Transoesophageal echocardiographic diagnosis of pulmonary arteriovenous malformation in a patient with ischaemic stroke

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Received 30 May 2008; accepted after revision 1 August 2008; online publish-ahead-of-print 23 August 2008

We report a case of an ischaemic stroke secondary to an isolated pulmonary arteriovenous malformation. In this case, transoesophageal echocardiogram played a pivotal role in shunt identification, exclusion of atrial septal defect, and definitive diagnosis and localization of the pulmonary arteriovenous malformations. Pulmonary arteriovenous malformations should be excluded in all patients with cryptogenic stroke.

KEYWORDS
Cerebral embolism; Pulmonary arteriovenous malformation; Stroke; Cerebrovascular accident; Transoesophageal echocardiography

Case report

A 48-year-old Taiwanese woman who had resided in the USA for the past 9 years, presented to the Emergency Department with symptoms of aphasia and right-sided facial droop. Her past medical history was significant for two episodes of transient ischaemic attack, 12 and 16 years prior to presentation. On initial evaluation, the patient exhibited receptive and expressive aphasia, right lower facial droop, and right upper extremity weakness. Mild dysmetria was present on right finger-nose testing. Right plantar response was mute.

Computerized axial tomographic (CT) examination of the head without contrast showed a small hyperdense focus in the left sylvian fissure along the distribution of the posterior left middle cerebral artery (MCA) division. Computerized tomography angiogram of the head revealed a penumbra involving the left posterior frontal and parietal lobes along the left posterior MCA distribution compatible with thrombus in the posterior division of the left MCA. Cerebral arteriography confirmed these findings. Intra-arterial tissue plasminogen activator was infused with resolution of symptoms. A complete hypercoagulable workup (including activated protein C resistance, antithrombin, beta-2-glycoprotein IgG and IgM, anti-phospholipid antibody, protein C and protein S activity, and prothrombin gene mutation) was negative. Venous duplex ultrasound of the lower extremities was negative for deep venous thrombosis.

A transoesophageal echocardiogram (TEE) revealed no structural heart disease with the exception of a markedly positive agitated contrast saline study (see Supplementary material online, Movie 1 and Figure 1). Closer inspection revealed no evidence for atrial septal defect or patent foramen ovale. An additional agitated saline study revealed a large quantity of microbubbles entering the left atrium via the confluence of the left pulmonary veins (see Supplementary material online, Movie 2 and Figure 2). Computerized tomography pulmonary angiography revealed a pulmonary arteriovenous malformations (PAVM) within the anterior basal segment of the left lower lobe (Figures 3 and 4). Pulmonary arteriography of the left main pulmonary artery demonstrated a 4 mm feeding vessel. Amplatzer coils were percutaneously inserted with subsequent obliteration of the AV malformation. The patient was discharged in stable condition on hospital day 6 with complete resolution of her neurological deficits.

Discussion

Pulmonary arteriovenous malformations are abnormal communications between pulmonary arteries and pulmonary veins that were first described by Churton in 1897.¹ The incidence of PAVMs is 2–3 per 100 000 population.² Pulmonary arteriovenous malformations can be single or multiple. The incidence of solitary PAVMs ranges from 42 to 72%². Most solitary PAVMs are present in the lower lobes, with the left lower lobe the most common location.² Pulmonary
arteriovenous malformations may be congenital or acquired. More than 80% are congenital of which 47–80% are associated with Rendu–Osler–Weber syndrome also known as Hereditary Hemorrhagic Telangiectasia (HHT).\(^2,3\) It is estimated that overall 5–15% of the population with HHT has a PAVM.\(^2\)

All PAVMs have an afferent supply, usually from one or more branches of the pulmonary artery. The efferent limb drains into one or more branches of the pulmonary vein or even directly into the left atrium or inferior vena cava.\(^4\) Pulmonary arteriovenous malformations are usually found in close proximity to the visceral pleura or embedded in the outer third of lung parenchyma. In a study of 110 patients\(^4\) with a single PAVM, 89 (81%) of the lesions were either sub-pleural or partially embedded in the lung parenchyma. Pulmonary arteriovenous malformations result in a right to left shunt; if the shunt fraction is \(>20\%\) of
the systemic cardiac output, the patient may have obvious cyanosis, clubbing, and polycythemia. Asymptomatic patients are common and account for 13–55% of patients in various case series.

Neurological involvement in HHT has been reported in ~4–12% of patients. However, the incidence of neurological manifestations in isolated PAVMs is unclear. Roman et al. reported that the most common neurological changes in HHT resulted from the presence of a PAVM. Several mechanisms for stroke have been postulated which include cerebral thrombosis resulting from polycythemia secondary to hypoxaemia, air embolism through a defect in the wall of the PAVM, and thromboemboli arising within the fistula.

In this case, TEE played a pivotal role in shunt identification, exclusion of atrial septal defect, and definitive diagnosis and localization of a PAVM. Pulmonary arteriovenous malformations should be excluded in all patients with cryptogenic stroke. Although both transthoracic echocardiography and transcranial Doppler may be useful adjunctive techniques in determining the presence of a shunt, neither will definitively differentiate between intracardiac and intrapulmonary aetiologies. Transoesophageal echocardiography should be performed routinely in patients with cryptogenic stroke.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

References