

Massive haemoptysis in a woman with left lower lobe pulmonary artery interruption—A rare clinical presentation

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Abstract

Proximal interruption of the pulmonary artery (PA) is a rare congenital vascular anomaly with varying presentation. These patients can be asymptomatic or symptomatic with breathlessness, haemoptysis, recurrent chest infections and pulmonary hypertension. Here, we present a patient who presented with massive haemoptysis secondary to interruption of the left lower lobe PA. To the best of our knowledge, massive haemoptysis due to isolated interruption of the left lower lobe PA has been rarely reported in the English medical literature.

KEYWORDS

congenital, haemoptysis, interruption of the pulmonary artery, lobe, pulmonary

INTRODUCTION

Proximal interruption of the pulmonary artery (PIPA), a condition secondary to congenital developmental anomaly of the sixth branchial arch, is described as absence of pulmonary artery (PA) or termination of it within 1 cm of its origin.¹ Most patients with proximal interruption of the left PA have associated cardiovascular congenital anomalies. In contrast, most of the right PA interruption patients are asymptomatic and may be diagnosed incidentally during the adulthood.^{1,2} The prevalence of interruption of the PA is approximately one in 200,000.^{1,2} The median age at diagnosis of isolated interruption of the PA is 14 years (range, 0.1–58 years).³ Massive haemoptysis due to isolated interruption of the left lower lobe PA has been rarely reported in the literature. In this report, we describe a middle-aged patient with PIPA presenting with massive haemoptysis with good surgical outcome.

CASE REPORT

A 45-year-old previously healthy Indonesian-born woman presented with acute intermittent massive haemoptysis over 2 days. Haemoptysis was estimated to be about 1000 ml of fresh blood over 24 h. There were no preceding infective symptoms or risk factors for pulmonary embolism. She had no contact or history of tuberculosis and was not on any regular medications including antiplatelet or anti-coagulation therapy. Despite a significant drop in haemoglobin from 152 to 114 g/L, she remained haemodynamically stable. White cell count, liver and renal function, connective tissue disease screening, vasculitis screening, thrombophilia work-up and coagulation profile were normal. Mantoux test was negative. A contrast computed tomography (CT) chest and a CT pulmonary angiogram demonstrated complete occlusion of the left lower lobe PA with associated bronchial arterial hypertrophy with multiple collateral vessels arising from the bronchial arteries (Figure 1). Reconstructed image of the CT

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pulmonary angiogram demonstrated absence of the left lower lobe PA (Figure 2). The patient was treated with tranexamic acid. Her haemoptysis resolved over a period of

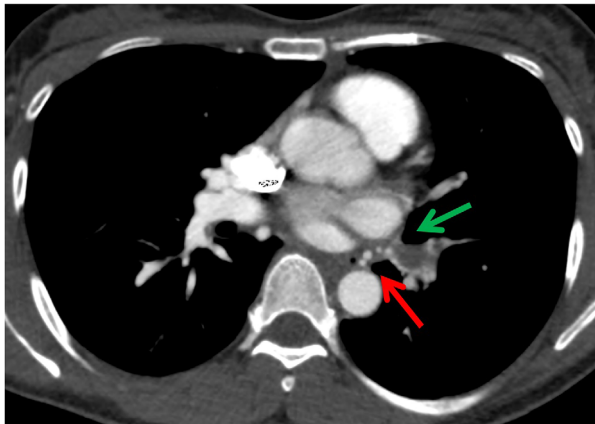


FIGURE 1 Computed tomography bronchial angiogram: abrupt cessation of opacification of the left lower lobe pulmonary artery (green arrow) and multiple hypertrophied collateral bronchial arterial supply (red arrow)

5 days without any active intervention. Bronchoscopy performed 2 days after presentation demonstrated complete occlusion of the left lower lobe bronchus with clotted blood. The bronchial anatomy was otherwise normal. Echocardiogram showed no cardiac abnormalities and PA pressure was not elevated.

She underwent a video-assisted left lower lobectomy without any major complications and recovered well and has had no further episodes of haemoptysis.

The resected left lower lobe macroscopically demonstrated termination of the left lower PA after 15 mm of origin. Thromboembolism or vasculitis was not identified. The findings were suggestive of left lower lobe PA interruption (Figure 3).

DISCUSSION

Haemoptysis is a common clinical presentation encountered in clinical practice. The source of bleeding could be from the bronchial circulation or the pulmonary circulation. Bleeding from a bronchial artery is the cause of massive haemoptysis

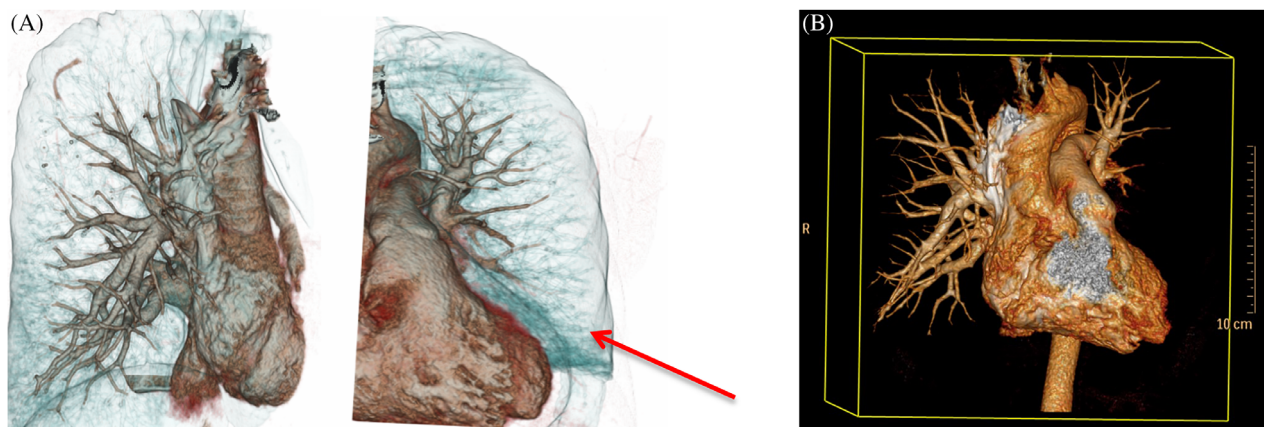


FIGURE 2 (A, B) Reconstructed computed tomography (CT) image of the right and left pulmonary arterial supply. Note the absence of the left lower lobe pulmonary arterial supply (arrow in A) when compared to the right lower lobe

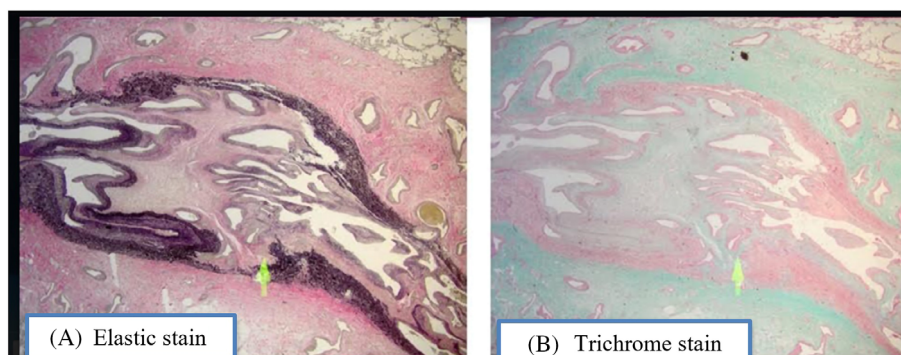


FIGURE 3 (A, B) Elastic stain and trichrome stain ($\times 20$ magnification). Arrows highlight the large muscular artery with abrupt transition to small muscular arteries. Histopathology shows the abrupt interruption of the left lower lobe pulmonary artery (PA). Approximately 8 mm from the proximal left lower lobe PA, there was an abrupt transition to tortuous muscular arteries, of variable wall thickness and size, occupying the lumen of the PA. There are also prominent muscular arteries within the adventitial layer. The PA appeared to be interrupted by these muscular arteries to a distance of approximately 19 mm.

in 90% of cases. PIPA is an uncommon developmental anomaly and interruption of the lobar PAs is even rarer.³ The term ‘interruption’ is preferred to ‘absence’ of the PA, as the portion of the vessel that is in the lung is usually intact and patent.² In proximal interruption, the PA ends blindly at the hilum, and blood is supplied to the lung through collateral systemic vessels, mainly bronchial arteries but also internal mammary, intercostal, innominate and subclavian vessels.^{3,4} In the majority of cases, the interrupted PA is contralateral to the side of the aortic arch.^{1,2} Most patients with PIPA are symptomatic, with recurrent pulmonary infections (37%), dyspnoea or limited exercise tolerance (40%), haemoptysis (20%) and pulmonary hypertension (19%–44%).^{3,5} The likely source of acute bleeding among patients presenting with massive haemoptysis is due to rupture anastomoses vessels between bronchial and pulmonary vasculature that have high systemic arterial pressure. However, haemoptysis is usually minor and self-limited, unlike in our patient.^{2,3,5} Patients with PIPA have ipsilateral volume loss with mediastinal shift, elevated ipsilateral hemidiaphragm and paucity of vascular markings and hyperinflation.² They also demonstrate fine reticular opacities in the periphery of the affected lung due to collaterals between systemic and pulmonary vasculature and post-inflammatory fibrosis. This can be mistaken for tuberculosis. Pleural thickening and rib notching due to collaterals may be seen.^{3,4} In our patient, these findings were not noted and we believe this was due to confinement of the anomaly only to the left lower lobe PA. The absence of risk factors for pulmonary embolism, along with absence of any evidence of vasculitis and thromboembolism on macroscopic examination of operative sample and on histology, excluded the possibility of chronic pulmonary embolism in this patient.

The patient described in this report presented with massive haemoptysis and underwent investigations to establish the diagnosis and rule out the possibility of other diagnosis including infection, pulmonary embolism and so on. The CT bronchial angiogram demonstrated multiple hypertrophied collateral bronchial vessels and no identifiable bleeding point. She was considered to be high risk for recurrent massive haemoptysis. Hence, a video-assisted left lower lobe lobectomy was performed to alleviate the chance of future bleeding. A previous review found that surgical procedures were performed in 17% of patients and included pneumonectomy and revascularization surgeries. The overall mortality rate was 7%.³

AUTHOR CONTRIBUTION

All authors contributed equally to the conception or design of the work, the acquisition, analysis or interpretation of data for the work; drafting of the work or revising it

critically for important intellectual content; and final approval of the version to be published.

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CONFLICT OF INTEREST

None declared.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analysed in this study.

ETHICS STATEMENT

Appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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