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Isolated unilateral pulmonary artery agenesis presenting with massive hemoptysis

Min Kyun Kang, Do Kyun Kang & Youn-Ho Hwang

Department of Thoracic and Cardiovascular Surgery, Haeundae Paik Hospital, Inje University College of Medicine, Busan, South Korea.

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Correspondence
Do Kyun Kang, Department of Thoracic and Cardiovascular Surgery, Haeundae Paik Hospital, College of Medicine, Inje University, Haeundae-ro 875, Haeundae-gu, 48108, Busan, South Korea.
E-mail: med7695@naver.com

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Abstract

This case was about a 21-year-old man with recurrent and persistent hemoptysis. Chest computed tomography scan showed a right-sided pulmonary artery agenesis and associated hypertrophic change of right side bronchial arteries. Thoracic aortogram revealed abnormal collateral arteries supplying the right lung. Minor hemoptysis continued even after endovascular embolization of collateral vessels was performed. Right pneumonectomy was performed and the patient was treated successfully. Physicians should be aware that unilateral pulmonary artery agenesis could be a source of hemoptysis and pneumonectomy may be the best treatment.

Introduction

Unilateral pulmonary artery agenesis (UPAA) is a rare congenital abnormality associated with other cardiac abnormalities such as ventricular septal defect, tetralogy of Fallot, truncus arteriosus communis, and right-sided aortic arch [1]. Isolated UPAA, which has no cardiac abnormalities, constitute about 30% of UPAA patients and might be undiagnosed until adulthood because it usually presents with mild clinical symptoms in infancy and childhood [2]. The symptoms of isolated UPAA patients include hemoptysis, repeated respiratory tract infection, dyspnea, and pulmonary hypertension. Less than 10% of UPAA patients presented with hemoptysis [3]. This case presents a UPAA patient with massive hemoptysis, which was treated with pneumonectomy.

Case Report

A 21-year-old man was admitted with a history of recurrent hemoptysis. The patient had had hemoptysis for the last 5 years and been treated with hemostatic agents. But in these episodes, massive hemoptysis presented with cough (about 300 cc blood and clots). The day before hospitalization, the patient suffered about 50 episodes of hemoptysis. His hemoglobin level at the time of admission was 16.2 g / dl, and 2 days later, it was measured at 13.9 g / dl. The patient had a history of craniectomy due to traumatic subdural hemorrhage. He was a smoker with a 2 pack-year smoking history. He had continuously experienced minor hemoptysis in the hospital.

A computed tomography (CT) demonstrated right-sided pulmonary artery agenesis and associated hypertrophic change of right side bronchial arteries (Fig. 1). No cardiac anomalies were detected using echocardiography. Bronchoscopy identified large amount of fresh blood in the trachea and localized in the right main bronchus.

Thoracic aortogram revealed abnormal collateral arteries supplying the right lung (Fig. 2). They originated from hypertrophied right thyrocervical trunk and right internal thoracic artery, which were embolized with glue. Angiogram performed after embolization revealed complete obstruction. However, minor hemoptysis continued for 24 h and surgical treatment was recommended after multidisciplinary discussion.

Pneumonectomy was performed with a posterolateral thoracotomy. Severe adhesions were observed around hilar

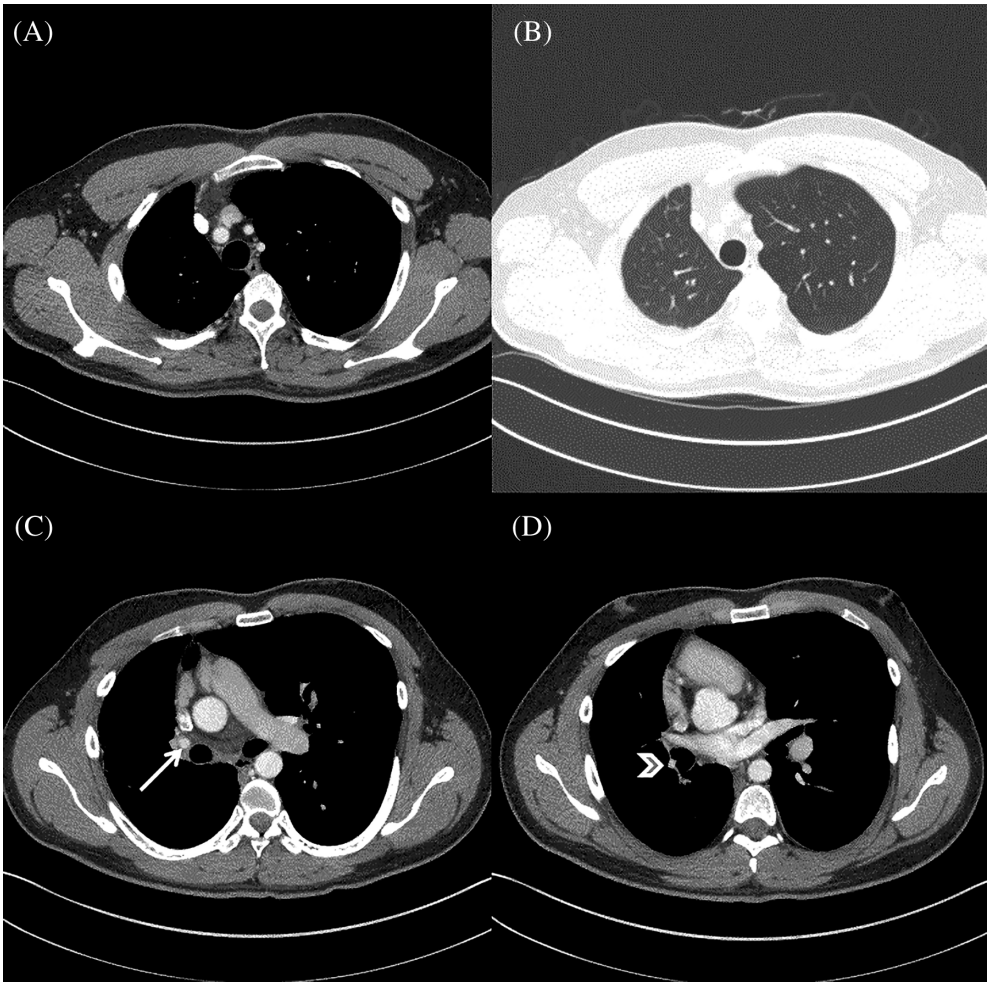

Figure 1. Computed tomography demonstrating small volume of right lung with mediastinal deviation to right side (A, B) and right pulmonary artery agenesis and bronchial artery aneurysm (arrow), hypertrophic change of bronchial artery (arrowhead) (C, D).

structures. Right pulmonary artery was absent. Rt. bronchial artery presented an aneurysmal change. Right superior and inferior pulmonary vein, and the right main bronchus were isolated, stapled, and divided. The histological findings revealed that hemorrhage of tortuous peribronchial blood vessels and no right pulmonary artery. The patient had no more hemoptysis and recovered well. He was discharged on postoperative day 16. The prolonged postoperative hospitalization was due to the thoracotomy wound dehiscence. In a recent follow-up 2 months after discharge, he had no symptoms such as hemoptysis or cough.

Discussion

UPAA was reported for the first time in 1868 [4] and less than 200 cases have been reported in the English literatures till now. UPAA is caused by the failure of the sixth aortic arch to connect with the pulmonary trunk. Right pulmonary artery agenesis is usually less frequently associated with abnormal cardiac anatomy than left pulmonary artery agenesis. The pulmonary arterial supply originates from collateral vessels, which is hypertrophied and easy to rupture to cause hemoptysis. The main diagnostic tool is the contrast-enhanced CT or magnetic resonance imaging. Treatment of UPAA depends on the age and symptoms. In pediatric patients with pulmonary hypertension, revascularization of the affected side could be the best treatment option. Endovascular embolization may be the possible option for hemoptysis cases [5], but the development of another collateral vessel could be the cause of recurrence. Pneumonectomy could be the treatment option in adults who are refractory to the other treatments, such as embolization, medication, especially in the isolated UPAA cases.

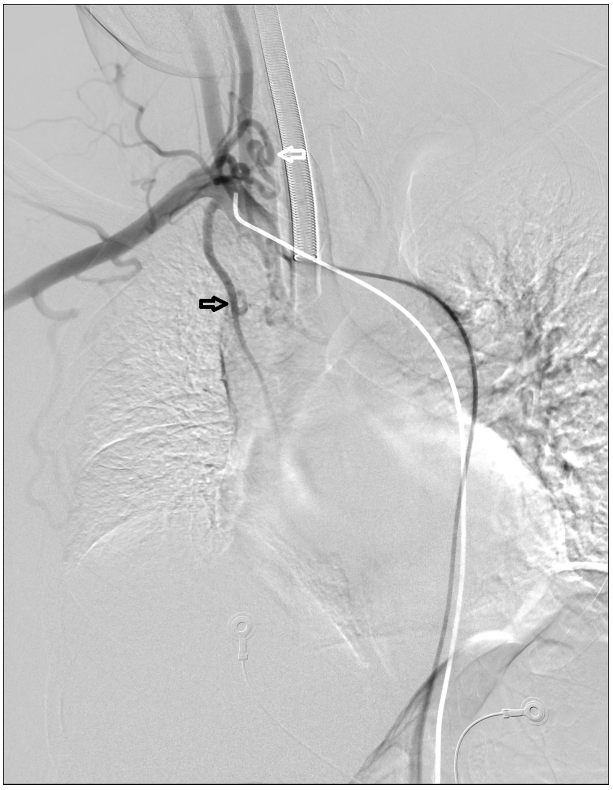

Figure 2. Conventional thoracic angiogram demonstrating abnormal feeding arteries arising from right thyrocervical trunk artery (white arrow) and right internal thoracic artery (black arrow).

The patient in this study case presented with a recurrent hemoptysis. Collateral vessels embolization was performed, but it was ineffective and hemoptysis continued. Pneumonectomy was the definite treatment method for the 21 year old patient.

Physicians should be aware that UPAA could be a source of hemoptysis presenting in adulthood and pneumonectomy may be the best treatment option for relieving hemoptysis.

Disclosure statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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