Case 12202
Paradoxical embolic posterior infarction in a patient with pulmonary arteriovenous malformation with Osler's disease

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Section: Neuroradiology
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Patient: 65 year(s), male

Clinical History
A patient complaining of right-sided hemianopsia.

Imaging Findings
Brain MRI was performed and showed multiple diffusion restrictions consistent with left posterior infarction (Fig. 1). MRA showed occlusion of the left posterior cerebral artery (Fig. 2). No embolic source was found in the extracranial arterial duplex, transthoracic or transoesophageal echocardiography.
The patient was known to have Osler's disease (hereditary haemorrhagic telangiectasia). A CT of the chest was performed and showed the presence of peripheral pulmonary embolism (Fig. 3). A small, well-circumscribed enhancing nodule, with a single feeding segmental artery and a single draining vein was found at the periphery of the same lobe and another smaller nodule in the left upper lobe, representing simple arteriovenous malformations (Fig. 4, 5).
Anticoagulation was started but with reduced therapeutic dose, due to the risk of bleeding from the malformation. Minimally invasive transcatheter occlusion of the pulmonary arteriovenous malformation (PAVM) is planned.

**Discussion**

Embolic stroke may arise from cardiac sources, vessel supplying the brain (e.g. internal carotid artery), or from other vascular sites in the presence of right-to-left shunt (paradoxical embolism). Shunts may be intracardiac; as in cases of patent foramen ovale, or extracardiac as in pulmonary arteriovenous malformation (PAVM), which was present in our patient with Osler's disease.

Osler's disease or hereditary haemorrhagic telangiectasia (HHT) is an autosomal dominant disorder characterised by forming abnormal vascular structures, whereby arteries connect directly to veins without the presence of intervening capillaries [1]. These abnormal connections can be found in the skin, nose, mucous membranes, lungs, GIT and brain. Its prevalence is about 1/5000 [1, 2].

The clinical diagnostic criteria include: epistaxis, telangiectasia, visceral lesions and family history of first-degree relatives with HHT. According to the advisory board, assembled in Curacao, a definite diagnosis results when there is a presence of at least three out of four criteria; a possible diagnosis in the presence of two criteria, and unlikely diagnosis if fewer than two [3, 4].

PAVMs are present in approximately 15-50% of people with HHT [2]. Complications occur due to low oxygen levels as the blood bypasses the gas-exchanging surface in the pulmonary capillary bed; rupture of the fragile nidus causing pulmonary or pleural haemorrhage, or due to failure of filtering function of the capillaries leading to paradoxical embolism or brain abscess [1, 2, 3]. Due to the seriousness of these complications, which could occur due to an AVM that has been previously silent; screening is advised. The best screening method for PAVMs is transthoracic contrast echocardiography (TTCE) with agitated saline [2]. A positive test, from the detection of bubbles in the left atrium, should be followed by a CT examination, which delineate the exact location, number and size of PAVMs [2].

Administration of anticoagulants in such patients is a challenging decision. The risk of bleeding from AVMs has to be balanced against the risk of recurrent cerebral embolism and should be discussed on a case-by-case basis [5].

Generally any PAVM of more than 3 mm in diameter should be treated, regardless of the presence or absence of symptoms. Angiographic occlusion is the treatment of choice. Follow-up using CT within 6-12 months following embolization and at three-year intervals thereafter is recommended [2].

Teaching point:
PAVM is a source of serious complications, including paradoxical emboli and needs to be screened for in patients with HHT.

**Final Diagnosis**
Paradoxical posterior infarction associated with PAVM and concurrent pulmonary embolism.

**Differential Diagnosis List**

Other causes of embolic infarction (e.g. cardioembolic), Other causes of pulmonary nodules (e.g. granuloma; hamartoma; carcinoma; metastasis), Other causes of PAVM (e.g. traumatic; hepatopulmonary syndrome; schistosomiasis)

**Figures**

**Figure 1 Axial DWI & ADC map**

Axial DWI & ADC map showing multiple diffusion restrictions in the region of the left posterior cerebral artery.

Area of Interest: Neuroradiology brain;
Imaging Technique: MR; MR-Diffusion/Perfusion;
Procedure: Diagnostic procedure;
Special Focus: Embolism / Thrombosis;

**Figure 2 3D phase-contrast intracranial angiography**
3D phase-contrast intracranial angiography showing occlusion of the left posterior cerebral artery.

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Area of Interest: Arteries / Aorta; Vascular;
Imaging Technique: Image manipulation / Reconstruction; MR; MR-Angiography;
Procedure: Diagnostic procedure;
Special Focus: Embolism / Thrombosis;

Figure 3 Magnified oblique axial view

Magnified oblique axial view of a CT of the chest with contrast filling defects due to pulmonary embolism in the segmental arteries of the right lower lobe.

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Area of Interest: Pulmonary vessels; Thorax; Vascular;
Imaging Technique: CT; CT-Angiography; Image manipulation / Reconstruction;
Procedure: Diagnostic procedure;
Coronal MIP of the chest-CT with evidence of an arteriovenous malformation at the right lower lobe and another in the left upper lobe.

Area of Interest: Arteries / Aorta; Pulmonary vessels; Thorax;
Imaging Technique: CT; CT-Angiography; Image manipulation / Reconstruction;
Procedure: Diagnostic procedure;
Special Focus: Arteriovenous malformations;

3D reconstruction of the chest-CT. Pulmonary arteries in blue, veins in red. In the right lower
lobe and the left upper lobe there are simple AVM each with a single feeding artery and draining vein.

Area of Interest: Cardiovascular system; Pulmonary vessels; Thorax;
Imaging Technique: CT; CT-Angiography; Image manipulation / Reconstruction;
Procedure: Computer Applications-3D; Diagnostic procedure;
Special Focus: Arteriovenous malformations;

References


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