Anomalous Pulmonary Vessels – An Unusual Cause of Hemoptysis

MA HOQUE[a], MMR MIA[b], NK SARKAR[c], SMZ RASHA[d]

Abstract:

Hemoptysis is a serious medical emergency and warrants prompt and comprehensive evaluation. Anomalies of thoracic vasculature constitute an uncommon cause of hemoptysis. Pulmonary vasculature is affected by a sizeable number of anomalies. Multidetector computed tomography (MDCT) of chest is an important imaging modality for evaluating such anomalies. It plays a pithy role in the diagnosis and planning of definitive treatment by identifying the origin and course of the aberrant artery. Definitive treatment is surgical (lobectomy or segmentectomy) or endovascular approach (e.g., coiling, embolization). We report a young man with hemoptysis originating from an aberrant systemic artery arising from the descending thoracic aorta to the left lower lobe. He underwent lower lobectomy. The definite treatment led to cessation of hemoptysis.

Keywords: Anomalous artery, hemoptysis, lobectomy

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Introduction:

Haemoptysis is defined as coughing out of blood originating from airways below the level of glottis.[1] A number of common different etiologies exist with lung cancer, infections such as tuberculosis, bronchiectasis, bronchitis, or pneumonia, and certain cardiovascular conditions. But other rare and uncommon conditions also exist posing diagnostic challenge. The presentation varies, ranging from mild, blood-tinged mucus to serious and potentially life-threatening hemorrhage.[2]

Hemoptysis originates from the bronchial circulation in 95% of cases, and from the pulmonary circulation in the rest.[2] Bleeding from the bronchial arteries has a greater propensity to cause massive hemoptysis, as it is a high-pressure circulatory system. Even for the most experienced clinicians, management of hemoptysis is often a nightmare. Ultimately, management of this condition requires a multi-disciplinary team approach, including interventional pulmonologists, intensivists, radiologists, interventional radiologists, and thoracic surgeons.[3] Sometimes, hemoptysis create a diagnostic dilemma because of difficulties in detecting the site of bleeding and underlying cause, especially if it is a rare one like, systemic vascular anomalies to normal lung.

Case study

We report a 21-year-old man who visited at the outpatient department of our facility with the complaints of recurrent hemoptysis and chest heaviness. He was in usual state of health 4 years back, since then he complained of recurrent episodes of hemoptysis which was scanty at earlier period, later moderate in amount. Blood was fresh; there was no fever, cough, weight loss, history of trauma or previous pulmonary tuberculosis, and bleeding from any other sites. He consulted several physicians, but was not getting improved. Six months before presentation, he felt heaviness in the left side of chest, marked on exertion.

On physical examination, he was found in usual state of health, with BMI 29 kg/m^2, pulse 96 beats/minute, respiratory rate 18 breaths/ minute, blood pressure 110/ 70 mmHg. He was not anemic, neck glands were not enlarged, there was no organomegaly or bony tenderness. Respiratory and cardiovascular system

a. Dr. Mohammad Anamul Hoque, Registrar, Department of Thoracic Surgery, NIDCH, Dhaka, Bangladesh

b. Dr. Md. Mofizur Rahman Mia, Associate Professor, Department of Thoracic Surgery, NIDCH, Dhaka, Bangladesh

c. Dr. Nirmal Kanti Sarkar, Assistant Professor, Department of Respiratory Medicine, NIDCH, Dhaka, Bangladesh

d. Dr. Sheikh Md. Zakirullah Rasha, Junior Consultant, Department of Thoracic Surgery, 250-Bedded TB Hospital, Dhaka, Bangladesh

Address of Correspondence: Dr. Mohammad Anamul Hoque, FCPS (Surgery), FCPS (Thoracic Surgery), Registrar, Department of Thoracic Surgery, National Institute of Diseases of the Chest and Hospital (NIDCH), Mohakhali, Dhaka, Bangladesh. E-mail: dr.anamul27@gmail.com

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examination revealed normal findings. Chest x-ray was unremarkable and plain computed tomography scan of chest showed dilated left lobar pulmonary arterial branch with prominence of segmental branch of the left lower lobe [Fig. 1]. Fiberoptic bronchoscopy showed normal findings. We decided to do contrast-enhanced computed tomography scan of chest, which revealed a large aberrant systemic artery originating from descending thoracic aorta supplying the left lower lobe (predominantly basal segment) without sequestration. Bronchial and pulmonary venous anatomy was normal [Fig. 2, A and B]. In view to ongoing hemoptysis, we shared the possible treatment options with the patient and caregivers and decided to move for the definite treatment – left lower lobectomy.

Under general anesthesia and one lung ventilation (OLV), postero-lateral thoracotomy incision was done. No collection was found in thoracic cavity, only mild adhesions were noted between lung and chest wall. All adhesions were freed meticulously and a thorough inspection was taken. Lower lobe was found grossly congested [Fig. 3A] with multiple small nodules and a normal upper lobe. During dissection, we found a large aberrant artery arising from descending thoracic aorta, supplying the lower lobe [Fig. 3B]. Multiple dilated bronchial arteries and dilated inferior pulmonary vein were also noted. All vessels were ligated and dissected. Lower lobar bronchus was separated, sutured and lower lobectomy was done. Surrounding lung injury and any air leak was checked, and ensured that remaining lung has been expanded. After securing haemostasias, a 32 Fr chest drain tube was kept in-situ. Wound was closed by layers and skin was closed by stapler. Resected specimen was sent for histopathology. Post-operative period was uneventful. On the 5th post-operative day, chest drain was removed and the patient was discharged on 10th post-operative day. On 3rd, 6th and 12th month follow-up, he was doing well without any further hemoptysis.

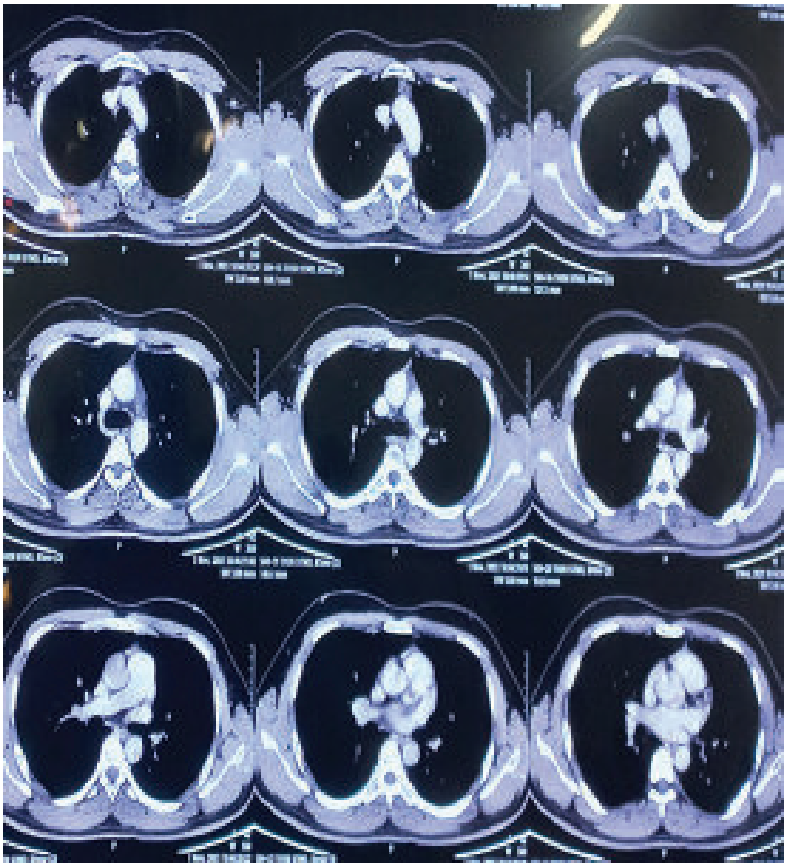

Figure 1: *Plain CT scan of chest.*

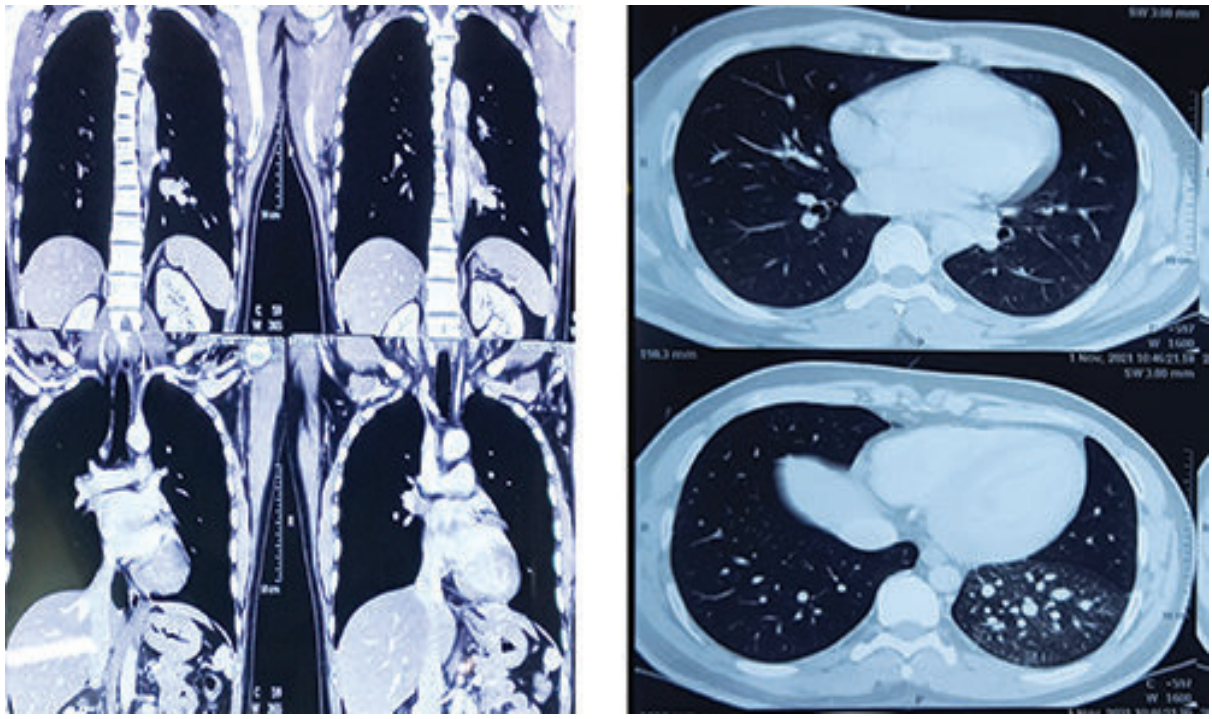

Figure 2: *Contrast-enhanced CT scan of chest [coronal view (A), and axial view (B)]*

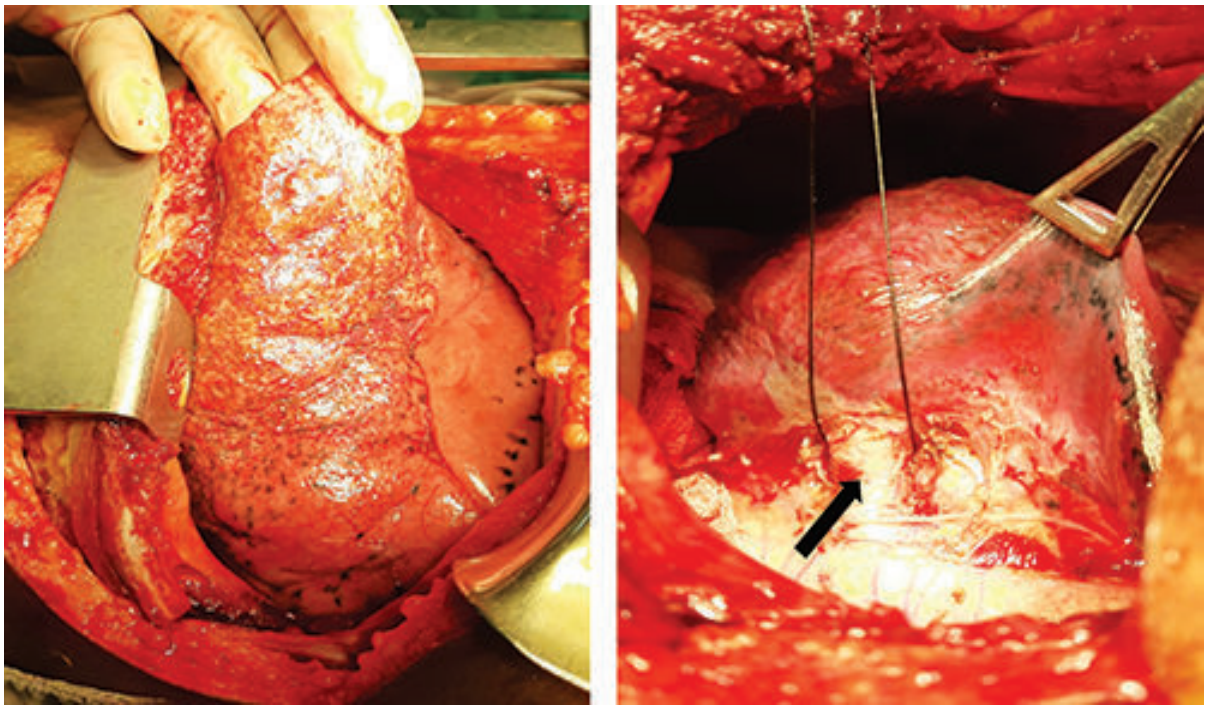

Figure 3: *Pathological lower lobe [A], and large aberrant artery (tied, black arrow) [B]*

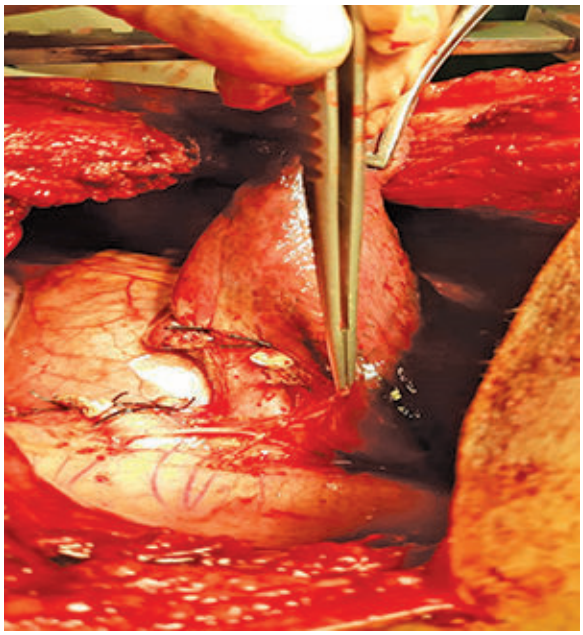

Figure 4: *Following left lower lobectomy*

Discussio

Hemoptysis is often an alarming and worrisome symptom for both the patient and the physician, having variable severity, ranging from blood-streaked sputum to life-threatening hemorrhage. Though massive hemoptysis constitutes less than 20% of all cases, it may lead to devastating consequences if left untreated or not managed properly.[4] Massive hemoptysis should be promptly identified as it poses a significant risk of mortality which has been reported to be as high as 80% without appropriate management.[5] The five most common causes of hemoptysis worldwide, in random order, remain bronchiectasis, lung cancer, tuberculosis, lower respiratory tract infections, and mycetoma[6], but sometimes the underlying cause may be a rare one, creating a diagnostic chameleon, like our case.

Systemic arterial supply to a lung with normal bronchial communication (without bronchial sequestration) and without cardiovascular or pulmonary abnormalities is a rare congenital abnormality. It is of two types: isolated systemic arterial supply to normal lung (ISSNL) and systemic arterial supply associated with normal pulmonary artery (dual supply).[7] In 1946, Pryce described this abnormal arterial supply to lung as intralobar pulmonary sequestration, but soon realized that it is a different entity. He categorized this condition in 3 groups: type I - isolated pulmonary arterial supply to normal lung (without sequestration), type II - the systemic arterial supply both normal and abnormal lung, having no communication with the tracheobronchial tree, and type III - only non-communicating abnormal lung receiving the aberrant systemic arterial supply.[8]

Embryologically, this condition is believed to be due to failure of regression of the primitive aortic branches to the developing lung bud. During normal lung development, the primary lung bud receives a vascular plexus from the dorsal aorta which regresses with the development of the main pulmonary artery.[9] Variation depends on different degrees of embryonic defects.

The hemodynamics of the involved lung segment is adversely affected by transmission of high systemic pressure to the low-pressure pulmonary vasculature. Focal pulmonary hypertension develops leading to development of high-output cardiac failure. The basal segments of left lower lobe are the most commonly affected site, although rarely other sites can be affected (right middle lobe).[9] The systemic artery most commonly arises from the thoracic aorta, but can arise from abdominal aorta or celiac axis and even more rarely from left subclavian and internal mammary artery. These vessels usually enter the lung at the level of pulmonary hilum or at the level of inferior pulmonary ligaments. The venous drainage is usually normal and accomplished by inferior pulmonary vein draining into the left atrium.

Presentation may be variable; ranging from cough, expectoration and recurrent pneumonia to different degrees of hemoptysis.[10-12] This is because the high-pressure artery arising from descending aorta exerts an increasing pressure in the pulmonary capillaries and veins, leading to rupture of vessels and intra-alveolar hemorrhage.[13] Persistent pulmonary venous hypertension may lead to left atrial dilation, congestive heart failure, exertional dyspnea and chest pain.[14] This is the possible explanation of exertional chest pain of our patient.

Chest x-ray may show ill-defined retrocardiac opacity corresponding to the anomalous artery. CT angiography can demonstrate the origin and course of aberrant artery with the advantage of multiplanar and 3D reconstructions.[15] There are different modalities of treatment. When aberrant systemic artery is the only supply, surgery (lobectomy or segmentectomy) is the

treatment of choice. When the involved segment has dual blood supply, either occlusion of the aberrant vessel by surgical ligation or endovascular treatment (coiling, embolization) may be tried.[15]

The index case had a systemic aberrant supply from the descending thoracic aorta to the left lower lobe with normal pulmonary circulation, henceforth we approached for left lower lobectomy and successfully managed the patient.

Conclusion

We present a case of hemoptysis having symptoms for many years. Though uncommon, the vascular anomalies should be kept in mind as an underlying cause of hemoptysis in an otherwise healthy person. In-depth evaluation with MDCT, pulmonary angiogram, and bronchoscopy are valuable armaments for diagnosis of underlying pathology and localization of bleeding point. Among different treatment options, the best one should be offered as per institutional practice and facility, that was done in our patient.

Conflict of interest

There is no potential conflict of interest to declare.

Disclosure

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

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