

## Diagnosis: Pulmonary arteriovenous malformation (PAVM)

Editor: Husn Frayha

Authors: Mohammed Khalid,\*Neelam Malik,† Suzanne Abbas,†

### Radiological Findings

The routine chest radiograph revealed a sharply defined lobulated soft tissue mass of uniform density in the right lower hemithorax with evidence of tubular densities radiating from the hilum. No calcification was noted within the mass. Likely differentials based on chest radiograph would be granuloma, sequestration, metastasis and AVM.

The lesions on the contrast-enhanced CT chest were identified as subpleural nodular, lobulated masses of uniform density in the right lower lobe anteriorly, with a single feeding artery and a draining vein consistent with PAVM.

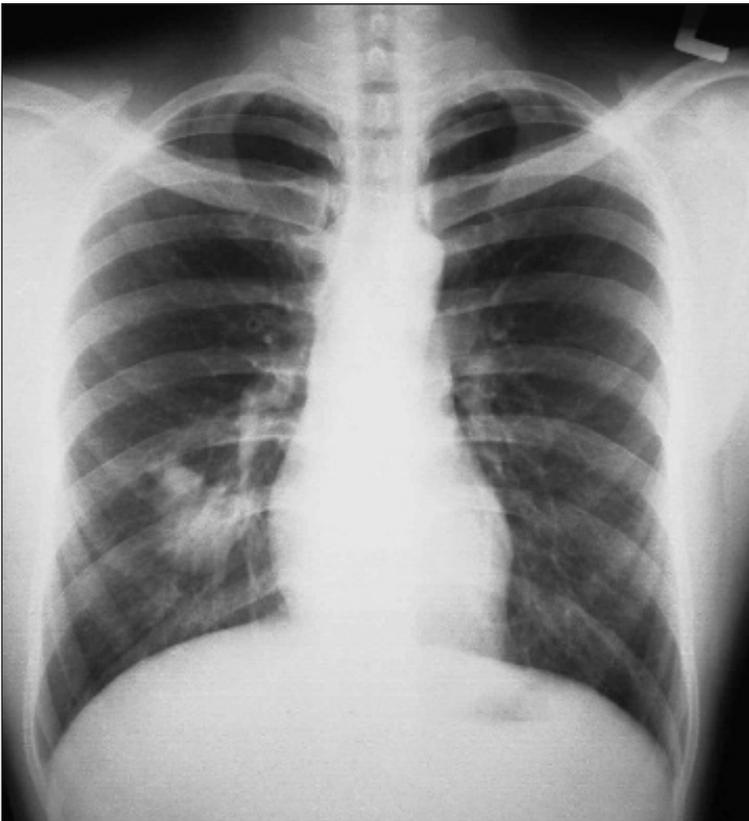
Another radiological test that could demonstrate the lesion is pulmonary angiography, which helps in defining the number, size, location and angioarchitecture of individual PAVM. Pulmonary angiography is considered the gold standard in the diagnosis of PAVM.

### Discussion

PAVMs are rare pulmonary vascular anomalies consisting of abnormal communication between arteries and veins. Since their first description in 1897 by Churton, more than 500 cases have been reported.<sup>1,2</sup> More than 80% of PAVMs are congenital in nature and of these, 47% to 80% are associated with Osler-Weber-Rendu disease or hereditary haemorrhagic telangiectasia (HHT). Conversely, it is estimated that overall 5% to 15% of the population with HHT have a PAVM.<sup>2,3</sup> Both conditions are congenital and the stigmata of either of these two conditions should alert the clinician to the possibility that they may coexist.

Patients are usually asymptomatic. Some patients may have frank cyanosis, or less severely, orthodeoxia. Symptoms often develop between the fourth and sixth decades of life and are more common in PAVM with HHT rather than without HHT.<sup>2,4</sup> The most common complaints in symptomatic patients are epistaxis, caused by bleeding from mucosal telangiectases. Dyspnoea is the second most common complaint followed by haemoptysis, which is the third most common symptom. Epistaxis, telangiectases of skin and mucus membrane and gastrointestinal haemorrhage are common extrathoracic manifestations in patients of PAVM with HHT. The most commonly reported complications relate to the central nervous system, resulting from right to left shunts or coexisting cerebral AVM, with the incidence being 19% to 59%. Such patients may suffer from brain abscess or stroke.<sup>2,4,5</sup>

About 53% to 70% of pulmonary AVMs are seen in the lower lobes, with the left lower lobe being the most common location. They are found in close proximity to the visceral pleura or embedded in the outer third of lung parenchyma. PAVMs have been



**Figure 1.** Chest radiograph showing a lobulated mass of uniform density in the right lower hemithorax with connecting vessels radiating to the hilum.

classified as simple or complex depending upon the number of feeding arteries and veins. Eighty to ninety percent of PAVMs are of the simple type, which is defined as those with a single feeding artery and a single draining vein. There have been cases described with both pulmonary and systemic arterial supply.<sup>5,6</sup>

The chest radiograph is an important diagnostic tool, not only in diagnosis but also in the follow-up of patients with a PAVM. Ninety percent of PAVMs are shown on a chest radiograph. Their classic radiographic features are a sharply defined round or oval mass of uniform density, frequently lobulated, ranging in size from 1 cm to 5 cm in diameter and more commonly in lower lobes.<sup>2,4,6</sup> A radiograph may show connecting vessels radiating from the hilum. Evaluation of a lesion under fluoroscopy during Valsalva and Mueller maneuvers may show pulsations and changes in size, which are highly suggestive of a PAVM.<sup>6</sup>

The contrast-enhanced CT scan is a valuable tool in diagnosis and defining the vascular anatomy of PAVM. It is more sensitive than conventional pulmonary angiography in detecting PAVM (98% vs. 60%). The superiority of CT scanning in detecting PAVM is attributed to the absence of superimposition of the lesion in transaxial views.<sup>7,8</sup> However, pulmonary angiography is better in determining the angioarchitecture when supplemented with hyperselective angiograms. Pulmonary angiography remains the gold standard in the diagnosis of PAVM, and is usually necessary if resection or embolotherapy is being considered.<sup>9</sup>

Magnetic resonance (MR) imaging of PAVM has been studied less than CT. Most lesions within the lung have a relatively long relaxation time and produce medium to high intensity signals.<sup>10</sup> Rapidly flowing blood in PAVMs and aneurysms results in an absent or minimal MR signal a so-called "flow void," thus making them indistinguishable from the adjacent air-filled lung on spin-echo MR imaging. For these reasons, spin-echo MR imaging results in reduced sensitivity and specificity for detection of PAVM.<sup>10,11</sup> However, ultrafast time-resolved 3D MRA is reportedly a better alternative to CT angiography and digital subtraction angiography.<sup>12</sup>

Radionuclide perfusion lung scanning is also a useful adjunct in the diagnosis of PAVM. A positive result at radionuclide scanning is not specific for PAVM, whereas a negative result essentially excludes the diagnosis.<sup>13</sup> Contrast echocardiography is also used to evaluate the cardiac and intrapulmonary shunts, and is able to identify small right-to-left shunts.<sup>14</sup>



Figure 2a, 2b. Contrast-enhanced chest CT scans demonstrating a lobulated enhancing subpleural mass with feeding artery and draining vein.

The treatment of choice in symptomatic patients includes embolotherapy with coils and/or detachable balloons. Alternatively, all patients with feeding vessels more than 3 mm should be embolized due to the attendant risk of paradoxical embolization. Ninety-six percent of PAVMs are undetectable or reduced in size at follow up obtained one or more year after embolotherapy.<sup>7,8,15</sup> No angiography was performed in our patient due to the complete asymptomatic nature of the lesion and because no therapeutic intervention was intended.

## References

1. Churton T. Multiple aneurysms of the pulmonary artery. *Br Med J*. 1897;1:1223-1225.
2. Khurshid I, Downie GH. Pulmonary arteriovenous malformation. *Postgrad Med J*. 2002;78(918):191-197.
3. Iqbal M, Rossoff LJ, Steinberg HN, Marzouk A, and Siegel DN. Pulmonary arteriovenous malformations: a clinical review. *Postgrad Med J*. July 1, 2000;76(897):390-394.
4. Gossage JR, Kanj G. Pulmonary Arteriovenous Malformations. A State of the Art Review. *Am J Respir Crit Care Med*. August 1998;158(2):643-661.
5. Dines DE, Arms RA, Bernatz PE, et al. Pulmonary AV Fistula. *Mayo Clin Proc*. 1974;49:460-5
6. Sluiter-Eringa H, Orié NGM, Slutier HJ. Pulmonary arteriovenous Fistula: diagnosis and prognosis in non-complaint patient. *Am Rev Respir Dis*. 1969;100:177-184.
7. Remy J, Remy-Jardin M, Giraud F, Watinne L. Angioarchitecture of Pulmonary arteriovenous malformations: Clinical utility of 3-Dimensional CT. *Rad*. 1994;191:657-664
8. Stienberg I, Finby N. Roentgen manifestation of pulmonary Arteriovenous Fistula: Diagnosis and treatment of 4 new cases. *A J Roentgenology*. 1957;78:234-246
9. White RI, Mitchell SE, Barth KH, Kaufman S, Kadir S, Chang R, Terry PB. Angioarchitecture of pulmonary arteriovenous malformations: an important consideration before embolotherapy. *AJR*. 1983;140:681-686
10. Silverman JM, Julien PJ, Hefkens RJ, Pelc NJ. Magnetic Resonance Imaging evaluation of Pulmonary vascular malformations. *Chest*. 1994;106:1333-1338
11. Brown JJ, Gilbert T, Gamsu G, Golden JA, Higgin CB. MR imaging of low signal intensity lesions using flow sensitive technique. *J Comp Assist Tomog*. 1988;12:560-564
12. Goyen M, Ruehm SG, Jagenburg A, Barkhausen J, et al. Pulmonary arteriovenous malformation: Characterization with time-resolved 3D MR angiography. *J Magn Reson Imaging*. 2001 Mar;13(3):458-460
13. Whyte MKB, Peters AM, Hughes JMB, Henderson BL, Billigan GJ, Jackson JE, Chulvers ER. Quantification of right to left shunt at rest and during exercise in patient with pulmonary arteriovenous malformation. *Thorax*. 1992;47:790-796
14. Barzilai BA, Wagonner AD, Spessert C, Picus D, Goodenbeger D. Two-dimensional echocardiography in detection and follow-up of congenital arteriovenous malformations. *Am J Cardiol*. 1991;68:1507-1510.
15. Lee DW, White RI, Egglin TK, Pollak JS, Fayad PB, Wirth JA, Rosenblatt MM, Dickey KW, Burdige CM. Embolotherapy of large pulmonary arteriovenous malformations: long-term results. *Ann Thorac Surg*. 1997;64:630-640