

Cerebral and pulmonary arteriovenous malformations

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A 1-year 5-month-old boy presented to the casualty department at Kalafong Hospital with diarrhoea, coughing, fever and weight loss. On examination he was found to be anaemic, peripherally cyanotic and had clubbing of his fingers. On auscultation there were crepitations in the right axilla and a pansystolic murmur over the entire chest. He had a pulse rate of 170/min, a respiratory rate of 44/min and the oxygen saturation at room air was 60 - 70% and on 100% oxygen, 60 - 80%. On the chest radiograph (CXR) a right middle and lower lobe consolidation was noted (Fig. 1). The diagnosis of multi-lobar pneumonia was made, and intravenous antibiotics were commenced. Follow-up radiographs were done which showed no change within a period of 1 week. On echocardiography a mild tricuspid incompetence was found, which did not explain the loud pansystolic murmur. On re-examination a separate murmur was heard over the anterior fontanelle.

Computed tomography (CT) examination of the brain with intravenous contrast revealed multiple arteriovenous malformations (AVMs), predominantly involving the left cerebral hemisphere and thalamic region (Fig. 2). On magnetic resonance imaging (MRI) multiple tightly packed masses of flow voids were found (Figs 3 and 4). CT examination of the chest was also done which revealed a large right pulmonary AVM (Fig. 5).



Fig. 1. Frontal radiograph demonstrating right lower and middle lobe consolidation.

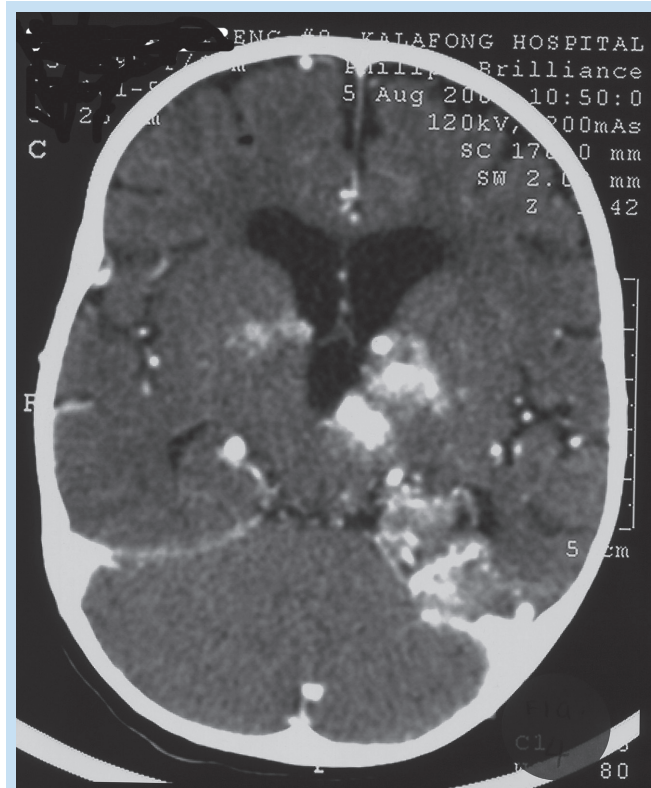


Fig. 2. Contrast-enhanced axial CT images demonstrating hyperdense serpiginous enlarged feeding and draining vessels with a strongly enhancing nidus. Multiple AVMs are demonstrated in the thalamic region.

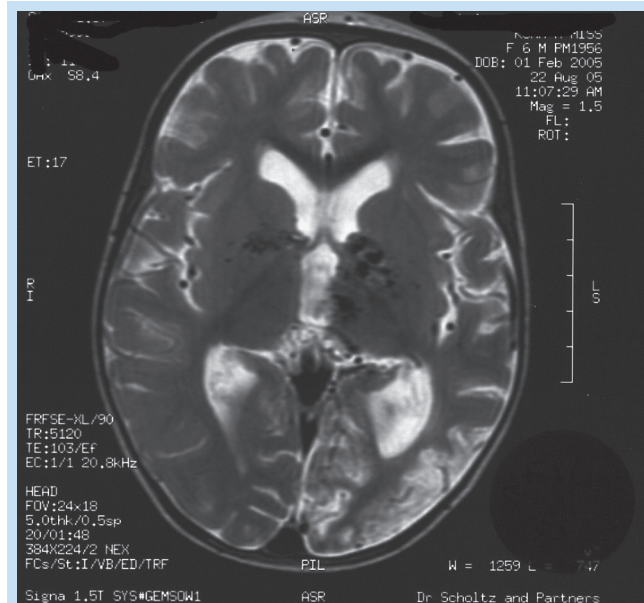


Fig. 3. T2-weighted MRI image demonstrating multiple flow voids with enlarged feeding and draining vessels. There is minimal mass effect as AVMs replace cerebral tissue and do not displace it.¹

CASE REPORT

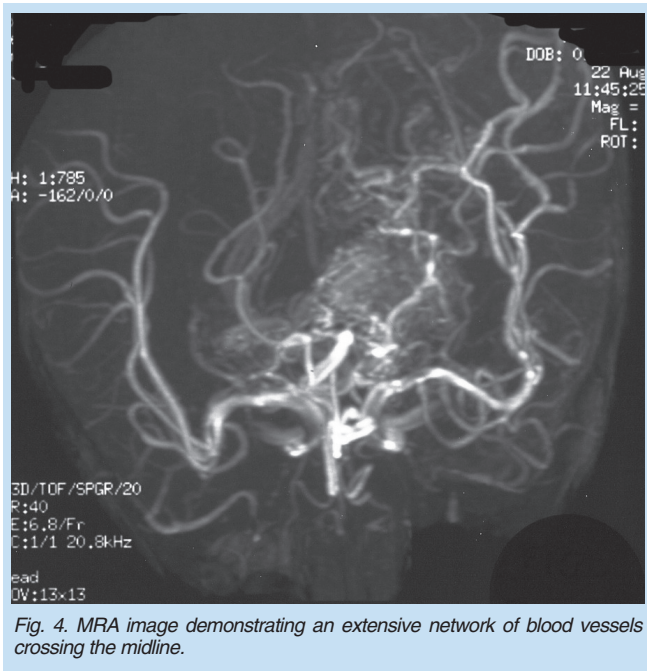


Fig. 4. MRA image demonstrating an extensive network of blood vessels crossing the midline.

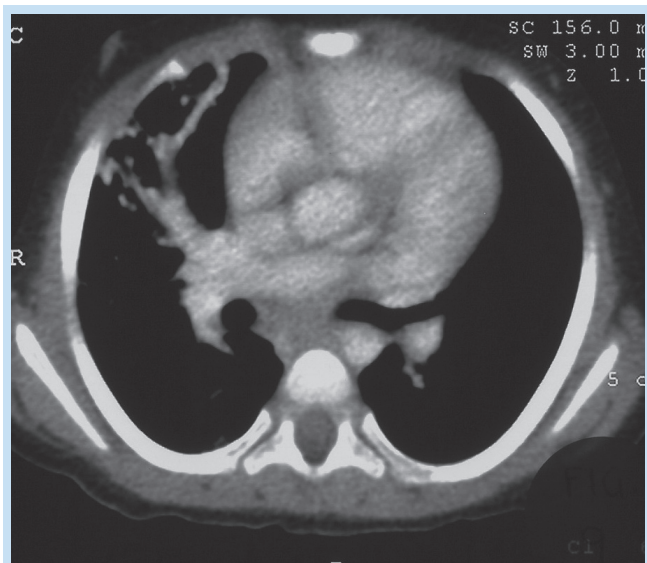


Fig. 5. Contrast-enhanced axial CT image of the chest demonstrating a large right pulmonary AVM.

Discussion

An AVM is a congenital abnormality consisting of a nidus of abnormal dilated tortuous arteries and veins resulting in shunting of blood from an arterial to a venous vessel without the intermediary capillary bed.²

Patients with cerebral AVMs present with headaches, seizures, mental deterioration, progressive hemispheric neurological deficits and ictus from acute intracranial haemorrhages. Eighty per cent of lesions occur by the end of the fourth decade; 20% occur in patients younger than 20 years.²

Congenital pulmonary AVMs are thought to occur secondary to failure of development of the pulmonary capillary network, with persistence of the primitive arteriovenous communications. The AVM is usually fed by a single artery and drained by a single vein, although multiple vessels have been reported. The association of a partial anomalous pulmonary venous return and a hypogenetic lung is perhaps more well-recognised as the scimitar syndrome (congenital pulmonary venolobar syndrome). In these cases the scimitar vein most commonly empties into the infradiaphragmatic inferior vena cava.

Pulmonary AVMs may be asymptomatic or present with haemoptysis, clubbing, cyanosis or polycythaemia. Although pulmonary AVMs are congenital, only about 5 - 10% present in childhood. Nearly two-thirds of pulmonary AVMs occur in patients with Rendu-Osler-Weber syndrome (hereditary telangiectasia). This autosomal-dominant condition is characterised by telangiectatic lesions in the mucous membranes, the skin, and in about 20% of patients, the lungs.³

The current classification of central nervous system vascular anomalies is pathoanatomically based. This assumes that vascular malformations are congenital or developmental hamartomas, rather than neoplasms. This classification is based on microscopic and gross pathological features of the lesions. Although most CNS vascular malformations occur in isolation, some are also found in association with cutaneous and mesodermal vascular anomalies; examples include the Wyburn-Mason syndrome (cerebral, retinal and maxillofacial AVMs) and the alternative name of 'unilateral retinocephalic vascular malformation'. Due to the metameric nature of this disorder there has been a revised classification of such craniofacial vascular malformation syndromes as the cerebrofacial arteriovenous metameric syndromes (CAMS). The Sturge-Weber syndrome involves capillary-venous malformations of the brain due to a lack of a cortical draining vein. It usually affects a single cerebral hemisphere, and is associated with retinal vascular malformations and cutaneous facial capillary malformations.⁴

Conclusion

Since this patient displayed no other ancillary findings, these features were thought to be due to isolated cerebral and pulmonary vascular malformations.

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3. Kuhn JP. *Caffey's Paediatric Diagnostic Imaging*. 10th ed. Philadelphia: Mosby, 2004: 1077.
4. Duncan IC. Vascular malformations part 2 - current classification of vascular malformations. *South African Journal of Radiology* 2004; 8: 23 - 30.