients, there will undoubtedly be a greater need to address the respiratory manifestations of the disease. The technique we describe here may help others to manage these complex problems.

This work was supported in part by the American Association for Thoracic Surgery's Summer Intern Scholarship.

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FEATURE ARTICLES

Pulmonary Artery Agenesis Associated With Emphysema and Multiple Invasive Non-Small Cell Lung Cancers

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Pulmonary artery (PA) agenesis in the absence of associated cardiac abnormalities is a rare congenital abnormality. It may remain undiagnosed until adulthood when patients present with respiratory symptoms such as hemoptysis, dyspnea, repeated respiratory infections, or pulmonary hypertension. Herein we present a case of a 50-year-old woman who was found to have multiple, morphologically distinct non-small cell lung cancers in association with agenesis of the PA. This instance represents the fourth reported case of such association in the English literature.

(Ann Thorac Surg 2015;99:2192–5)

Pulmonary artery agenesis is a rare congenital abnormality that may present in varied clinical scenarios, depending on the associated abnormalities. It is secondary

Accepted for publication Aug 29, 2014.

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0003-4975/$36.00
http://dx.doi.org/10.1016/j.athoracsur.2014.08.062

to a failure in the connection of the sixth aortic arch with the pulmonary trunk [1]. Since it was described in 1868 less than 200 cases have been reported. The prevalence of pulmonary artery agenesis is estimated to be around 1 in 200,000 individuals and it is usually accompanied by congenital cardiac malformations such as ventricular septal defect, tetralogy of Fallot, truncus arteriosus, and right-sided aortic arch [1]. In the absence of cardiac abnormalities it may remain undiagnosed until adulthood when patients begin having respiratory symptoms such as hemoptysis, dyspnea, repeated respiratory infections, or pulmonary hypertension [1, 2].

A 50-year-old woman presented with episodes of recurrent hemoptysis. She noted previous episodes of hemoptysis (in her 20s) which spontaneously resolved, and no further workup was performed at that time. The current episodes are characterized by a few ounces of bright red blood, accompanied by frothy sputum. The patient reports a history of shortness of breath throughout her life and notes that she has difficulty keeping up with her children's activities. She is a former smoker (25 pack-years), having quit 12 years ago. A chest X-ray revealed absence of the right pulmonary artery shadow, right cardiac and mediastinal displacement, and smaller right hemithorax, and grossly diminished right pulmonary vascular markings.

Computed tomographic (CT) scan of her chest revealed absence of the right pulmonary artery and emphysema changes in the right lung (Figs 1, 2). There was hypertrophy of the right internal mammary artery with multiple systemic collaterals identified along the pleural surface, particularly along the right hemidiaphragmatic pleural surface. The bronchial arteries were dilated, reflecting significant bronchial collateral circulation. Additionally, 4 ground glass opacities, ranging from 1.0 to 2.5 cm, were identified within the right lung; no similar ground glass opacities were identified within the left lung. A bronchoscopy revealed multiple dilated blood vessels on the mucosal surface of the right bronchial tree, while the left side was unremarkable.

Her recurrent hemoptysis was attributed to the hypervascularity of her airway from bronchial collaterals as a result of her right-sided pulmonary artery agenesis. It was determined that a pneumonectomy would be the best option for management of her recurrent hemoptysis. Intraoperative findings included a grossly abnormal right lung with prominent visceral pleural vasculature, excessive collaterals arising from the subclavian artery, within the mediastinum, diaphragm, chest wall, and lung apex. Additionally, the lung had bullous changes with parenchymal destruction. Upon careful dissection of the hilum, fibrous tissue was present in place of normal pulmonary arteries. A right pneumonectomy and a mediastinal lymph node dissection were performed.

Examination of the pneumonectomy specimen in pathology revealed, in addition to the markedly abnormal vasculature, multiple (6) distinct lung nodules involving all 3 lung lobes. Histologically, the medium- and small-caliber pulmonary arteries were markedly atretic and the bronchial arteries exhibited engorgement and significant remodeling. The 2 largest nodules (2.2 cm and 1.5 cm), located in the upper lobe, were ultimately classified as adenocarcinomas with predominately lepidic growth patterns. Two nodules located in the middle lobe (1.2 cm and 0.9 cm) proved to be an adenocarcinoma with papillary features and an adenocarcinoma with acinar growth pattern, respectively. In situ adenocarcinomas (0.4 cm and 0.3 cm) were also identified in the upper and lower lobes, respectively. No visceral pleural involvement was identified with any of these lesions and the parenchymal and hilar lymph nodes were uninvolved. Given the morphologic disparity between these lesions, all tumors were considered independent primary lung cancers; the final stage was pT1b, N0, stage Ia. Mutational analysis of the *EGFR* gene [epidermal growth factor receptor] was performed on the 4 invasive tumors, 3 of which proved to be wild-type (mutation-negative). The middle lobe tumor, however, harbored an exon 21 Leu861Gln mutation.

The patient's postoperative course was marked by transient atrial fibrillation with a rapid ventricular response, which was treated medically. She was discharged home on postoperative day number 5. Adjuvant therapy was not recommended. The follow-up plan was to maintain surveillance with annual low-dose screening computed tomographic scans. At a recent 1-year follow-up appointment the patient was asymptomatic and there was no evidence of recurrent or metastatic disease.

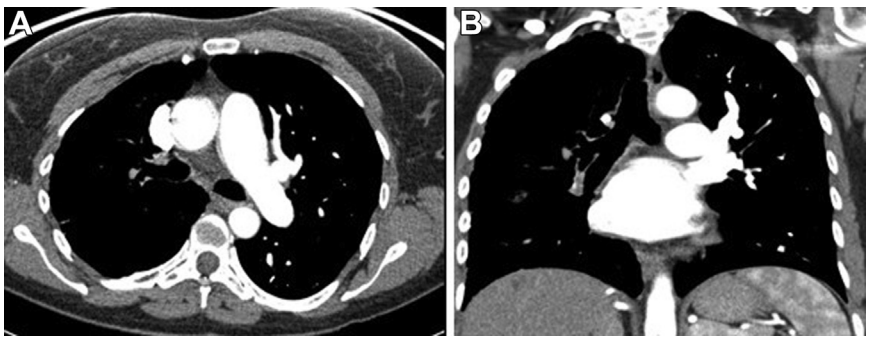

Fig 1. Chest computed tomographic scan; (A) and (B) images reveal the absence of the right pulmonary artery and the absence of the arterial vasculature in the right lung.