

Successful Lobectomy of Pulmonary Arteriovenous Malformations in a Patient Misdiagnosed as Tuberculosis and Polycythemia Vera: A Case Report

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Case report

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Abstract

Background: Pulmonary arteriovenous malformations are uncommon conditions of abnormal communications between pulmonary arteries and veins, which are most commonly congenital in nature. Although such condition is not extremely rare, it is an challenge in the differential diagnosis of pulmonary problems such as hypoxemia and pulmonary lesions.

Case presentation: We report a meaningful case of a 23-year-old male presented with elevated hemoglobin (23.0 g/dl) on admission. Physical examination revealed cyanosis, digital clubbing and low oxygen saturation on room air. The patient was initially diagnosed as polycythemia vera while the subsequent result of bone marrow aspiration was negative. During further assessment, pulmonary arteriovenous malformations were detected by computed tomographic angiography of pulmonary artery. Lobectomy was successfully performed with significant increase of oxygen saturation from 86% to 98%. The patient received an uneventful postoperative recovery and was followed up for nearly 5 years without recurrence.

Conclusions: Pulmonary arteriovenous malformations should be suspected in the patients with central cyanosis, digital clubbing, polycythemia and pulmonary lesion and without cardiac malformations. Embolization or surgery is highly recommended to reduce the risks caused by pulmonary arteriovenous malformations.

Introduction

Pulmonary arteriovenous malformations (PAVMs) are an uncommon disease with an latest estimated incidence of about 1 in 2630 in population scanned by computed tomography (CT)[1]. It was first described by Churton in 1897[2]. Owing to the lack of intervening capillary bed, the patients with PAVMs have the predisposition to complications like hypoxemia, hemoptysis, ischemic stroke and cerebral abscess. Treatment is recommended to prevent later complications by embolization or surgical lobectomy. Herein, we report a 23-year-old male who presented with elevated hemoglobin which was initially misdiagnosed as polycythemia vera on admission. His clubbed fingers and pulmonary lesion were regarded as the sequela of previous tuberculosis (TB). Our case showed great learning value of differential diagnosis in patients with PAVMs. Furthermore, a brief literature review of PAVMs was performed.

Case Report

A 23-year-old male was admitted to the department of hematology because of elevated hemoglobin during routine check-up. Physical examination revealed digital clubbing, cyanosis of oral mucosa and extremities. Remarkable signs on laboratory work up included decreased oxygen saturation on room air of about 85% and elevated hemoglobin of about 23.0 g/dl. Chest roentgenogram suggested remote infectious lesion in the left upper lobe (Fig. 1A) and echocardiography showed no cardiac malformation.

Additionally, the patient reported history of tuberculosis (TB) in his childhood and had been treated with standard anti-tuberculosis therapy for almost 1 year. Family members reported no repeated nosebleed, anemia or similar features mentioned above. The patient had been informed by his doctor that his clubbed fingers were sequela of chronic hypoxia caused by TB and the left pulmonary lesion were residual TB lesion after standard treatment. Bone marrow examination demonstrated non-specific erythroid hyperplasia. The initial diagnosis made by hematologist was polycythemia vera and hydroxyurea was applied. After several courses of treatment, the patient's hemoglobin still kept high (22.5 g/dl) without obvious change and cardiothoracic consultation was scheduled. Considering the digital clubbing, cyanosis of oral mucosa and extremities, elevated hemoglobin, low oxygen saturation and left pulmonary lesion, PAVMs were suspected. Computed tomographic angiography (CTA) of pulmonary artery showed abnormal communications between the left upper pulmonary arteries and veins and PAVMs of left lingual lobe was confirmed (Fig. 1B). No extrapulmonary vascular malformations were detected.

Since PAVMs were diffuse and restricted to left upper lobe of lung, complete feeding arteries embolization was difficult and the patient finally received video assisted thoracoscopic left upper lobectomy. Intraoperatively, diffusely dilated and angiomatous vessels with hemorrhagic tendency were detected on the surface of left upper lobe (Fig. 1C, 1D). The left superior pulmonary vein, left upper lobe bronchus and left superior pulmonary artery were resected by endo-stapler, respectively. After left upper lobectomy, the O₂ saturation immediately increased to 98%.

The histopathological examination confirmed the diagnosis of PAVMs. The patient had an uneventful recovery without any complications and he was discharged home on the 5th postoperative day. The patient has been followed up for about 5 year without cyanosis, dyspnea and other signs of recurrence.

Discussion

PAVMs are defined as abnormal communications between pulmonary arteries and veins without intervening capillary bed. Its prevalence is about 1 in 2630 in population scanned by computed tomography (CT) with slight female predilection[1, 3]. PAVMs can be sporadic, multiple or even diffuse lesions as well as uni- or bi-lateral involved. PAVMs often have lower lobe predominance. Histopathologically, PAVMs manifest as tortuous or direct aneurysmal connection developing between arteries and veins without intervening capillary bed, which caused the loss of "filter capacity".

The etiology of PAVMs could be congenital or acquired. The majority of PAVMs are associated with autosomal dominant disorder called hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome)[4]. Acquired PAVMs are usually secondary to liver cirrhosis, infections, metastatic carcinomas, chest trauma and iatrogenic procedures[3]. Patients with PAVMs are usually asymptomatic, especially in children. The common pulmonary symptoms include cyanosis of oral mucosa and extremities, digital clubbing, hemoptysis and chest pain. Extrapulmonary manifestations are polycythemia, cerebral abscess, stroke or transient ischemic attack, epistaxis and so on[5].

In our case, the patient was initially misdiagnosed as polycythemia vera. There might be several reasons why the other clinical features were ignored when making diagnosis. First, the patient reported the history of TB which made the doctors regard the pulmonary lesion as residual TB lesion. Second, TB is able to cause cyanosis and digital clubbing when severe hypoxemia occurs, which can explain the patient's cyanosis and clubbed fingers since he was a child. We should draw lessons from this case and PAVMs should be suspected when the following combination of clinical manifestations coexists: central cyanosis, digital clubbing, hypoxemia, hemoptysis, elevated hemoglobin, pulmonary lesions on chest roentgenogram or CT, history of cerebral abscess and so on.

Echocardiography findings of PAVMs are usually normal or increased left heart volume overload. Chest roentgenogram could detect abnormal lesions in PAVMs region but with low specificity. CTA of pulmonary artery is of great diagnostic value. Pulmonary angiography is regarded as the gold standard in diagnosis, especially when a therapeutic intervention is planned. In patients who are allergic to iodinated contrast, contrast-enhanced transthoracic echocardiography is highly recommended to be used as a diagnostic method, in which early appearance of microbubbles in the left atrium strongly suggests the existence of PAVMs[3].

The natural history of PAVMs is relatively predictable because of its tendency to increase in size. If untreated, the mortality rate of untreated symptomatic patients ranges from 4 to 22% and even up to 40% in severe cases[6]. Spontaneous regression has rarely been reported.

There is consensus about early intervention in patients with PAVMs to prevent later complications like systemic embolization, pulmonary hemorrhage, ischemic stroke, cerebral abscess, congestive heart failure and so on. Treatment of PAVMs should be made by taking multiple factors such as size, number, location and complications into consideration. Embolization is recommended for first-line treatment of PAVMs[7] while surgical lobectomy is recommended when PAVMs are diffuse, large and restricted to one lobe[8].

Conclusion

We experienced a meaningful case with PAVMs which was initially misdiagnosed as TB and polycythemia vera. We emphasize the importance of maintaining clinical suspicion of PAVMs in patients with features of central cyanosis, digital clubbing, hypoxemia, hemoptysis, elevated hemoglobin, pulmonary lesions on chest roentgenogram or CT, history of cerebral abscess and so on. Surgical lobectomy is suitable for PAVMs which are diffuse, large and restricted to one lobe.

Abbreviations

PAVM

pulmonary arteriovenous malformation; CT:computed tomography; TB:tuberculosis; CTA:computed tomographic angiography.

Declarations

Acknowledgments: Not applicable.

Authors' contributions

XHF AND SW wrote the draft of the manuscript. ZYR revised the article. XHF, NYM, and LWD performed the surgery and contributed to the perioperative care. All authors read and approved the final manuscript.

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Availability of data and materials

The authors declare that all data in this article are available within this published article.

Ethics approval and consent to participate

Informed consent was obtained from the patient.

Consent for publication

Written informed consent for publication of this case report was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

References

1. Nakayama M, Nawa T, Chonan T, Endo K, Morikawa S, Bando M, et al. Prevalence of pulmonary arteriovenous malformations as estimated by low-dose thoracic CT screening. *Intern Med.* 2012;51(13):1677–81.
2. Churton T. Multiple aneurysm of pulmonary artery. *Br Med J*, 1897. 1: 1223–1225.
3. Majumdar S, McWilliams JP. Approach to Pulmonary Arteriovenous Malformations: A Comprehensive Update. *J Clin Med*, 2020. 9(6).
4. Saboo SS, Chamrathy M, Bhalla S, Park H, Sutphin P, Kay F, et al. Pulmonary arteriovenous malformations: diagnosis. *Cardiovasc Diagn Ther.* 2018;8(3):325–37.
5. Shovlin CL. Pulmonary arteriovenous malformations. *Am J Respir Crit Care Med.* 2014;190(11):1217–28.
6. Chowdhury UK, Kothari SS, Bishnoi AK, Gupta R, Mittal CM, Reddy S. Successful lobectomy for pulmonary arteriovenous malformation causing recurrent massive haemoptysis. *Heart Lung Circ.* 2009;18(2):135–9.

7. Hsu CC, Kwan GN, Evans-Barns Hvan Driel ML. Embolisation for pulmonary arteriovenous malformation. Cochrane Database Syst Rev. 2018;1:CD008017.
8. Hayashi K, Motoishi M, Horimoto K, Sawai SHanaoka J. Left upper division segmentectomy with a simultaneous displaced bronchus and pulmonary arteriovenous anomalies: a case report. J Cardiothorac Surg. 2018;13(1):40.

Figures

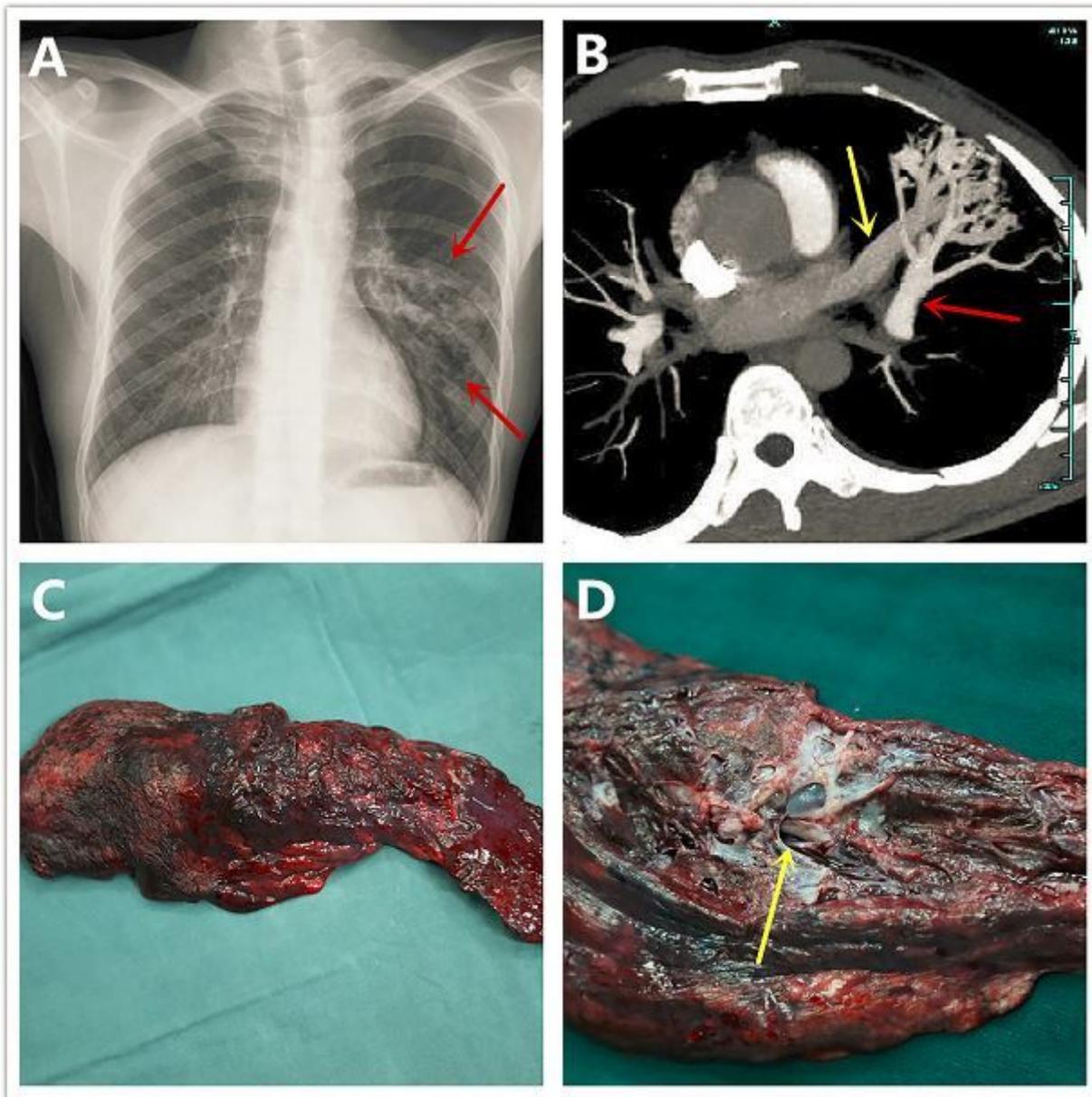


Figure 1

(A) Chest roentgenogram detected the lesions (in red arrows) in the left upper lobe which were initially considered as residual TB lesions. (B) CTA of pulmonary artery showed the abnormal communications between the left superior pulmonary vein (in yellow arrow) and left superior pulmonary artery (in red

arrow). (C, D) Surgical specimen (excised left upper lobe) showed the diffusely tortuous vessels all over the lobe and the vascular cavities were extremely dilated (in yellow arrow). TB: tuberculosis; CTA: computed tomographic angiography.

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