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## Case report

# Pulmonary arteriovenous malformations presenting with upper back pain in an adult: A case report and literature review <sup>☆,☆☆,★</sup>

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## ABSTRACT

Pulmonary arteriovenous malformations (PAVMs) are the abnormal connections between the pulmonary artery and pulmonary veins branches without intervening in the pulmonary vascular bed. Although the most common cause of PAVMs is hereditary hemorrhagic telangiectasia, the etiology of single PAVMs appears to be idiopathic. Dyspnea, cyanosis, cerebrovascular events, and brain abscess are the common clinical manifestations of these lesions, though they may present with a nonspecific symptom as upper back pain. Computed tomography is the gold standard investigation for diagnosing pulmonary arteriovenous malformations, and demonstrating their size and extent before therapy. Transcatheter embolization is the first-choice treatment for patients without severe complications. Surgical intervention is reserved for the lesions not amenable to embolotherapy.

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## Background

Pulmonary arteriovenous malformations are rare vascular abnormalities that occur when the branches of the pulmonary artery and veins directly connect without intervening in the pulmonary vascular bed, resulting in right-to-left shunts [1,2].

Although the most common cause of PAVMs is hereditary hemorrhagic telangiectasia, the etiology of single PAVMs appears to be sporadic. The treatment-related complication of cyanotic congenital heart disease, trauma, and gestational trophoblastic disease may be the rare causes of PVMs [3].

Dyspnea is the common symptom of PAVMs, though this may only be elicited on direct questioning. Hemoptysis and haemothorax may also occur, but the usual presenting symp-

*Abbreviations:* PAVMs, pulmonary arteriovenous malformations; CT, computed tomography.

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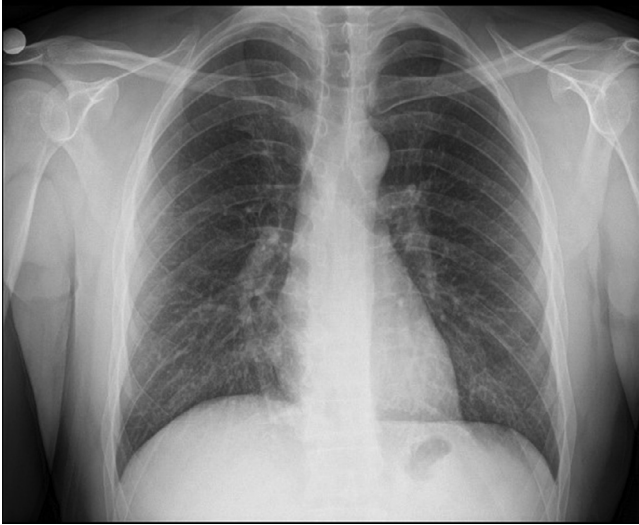
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**Fig. 1 – Normal PA chest x-ray**

toms of an index case are cerebrovascular events including transient ischemia attacks, stroke, or brain abscess [4].

Computed tomography is generally considered the gold standard investigation for diagnosis of PAVMs and demonstrating their size and extent before therapy. However, PAVMs may be visible on chest radiographs in only 60%-90% of instances, where clinically significant PAVMs are present [3].

Transcatheter embolization is often used for the treatment of PAVMs because it is less invasive and easy to repeat, comparing the surgical intervention [5].

The authors present here an unusual case of PAVMs in a 35-year-old male, presented with upper back pain.

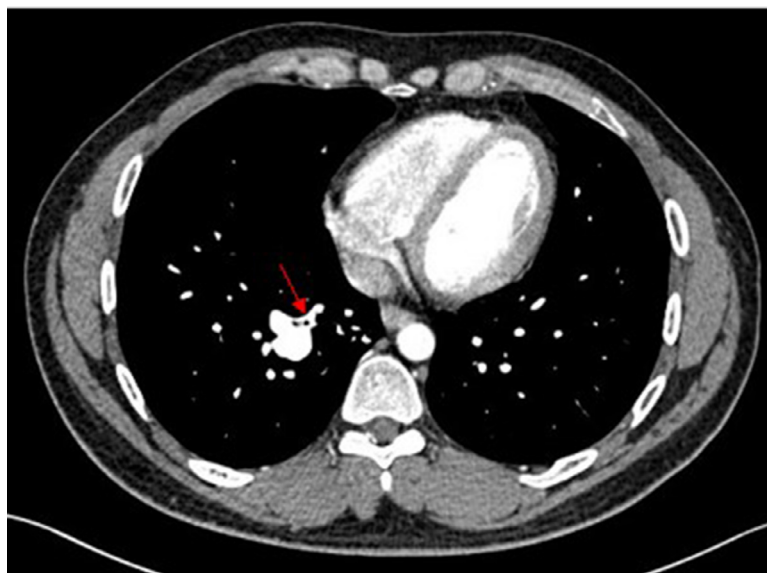
## Case presentation

A 35-year-old male was presented to our hospital, complaining of back pain after getting a cold for one month. He had no known disease in the past and he did not give any history of food or drug allergy and social problems. In physical examination, the patient was conscious, oriented, and cooperative. The findings of cardiovascular, respiratory, abdominal, and musculoskeletal examinations were unremarkable. No findings were noted in favor of hereditary hemorrhagic telangiectasia.

The laboratory analysis yielded hemoglobin: 18.2 g/dl (12–16), white blood cells: 6100/mm<sup>3</sup> (4500–11000), Urea: 23 mg/dl, and Creatinine: 1mg/dl. He was negative for Covid-19. The PaO<sub>2</sub> level was found to be 91 mm Hg, using a nasal prong at 3 L/min.

The chest x-ray was interpreted as normal (Fig. 1). In the chest CT angiogram, a pulmonary arteriovenous malformation with approximately 5.5 cm nidus was observed in the lower lobe of the right lung with a feeding artery and draining vein. The venous drainage of this arteriovenous malformation was draining into the right inferior pulmonary vein (Figs. 2 and 3).

Following multidisciplinary consultation, the patient was referred to the interventional radiology unite for the endovascular procedure and transcatheter embolization. Under general anesthesia and after heparinization, the pulmonary angiogram was obtained with right femoral vein intervention. Multiple PAVMs, some with high output were observed in the lower lobe of the right lung associated with varicose venous enlargements on the venous side of the PAVMs. The transcatheter embolization was successfully performed without any complication and complete embolization was observed on control pulmonary angiography (Fig. 4). The patient was transferred to the intensive care unite for further manage-



**Fig. 2 – : (A) Axial chest CT angiogram, showing a large contrast-enhanced nidus with feeding artery (arrow) in the lower lobe of the right lung. (B) Coronal and sagittal chest CT angiograms, showing a large contrast-enhanced nidus with feeding artery and draining vein (arrow) in the lower lobe of the right lung.**



Fig. 2 – Continued



Fig. 3 – Volume rendering 3D CT image, showing a large nidus with feeding artery (red arrow) and draining vein (green arrow) in the lower lobe of the right lung (color version of figure is available online.)

ment and close monitoring. He had an uneventful recovery and discharged on post-procedural day 3 with a control CT angiogram recommendation after 3 months.

## Discussion

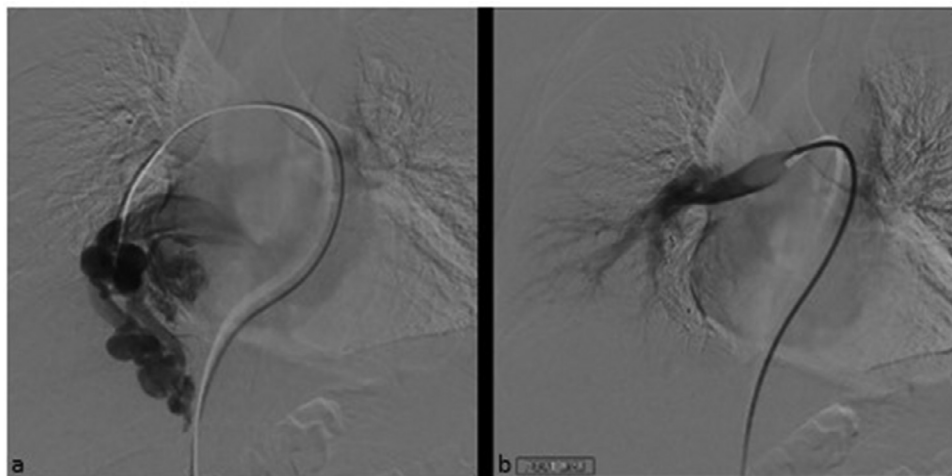
Pulmonary arteriovenous malformations are the abnormal connections between the pulmonary artery and pulmonary veins branches without an intervening the pulmonary vascular bed [2,1]. The incidence of PAVMs is 1 /100 000 population with a male to female ratio ranging from 1:1.5 to 1.8, and often involves the lower lung zones [5].

The first case of PAVM was described by Churton in 1897, and more than 500 cases have been reported in the literature [6].

Although the most common cause of PAVMs is hereditary hemorrhagic telangiectasia, the etiology of single PAVMs appears to be sporadic. The treatment-related complication of cyanotic congenital heart disease, trauma, and gestational trophoblastic disease may be the rare causes of PAVMs [3]. In this case, the patient had no findings for hereditary hemorrhagic telangiectasia, trauma, or other comorbidity resulting in PAVMs. Thus, we postulated that the cause of PAVMs in our patient was idiopathic.

PVMs can be simple; consist of an aneurysmal venous sac communicating with a dilated feeding artery and draining vein, complex plexiform masses with multiple afferent and efferent vessels, and diffuse having multiple small PAVMs affecting a single segment, or every segment of one or more lobes [3]. Our findings were compatible with a complex form of PAVMs.

These lesions can present with dyspnoea, epistaxis, hemoptysis, telangiectasias, cyanosis, clubbing, and gastrointestinal bleeding [1]. However, many patients with pulmonary AVM are reported asymptomatic despite having hypoxia. This phenomenon is thought to be due to chronic compensation including the secondary erythrocytic response [2]. This was consistent with our case. As in the present case, our patient has polycythemia without subjective dyspnea. In our case, the patient complained of upper back pain, which is unusual and



**Fig. 4 – Transcatheter pulmonary angiogram, showing pre (A) and post (B) embolization images.**

described in only 10% of patients with PAVMs, as reported by Shovlin C.L [3].

The most common complications of PAVMs include stroke and brain abscess due to paradoxical systemic embolization resulting from right to left shunting through the PAVM. The PAVM with a feeding artery diameter of  $>3$  mm is strongly associated with various neurologic manifestations like infarction, abscess, and seizure [7].

Imaging techniques including chest radiography, transthoracic contrast echocardiography, CT, catheter angiography, and contrast-enhanced magnetic resonance angiography can be used for screening and diagnosis of PAVMs [7]. However, Computed tomography is generally considered the gold standard investigation for diagnosing PAVMs and demonstrating their size and extent before therapy [3]. The characteristic appearance of a PAVM is a well-defined peripheral nodule, connected into feeding artery and draining veins. Following intravenous injection of contrast material, pulmonary arterial phase CT reveals enhancement of the PAVM sac along with its feeding and draining vessels [7].

Endovascular treatment will be the first choice for patients without severe complications such as severe pulmonary hypertension, renal failure, and early pregnancy [8]. Current evidence suggests that serious complications including stroke and brain abscess may occur in PAVMs with feeding artery diameter  $<3$  mm. Thus, it will be reasonable to perform embolization for any PAVM with a feeding artery diameter 2 mm or larger, and any symptomatic PAVM [9].

Surgical resection of PAVMs is indicated in patients who fail embolotherapy, develop serious bleeding complications despite embolotherapy, having intrapleural rupture of the PAVM, or untreatable contrast allergy, and lesions not amenable to embolotherapy [6].

Historically, coil embolization of the feeding vessel has been the primary treatment of pulmonary AVMs with the anchor and scaffold technique. However, a significant rate of recanalization and adjacent vessel damage is the important limitations of this technique. Therefore, novel Amplatzer vascular plugs with or without coils are recently used for en-

dovascular management of PAVMs with lower recanalization rates (7%), comparing the coil recanalization rate (33%). Nevertheless, vascular tortuous anatomy, small size feeding arteries, and sheath size requirement for plug deployment are the technical problems against plugs usage [10].

The study conducted by Vorselaars, V. M. M., et al showed that a decrease in stroke volume and cardiac output directly occur after embolization of pulmonary arteriovenous malformations that may provide additional insights into the hemodynamic responses after PAVM embolization in patients prone to pulmonary hypertension [11]. Thus, Clinical examination and imaging follow-up evaluation should be carried out 3-6 months after the embolization procedure. The reduction in PAVM sac size and complete resolution of the PAVMs occur within 6 months, though the SpO<sub>2</sub> increases shortly after the procedure [12].

Although this is one of the documented cases, proved with imaging studies and managed by endovascular intervention, lack of a long time follow up may be the only limitation for this case.

## Conclusion

Pulmonary arteriovenous malformations are rare abnormalities of the pulmonary vasculature, resulting in right-to-left shunts. Dyspnea, cyanosis, cerebrovascular events, and brain abscess are the common clinical manifestations of these lesions, though they may present with the nonspecific symptom as upper back pain. Computed tomography is the gold standard investigation for diagnosis of pulmonary arteriovenous malformations and demonstrating their size and extent before therapy. Transcatheter embolization is the first-choice treatment for patients without severe complications and surgical intervention is reserved for the lesions not amenable to endovascular therapy. Long time imaging follow-up is necessary because these lesions may recur after treatment.

## Ethics approval and consent to participate

The manuscript has got an ethical review exemption from the Ethical Review Committee (ERC) of our hospital, as case reports are exempted from review according to the institutional ethical review committee's policy.

## Patient consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

## Author contributions

Concept - HAE; Design – HAE; Supervision - HAE; Resources and data Collection- HAE; Literature Search - MTA; Writing Manuscript – HAE; Critical Review -MTA. All authors have read and approved the final manuscript.

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