Massive Hemoptysis due to Left Inferior Phrenic Artery-to-Left Pulmonary Artery Fistula in the Lingular Lobe of the Lung: A Case Report and Literature Review

Ching-Chieh Lin, Tsai-Wang Huang*, Kai-Hsiung Ko**, Wann-Cherng Perng***, Chih-Feng Gilan***, Ying-Chieh Chen***

Massive hemoptysis is a pulmonary emergency requiring immediate management, such as bronchial angiographic embolization or surgical intervention. It occurs in various pulmonary diseases and typically derives from the bronchial arteries. We herein report a very rare case of a patient bleeding from a left inferior phrenic artery-to-pulmonary artery fistula, accompanied by focal bronchiectasis in the left lingular lobe of the lung. In this case, pulmonary angiography was useful for clarifying the etiology and the abnormal anastomosis. In cases of hemoptysis with an uncommon etiology, video-assisted thoracic surgery with surgical resection of the bleeding vessel is the definitive management. *(Thorac Med 2018; 33: 20-26)*

Key words: bronchiectasis, fistula, hemoptysis, non-bronchial artery

Introduction

Massive hemoptysis is defined as expectorated blood in excess of 200 mL within 24 h, and is life-threatening because of concomitant hypotension, airway obstruction, or blood loss. It is a pulmonary emergency caused by various underlying conditions [1], typically lung malignancies and chronic inflammatory conditions (including tuberculosis, bronchiectasis, and lung abscess) [2].

Non-bronchial systemic arteries have been identified recently as a crucial origin of bleeding in cases with massive hemoptysis. The progressiveness of the clinical presentation and the unpredictable development of life-threatening hemoptysis demand intensive evaluation and management. We herein report a rare case of
massive hemoptysis in a patient with focal bronchiectasis and left inferior phrenic artery (IPA)-to-pulmonary artery fistula. Bleeding was terminated through a surgical approach.

Case Report

The patient was a 71-year-old non-smoking woman who had a medical history of old pulmonary tuberculosis and bronchiectasis. She had symptoms of non-productive cough lasting 1 month and hemoptysis on the day before admission. The initial amount of expectorated blood was 2 bowlfuls. After treatment with a hemostatic agent, the patient expectorated 80 mL of bright-red blood within 6 h. No coagulopathy or thrombocytopenia was detected.

Physical examination revealed mild bilateral crackles. She was not febrile or hypoxic. Chest radiograph revealed left lower lobe infiltrates. Chest contrast-enhanced computed tomography (CT) showed a 2.4-cm serpentine hypervascular lesion in the left lingular lobe abutting the pericardial region (Figure 1A), with focal bronchiectasis in the left lingular lobe (Figure 1B). Arteriographic embolization was performed due to the initial impression of bronchiectasis with left IPA feeding, which we suspected to be the possible source of the massive hemoptysis. However, angiography disclosed an artery-to-artery fistula at the left lingular lobe, abutting the pericardial region, in which the left IPA communicated with the inferior branch of the left pulmonary artery (Figure 2). The finding of vascular anastomosis

Fig. 1. Chest Contrast-Enhanced Computed Tomography. A hypervascular lesion (arrow) in the left lingular lobe, abutting the pericardial region (A). Focal bronchiectasis (arrow) in the left lingular lobe (B).

Fig. 2. Angiography. An engorged vascular anastomosis observed between the inferior branch of the left pulmonary artery (black arrow) and the left inferior phrenic artery (white arrow).
was also found on 3-dimensional reconstruction imaging (Figure 3). Due to the high flow of the pulmonary artery and the tortuosity of the IPA, we aborted the above therapeutic procedure.

We decided subsequently to perform video-assisted thoracoscopic surgery (VATS) with segmentectomy of the lingular lobe to prevent ongoing massive hemoptysis. The main supply artery arose from the left IPA and progressed to the lingular lobe of the lung (Figure 4A), and bronchiectasis (Figure 4B) was observed during VATS. After ligation of the supply artery, the patient gradually recovered, with progressive improvement of hemoptysis. She was discharged after 14 days in the hospital.

Discussion

The present case report describes an elderly woman who had a left IPA-to-pulmonary artery fistula with focal bronchiectasis, resulting in life-threatening hemoptysis. This condition is a respiratory emergency with a mortality rate of about 80% in the absence of further appropriate management [3-4]. It is important to confirm that lung vessels are the source of bleeding, and this involves excluding bleeding from vessels of the gastrointestinal tract or nasopharynx. The volume of bleeding usually correlates with patient outcome and duration of hospital stay. While patients with mild hemoptysis have a shorter hospitalization and usually have a better prognosis, patients with massive hemoptysis need a longer hospital stay and more surgical intervention [5]. The major causes of hemoptysis include bronchiectasis, bronchitis, pneumonia, and lung cancer. In a series reported in 1997, bronchiectasis accounted for 20% of
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In another study, bronchiectasis accounted for about 35% of cases with life-threatening hemoptysis [2].

The bronchial artery, from the systemic circulation, accounts for about 90% of bleeding sources in massive hemoptysis, and the pulmonary artery, from the pulmonary circulation, accounts for the remainder [6]. However, the non-bronchial systemic circulation may be involved in 10-30% of life-threatening hemoptysis cases [7-8]. Missing the non-bronchial systemic arteries in the angiography examination may result in recurrent bleeding. Many experts have suggested that a comprehensive examination of arteries involved in non-bronchial systemic supply should be performed in hemoptysis cases [9]. Also, some systemic arteries may induce hemoptysis, including the internal mammary, intercostal, and thyrocervical arteries, and the IPA. The IPA arises mainly from the celiac artery or aorta, and is well known as a provider of extrahepatic blood supply for hepatocellular carcinomas [6]. In many published cases of hemoptysis of IPA origin, the lower lobe of the lung was impacted with chronic inflammation and was apparently the source of bleeding [10-13]. Transpleural systemic-pulmonary artery anastomosis may occur in patients with tuberculosis, bronchiectasis, cystic fibrosis, or chronic pneumonia [10]. The possible mechanism underlying the development of this kind of transpleural systemic-pulmonary artery anastomosis is decreased pulmonary blood flow and pleural fibrosis. Some investigators have observed a tendency for a higher incidence of recurrent hemoptysis in patients with a systemic-pulmonary artery shunt [14-15]. In the present case, a left IPA-to-pulmonary artery fistula was the origin of bleeding in the lingular lobe, rather than in the lower lobe. The patient had focal bronchiectasis, and an abnormal systemic-to-pulmonary artery fistula developed due to chronic inflammation. Increased blood flow led to dilatation of the systemic arteries; thus, small vessels easily ruptured, with bleeding resulting from systemic pressure.

Pulmonary sequestration is a congenital disorder characterized by focal areas of anomalous lung tissue that lack normal communication with the tracheobronchial tree, and receive systemic arterial supply. In our case, there was non-bronchial systemic arterial circulation in the left lingular lobe. Therefore, pulmonary sequestration should be included in the differential diagnosis for the patient. However, chest contrast-enhanced CT and angiography of our patient clearly revealed focal bronchiectasis adjacent to an artery-to-artery fistula in the left lingular lobe. Clearly, there was normal communication with the tracheobronchial system in the present case. Furthermore, the lesion in our case was located in the lingular lobe, but a majority of pulmonary sequestration has been located in the lower lobes (96%) [16]. Based on the above findings, pulmonary sequestration could be excluded from the differential diagnosis of our patient.

Resuscitation and protection of the airway are the initial approaches to managing life-threatening hemoptysis. The next step is directed at localizing the source and cause of bleeding, and the final step includes intervention using definitive treatments to prevent recurrent bleeding. Bronchoscopy and angiography are the examinations of choice to confirm the site of bleeding and to perform therapeutic intervention. Bronchial artery embolization (BAE) is performed extensively for the treatment of hemoptysis, especially in severe, non-surgical cases. However, other sources of non-bronchial
systemic arterial supply and the IPA are accordingly assumed to diminish the therapeutic outcome of embolization. Furthermore, technical failure of BAE occurs in approximately 13% of cases, and is commonly caused by non-bronchial arterial supply from systemic vessels, such as the mammary, phrenic, intercostal, or subclavian arteries [17]. Complications of BAE include systemic embolization, vessel perforation, intimal tears, hemoptysis, pyrexia, and neurological complications. Surgery is effective for the management of localized lesions related to hemoptysis. Surgical resection is considered when BAE is unavailable, or if it is unlikely that the bleeding can be controlled by embolization. Surgical resection remains the management of choice for the treatment of life-threatening hemoptysis caused by particular cases of hydatid cyst, arteriovenous malformations, leaking aortic aneurysms, bronchial adenoma, iatrogenic pulmonary rupture, or hemoptysis associated with mycetoma, which are resistant to other means of management [18]. Compared to other therapies, surgical resection of the bleeding vessel is considered the definitive treatment. Furthermore, hemoptysis is usually complicated with respiratory distress, which can lead to desaturation after respiratory failure.

In our case, there were 2 reasons for the failure of BAE. First, the lumen of the artery-to-artery fistula was too large, which led to a very high flow rate. As a consequence, a steel coil could not be fixed to the precise site of the feeding vessel. Second, the IPA of our patient was too tortuous, and the coaxial microcatheter could not be extended to the distal part of the IPA. For patients with life-threatening hemoptysis for which BAE is ineffective, immediate surgery can be used to prevent the development of respiratory failure and the progression of bleeding.

References


左肺舌葉之左下橫膈動脈到左肺動脈廔管合併大量咳血：
病例報告與文獻回顧

林靖傑 黃才旺 * 柯凱雄 ** 彭萬誠 *** 簡志峯 *** 陳盈潔 ***

大量咳血是一種肺部急症，需要立即進行支氣管血管造影栓塞或外科手術。它發生於各種的肺部疾病，通常起源於支氣管動脈。我們的案例報告是一個71歲女性，曾有肺結核和支氣管擴張症的病史，最近1個月陸續有咳嗽症狀，入院前1天開始出現大量咳血，胸部電腦斷層發現左側肺部舌葉有異常的血管顯影合併支氣管擴張症，肺血管造影證實為一左下橫膈動脈到左肺動脈之廔管。最後，藉由胸腔內視鏡輔助手術切除異常部分的肺葉，並改善咳血的症狀。(胸腔醫學 2018; 33: 20-26)

關鍵詞：支氣管擴張症，廔管，咳血，非支氣管動脈