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Pulmonary arteriovenous malformation in chronic thromboembolic pulmonary hypertension

Arun Sharma, Gurpreet S Gulati, Neeraj Parakh[1], Abhinav Aggarwal[1]
Departments of Cardiac Radiology and [1]Cardiology, AIIMS, New Delhi, India

Correspondence: Dr. Gurpreet S Gulati, Cardiac Radiology, AIIMS, New Delhi, India. E-mail: gulatigurpreet@rediffmail.com

Abstract

Chronic thromboembolic pulmonary hypertension is a morbid condition associated with complications such as hemoptysis, right heart failure, paradoxical embolism, and even death. There is no known association of chronic thromboembolic pulmonary hypertension with pulmonary arteriovenous malformation. Possible hypothesis for this association is an increased pulmonary vascular resistance leading to the compensatory formation of pulmonary arteriovenous malformation. We present one such case presenting with hemoptysis that was managed with endovascular treatment.

Key words: Bronchial artery embolization; chronic thromboembolic pulmonary hypertension; pulmonary arteriovenous malformations

Introduction

Pulmonary arteriovenous malformations (PAVMs) are abnormal communications between pulmonary arteries and pulmonary veins without an intervening capillary bed.[1] They are most commonly congenital in origin. Acquired causes include hepatic cirrhosis, post bidirectional Glenn shunt, and less commonly schistosomiasis, mitral stenosis, trauma, actinomycosis, Fanconi's syndrome, and metastatic thyroid carcinoma.[1,2] In chronic thromboembolic pulmonary hypertension (CTEPH), hypertrophy of the bronchial circulation may occur as collateral compensation, and this may lead to hemoptysis, seen in 0.1–10% of patients.[3,4] Treatment of hemoptysis in CTEPH can be a therapeutic dilemma as most CTEPH patients require lifelong anticoagulation. We report a case of CTEPH associated with large PAVM, possibly the first of its kind presenting with hemoptysis. The case was managed with bronchial artery embolization and simultaneous vascular plug occlusion of PAVM.

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Case Report

A 27-year-old male presented with dyspnea (grade II) and massive hemoptysis (three episodes, each of 200 ml over 24 h) requiring blood transfusion. He also gave a history of multiple episodes of hemoptysis (150–200 ml/day) in the preceding 2 weeks period. There was no history of cyanosis or epistaxis. Clinical examination showed no mucocutaneous lesions. Chest radiograph showed features consistent with pulmonary artery hypertension (PAH), including prominent pulmonary artery segment, dilated right descending pulmonary artery, and peripheral pruning. In addition, there was a well-defined opacity in the left lower lung zone. Echocardiography suggested moderate

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PAH with normal biventricular function. Computed tomography (CT) angiography confirmed presence of chronic pulmonary thromboembolism [Figure 1A] with chronic thrombus in right descending pulmonary artery, multiple webs in the left pulmonary artery branches and dilated central pulmonary arteries. In addition, it revealed presence of a large (4.9 × 4.6 × 2.0 cm), solitary, simple, PAVM [Figure 1A and B] in inferior lingula with a large (11 mm) segmental feeding artery and corresponding draining vein finally draining into the left atrium. No aneurysm was noted in the feeding artery. CT of the brain and abdomen and bilateral lower limb venous Doppler was normal. Oximetry analysis showed arterial oxygen saturation of 90%. An invasive cardiac catheterization and pulmonary and aortic angiography were then performed to further evaluate for PAH, cause of hemoptysis, and anatomy of the PAVM. The mean pulmonary artery pressure was 36 mmHg with right ventricular systolic pressure of 50 mmHg, suggestive of moderate PAH. Pulmonary angiogram showed the presence of large PAVM [Figure 1C and D]. A descending thoracic aortic angiogram revealed presence of a hypertrophied common bronchial artery [Figure 2A]. This was embolized using gel foam slurry [Figure 2B]. PAVM was occluded using a 16 mm Amplatzer vascular plug (AVP II), delivered through a7F Amplatzer TorqVue delivery sheath in distal segmental feeding artery, and deployment started just proximal to the sac. Check angiogram after 3 min showed complete occlusion of the PAVM [Figure 2C]. Repeat oximetry showed an increase in the arterial oxygen saturation from 90 to 98%. He was discharged in a stable condition after 3 days. Apart from an episode of mild hemoptysis 10 days later that was managed conservatively, no further hemoptysis was seen at 6 months follow-up. Patient was advised lifelong oral anticoagulation with regular follow up. Repeat CT scan at 6-month follow-up showed vascular plug *in situ* without any residual PAVM [Figure 2D].

Discussion

PAVM is a rare vascular anomaly. The lesion may present with epistaxis, dyspnea, stroke, hemoptysis, or hemothorax in 79, 71, 36, 13 and 9% of patients, respectively.[5] An association with hereditary hemorrhagic telangiectasia is seen in up to 80% of cases.[6] Evaluation using CT abdomen is important to look for additional AVMs in the liver and gastrointestinal system. CT head is important to rule out AVMs, brain abscesses, and embolic lesions. Acquired causes include hepatic cirrhosis, post bidirectional Glenn shunt, and less commonly schistosomiasis, trauma, actinomycosis, Fanconi's syndrome, and metastatic thyroid carcinoma.[1,2] Extremely rarely, PAVM may occur in constrictive pericarditis[7] and mitral stenosis.[8] Possible hypothesis put forward to explain its occurrence in these

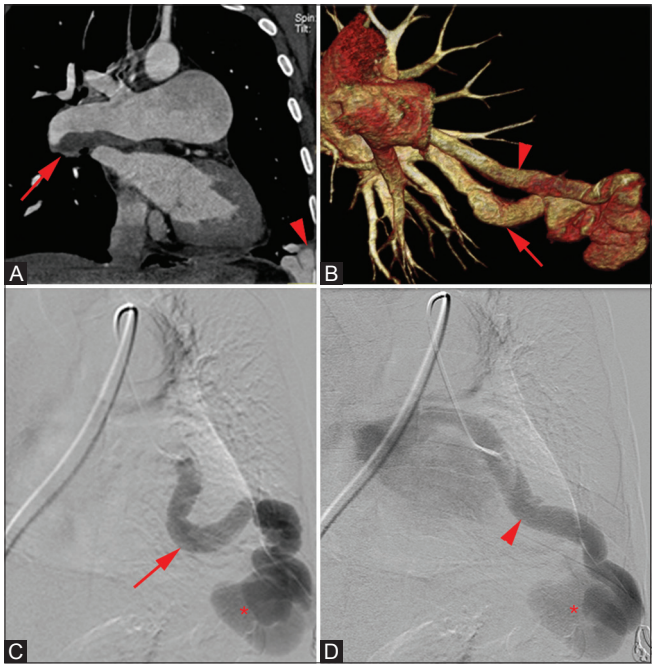

Figure 1 (A-D): (A) Coronal oblique CT image showing partial thrombus in the right descending pulmonary artery (arrow) with presence of left lung PAVM (arrowhead). (B) Volume rendered CT image showing PAVM with large segmental tortuous feeding artery (arrow), sac, and the draining vein (arrowhead). (C, D) Selective pulmonary artery angiograms demonstrating PAVM (*) with tortuous feeding artery (arrow) and corresponding draining vein (arrowhead)

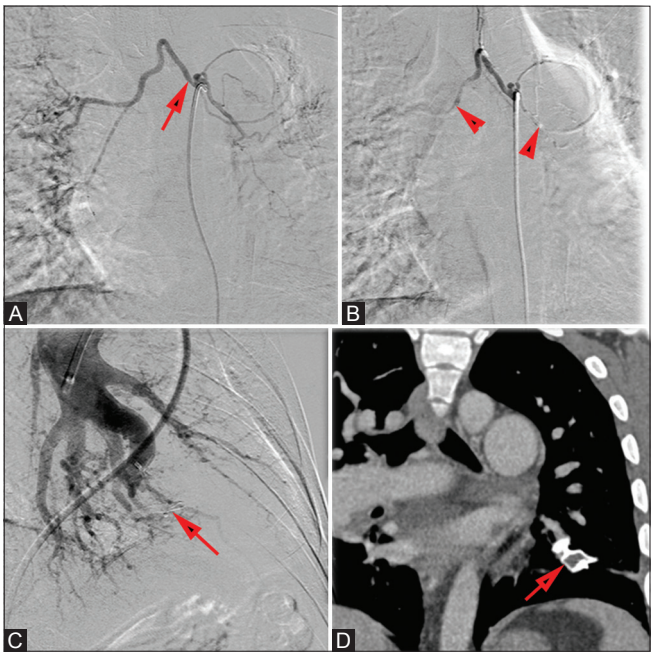

Figure 2 (A-D): (A) Selective bronchial artery angiogram revealing hypertrophied common bronchial artery (arrow). (B) Angiogram after gel foam embolization of common bronchial artery depicts successful embolization (arrow and arrowhead). (C) Descending pulmonary artery angiogram after successful vascular plug deployment depicts complete occlusion of PAVM with device *in situ* (arrow). (D) Coronal CT image at 6-month follow-up depicts no residual PAVM with vascular plug *in situ* (arrow)

conditions is that PAVM being a low resistance part of the circuit develops to compensate for the high pulmonary vascular resistance.

CTEPH is defined as the persistence of pulmonary hypertension (defined as mean pulmonary artery pressure >25 mmHg) after a single or recurrent pulmonary embolism.[9] Hemoptysis is a rare presentation (0.1–10%) that usually develops as a result of bronchial artery hypertrophy and is managed with bronchial artery embolization (as in our case). It is possible that the PAVM in our case developed secondary to the CTEPH, as this can be explained in a manner similar to that in constrictive pericarditis or mitral stenosis. Though difficult to prove, PAVM may also have been the cause for hemoptysis in this case. Given the increased risk of neurological complications in this PAVM with large feeding artery, we performed vascular plug occlusion of PAVM. Treatment of large PAVM (feeding artery diameter >8 mm) with Amplatzer vascular plug is effective with excellent long-term results without major complications.[10] Chance of migration and recanalization with Amplatzer vascular plugs (0–7%) is much less as compared to that of coils (12–15%).[1] In addition, these devices can be repositioned before their final deployment. Moreover, large number of coils may be needed in case of larger vessel, which carries the risk of dislodgement or systemic embolism. Initial follow up in PAVM cases is usually done at 3 months with clinical evaluation, oxygenation, and CT to see residual mass. Further follow-up is done at 6 and 12 months and every 3 years thereafter depending upon the symptomatology. Repeat angiography is performed in cases of persistent or enlarging soft tissue mass which may show residual filling of the feeding artery/sac and persistent early opacification of the draining vein in cases of device failure.

In summary, this is the first reported case illustrating the association between CTEPH and PAVM. Hemoptysis in this case was successfully managed by bronchial artery embolization with simultaneous vascular plug occlusion of PAVM.

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Conflicts of interest

There are no conflicts of interest.

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