Multiple cerebral arteriovenous malformations associated with soft-tissue vascular malformations

Case report


Department of Neurosurgery, Royal Adelaide Hospital, and Division of Tissue Pathology, Institute of Medical and Veterinary Science, Adelaide, South Australia, Australia

A patient found unconscious, probably due to a seizure, was discovered to have two intracranial arteriovenous malformations. Multiple arteriovenous malformations is a rare condition, and both lesions were excised successfully.

KEY WORDS • multiple cerebral arteriovenous malformations • multiple lesions

INcidents of multiple capillary telangiectasis and cavernous angioma are found frequently; however, multiple cerebral arteriovenous malformations (AVM's) are extremely rare. The association of central nervous system hemangioblastomas with visceral angioma is well recognized. We are reporting a case of multiple cerebral AVM's associated with multiple soft-tissue vascular malformations.

Case Report

This 31-year-old woman was found unconscious. On admission to the hospital she was restless and confused. The day after admission she recovered consciousness.

Examination. No neurological deficits were found. No bruit was heard in the neck or the head. Computerized tomography (CT) showed two high-density shadows in the right frontal and left parietal regions (Fig. 1). General examination showed three bluish, pulsatile swellings (10 × 15 × 10 mm) in the left hand, which were thought to be subcutaneous vascular malformations. Extensive investigation for bleeding and clotting disorders revealed no abnormality.

Two weeks later, repeat CT revealed no change in the appearance of the lesion other than operative effect (Fig. 2). Bilateral carotid angiography showed two avascular masses.

Operations. Eight weeks after admission, the right frontal lesion was excised. A well demarcated hemorrhagic lesion measuring 40 × 40 mm was found, with a tiny arterial supply. Two months later, a similar hemorrhagic encapsulated lesion (25 × 20 mm) in the left parietal region was excised.

Neuropathology. The surgical specimen from the frontal lobe lesion consisted of blood clot (40 × 40 × 15 mm) surrounded by a membrane and small fragments of nearby hemorrhagic gray-white brain tissue. Microscopic examination of the latter showed a vascular malformation abutting on the leptomeninges and consisting of an admixture of arteries, veins, and malformed, thick- and thin-walled, hyalinized vessels of indeterminate type (Fig. 3 left). The presence of elastic tissue was confirmed by the Miller-van Gieson stain (Fig. 3 right). Sections of the encapsulated blood clot showed this to be completely surrounded by an endothelial-lined fibrous wall of varying thickness. Some segments showed hyaline sclerosis and others smooth muscle with focal leiomyomatous proliferation. Only minimal microscopic calcification was demonstrated, and thromboses in various stages of organization were present. The surrounding and intervening brain tissue showed severe fibrillary gliosis, fer-
Multiple cerebral AVM's

**Fig. 1.** Computerized tomography scans showing two high-density lesions in the right frontal and left parietal regions.

**Fig. 2.** Computerized tomography scans 2 weeks after the first operation showing no change in the appearance of the lesion other than the effect of the operation.

**Fig. 3.** Photomicrographs of the frontal arteriovenous malformation. *Left:* Low-power view showing an artery and vascular channels of indeterminate nature. There are thrombi in various stages of organization and a fragment of white matter showing gliosis and hemosiderin deposition. H & E, × 25. *Right:* Same field as left showing extensive presence of elastic lamellae. Miller-van Gieson, × 100.
A. Hanieh, P. C. Blumbergs and P. G. Carney

Our patient presented with an episode of loss of consciousness, the cause of which is uncertain but most probably due to epilepsy. Evaluation led to the discovery of two high-density lesions in the right frontal and left parietal regions which were interpreted as areas of intracerebral hemorrhage. There was no change in the CT appearances over the next 2 weeks of observation, and it was only on histological examination that their true nature as AVM's was discovered. The presence of a high-density area without mass effect on plain CT scan in patients with an AVM who present with a seizure disorder has been noted previously, and in some cases is due to calcification. We believe that this CT appearance in our patient, in the absence of significant calcification, was due to the pooling of blood in the massive segmental saccular dilatation of the component vessels of the AVM which was demonstrated on pathological examination. Thus, this CT finding led to the erroneous clinical diagnosis of bilateral intracerebral hemorrhages.

**Postoperative Course.** The postoperative course was uneventful, and the patient showed no change in neurological state. The hand lesions were not treated.

**Discussion**

Reports in the literature describe AVM's as solitary lesions. Our case is unusual in that the AVM's were multiple and associated with soft-tissue vascular malformations, a rare association. In the last 10 years, only two multiple cases have been reported in the English literature. Hash, et al., reported a concurrent intracranial and spinal cord AVM, and Tamaki, et al., described a case of multiple AVM's involving scalp, dura, retina, cerebrum, and posterior fossa.

**Acknowledgment**

We are grateful to Dr L. V. Perrett for reporting on the radiological investigations.

**References**


Address reprint requests to: Paul G. Carney, Esq., F.R.A.C.S., Neurosurgical Unit, Royal Adelaide Hospital, North Adelaide, South Australia 5000, Australia.

J. Neurosurg. / Volume 54 / May, 1981