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Commentary: Isolated unilateral absence of pulmonary artery, a rare disease with diverse presentations

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CENTRAL MESSAGE

The unilateral absence of pulmonary artery present in various spectrums of disease. To provide the best treatment, a comprehensive understanding of the natural history of the disease is mandatory.

Isolated unilateral absence of the pulmonary artery (UAPA) is a rare disease, and the diagnosis is often delayed as the symptoms are usually nonspecific.[1] Depending on the severity of the disease and the patient's age at the time of diagnosis, the main issues related to the disease vary and, therefore, cardiothoracic surgeons encounter diverse clinical situations.

The 3 major issues in patients with UAPA are pulmonary hypertension, recurrent pulmonary infections, and hemoptysis.[2] In patients with severe disease and diagnosed during the neonatal period, pulmonary arterial hypertension is the main issue.[3] Early revascularization of the absent pulmonary artery may provide near-normal growth of the affected side of the lung.[4]

If the disease is not severe, the diagnosis of the UAPA is frequently delayed until adulthood. Various treatments are available for the treatment of late-diagnosed UAPA, mainly depending on the symptom.[2] For patients with pulmonary hypertension, long-term vasodilator therapy may be helpful. Recurrent pulmonary infections or hemoptysis will require appropriate treatment such as systemic antibiotics or selective bronchial artery embolization.[5] Those symptoms may be self-limiting, but if they become severe, surgical intervention, usually pneumonectomy, may be mandatory.[6]

In the paper entitled "Thoracoscopic Lobectomy for Lung Cancer With Unilateral Absence of Pulmonary Artery," Matsumoto and colleagues[7] presented a rare case of UAPA diagnosed at the age of 80 years in a patient with lung cancer in the affected side of the lower lobe. It is a compelling case, as it represents an end of asymptomatic UAPA, where no treatment would have been required if the lung cancer did not develop.

Given the age of 80 years and the expectation of abundant systemic collateral arteries, surgical treatment might be challenging. A careful multidisciplinary discussion seemed to be conducted. Once surgical treatment is decided, the optimal extent of surgery should be decided by the surgeon. If the patient's age were 50 years and they had several hemoptysis episodes, what would have been the best choice? I would have done pneumonectomy rather than a lobectomy because the patient may experience serious hemoptysis during the rest of their life. However, for the 80-year-old patient, lobectomy may be appropriate. If the collateral arteries made anatomic resection dangerous, wide wedge resection might have been a choice. If the fissure was challenging to separate, a fissureless technique might have been appropriate. The authors avoided dissecting the remaining lungs from the chest wall, through which abundant collateral arteries supplied lung parenchyma. Given the age of 80 years, trading the benefit of paratracheal lymph node dissection

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with the risk of postoperative bleeding seemed to be a wise choice.

This case report reminds us of the fact that the UAPA presents in various spectrums of disease. As cardiothoracic surgeons, we will encounter it either as a congenital cardiac surgeon or a thoracic surgeon. To provide the best treatment, a comprehensive understanding of the natural history of the disease and the patient's clinical condition is mandatory.

References

1. Morales P, Miravet L, Marco V. Agenesis of the right pulmonary artery in a young asymptomatic girl. *Eur Respir J*. 1991;4:1301-2.
2. Wang P, Yuan L, Shi J, Xu Z. Isolated unilateral absence of pulmonary artery in adulthood: a clinical analysis of 65 cases from a case series and systematic review. *J Thorac Dis*. 2017;9:4988-96.
3. Ten Harkel AD, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery: a case report and review of the literature. *Chest*. 2002;122:1471-7.
4. Kim GB, Ban JE, Bae EJ, Noh CI, Kim WH, Lee JR, et al. Rehabilitation of pulmonary artery in congenital unilateral absence of intrapericardial pulmonary artery. *J Thorac Cardiovasc Surg*. 2011;141:171-8.
5. Dhanoa D, Jaskolka JD, Darling G, Hanna W. Particle embolization as primary endovascular management of a patient with massive hemoptysis and isolated unilateral absence of the left pulmonary artery. *J Vasc Interv Radiol*. 2011;22:256-8.
6. Ohtsuka T, Nomori H, Watanabe K, Kaji M, Ebihara A, Naruke T, et al. Isolated unilateral absence of a pulmonary artery treated by pneumonectomy in an adult: report of a case. *Surg Today*. 2006;36:525-7.
7. Matsumoto D, Toba H, Kondo K, Tangoku A. Thoracoscopic lobectomy for lung cancer with unilateral absence of pulmonary artery. *J Thorac Cardiovasc Surg Tech*. 2020;3:343-5.