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Multiple intracranial arteriovenous malformations: a case report

K. Ericson[1], M. Söderman[1], B. Karlsson[2], W. Y. Guo[1], C. Lindquist[2]

[1] Department of Neuroradiology, Karolinska Hospital, S-104 01 Stockholm, Sweden
[2] Department of Neurosurgery, Karolinska Hospital, S-104 01 Stockholm, Sweden

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Abstract. Multiple intracranial arteriovenous malformations are rare. There are a few cases in the literature with up to three malformations in one patient. A child with seven separate cerebral malformations is now described. There was no history of haemorrhage, but only of febrile seizures. The malformations were discovered at CT and verified at angiography. Six were selected for stereotaxic irradiation with the multicobalt unit. The seventh was considered too big for irradiation and suitable for surgery.

Key words: AVM – Stereotaxic irradiation – Angiography

Intracranial arteriovenous malformation (AVM) is a congenital lesion that is usually single. Although true AVMs are often supplied from different arterial territories, each feeding artery supplying its own part of the malformation, the malformation usually forms a continuous nidus. AVMs are rare, with a reported incidence of 0.1–0.2 % [1]; multiple intracranial AVMs are extremely rare, their incidence, estimated by Stone et al. [2] being 0.002 %.

Cavernous haemangiomas are often multiple. They are now found more commonly, because of increased use of magnetic resonance imaging [3, 4]. Patients with different kinds of vascular malformation have been described [5, 6]; cavernous haemangiomas are sometimes seen in combination with AVMs or venous anomalies [5].

Case report

A girl aged 7 years had fits during periods of fever. There were no symptoms or signs, in particular none suggesting generalised vascular disorder. Such disease could not be found in the family either.

Contrast-enhanced CT (8 mm slice thickness) disclosed two AVMs, but angiography performed revealed six separate AVMs. A second contrast-enhanced CT (4 mm slice thickness) 18 months later showed five AVMs, three of which then showed evidence of previous haemorrhage. The patient was subsequently referred for treatment by stereotaxic irradiation. It was noted that one AVM in the right temporal lobe was rather large, but could probably be treated with an acceptable risk of undue radiation effects. The other AVMs were small. The quality of the images did not, however, permit more detailed analysis of the actual size and shape of the niduses.

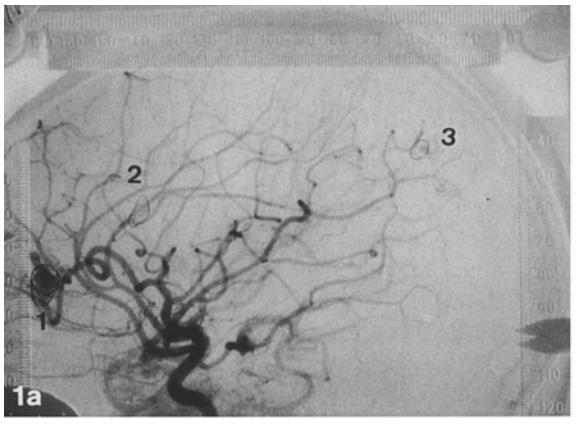
Angiography with a stereotaxic frame prior to treatment showed seven separate intracranial AVMs (Figs. 1, 2) in the cerebral hemispheres. The one in the right temporal lobe was large, its diameter exceeding 3 cm. It was superficially located, and was judged unsuitable for irradiation but suitable for open surgery. The remaining six malformations were considerably smaller and ideal targets for stereotaxic irradiation.

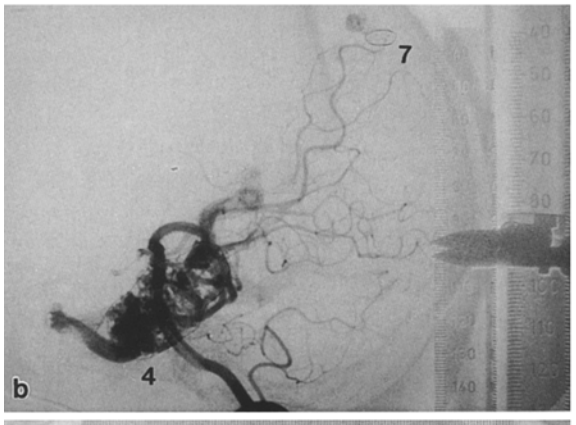
Discussion

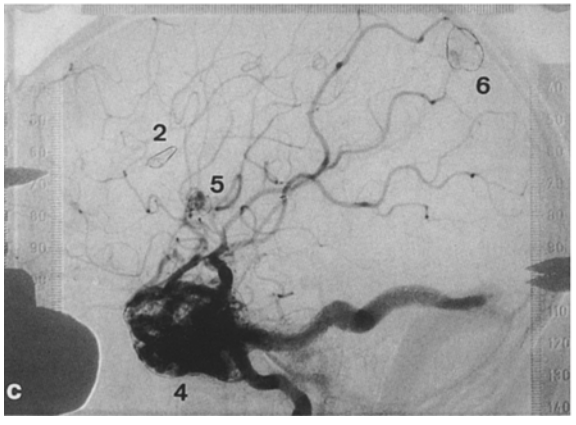
Karolinska Hospital has more than 20 years experience with irradiation of AVMs by the multicobalt unit (gamma knife radiosurgery) [7–12]. From over 3000 AVMs reviewed at our institution more than 1000 have been treated with gamma knife surgery. Among these 3000 cases were ten with more than one intracranial nidus. Seven patients with two or more separate nidi have been treated by stereotaxic irradiation, including the present patient. Two patients are lost to angiographic follow-up, one has recently been treated and one is awaiting treatment. Of two patients treated more than two years ago the lesions have been totally obliterated in one and partially in the other. The remaining three patients, all of whom had two nidi, were not accepted for treatment.

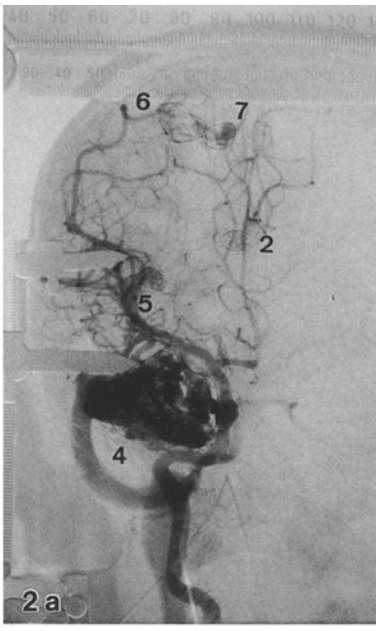
The combination of multiple intracranial, spinal and pulmonary AVMs is recognised [13–15], as is an association with hereditary haemorrhagic telangiectasia (Rendu-Osler-Weber disease). It was stated by Maryama et al. [15] that in this disease pulmonary arteriovenous fistulae are more frequent than other vascular lesions in liver or brain. Multiple cerebral AVMs and soft-tissue vascular malformations, possibly representing Rendu-Osler-Weber disease, were also described by Hanieh et al. [16]. Occasional patients with multiple AVMs have been reported by others [17–22]. Voigt et al. [23] described multiple AVMs in a patient who also had multiple arterial aneurysms and general arterial ectasia.

Correspondence to: K. Ericson









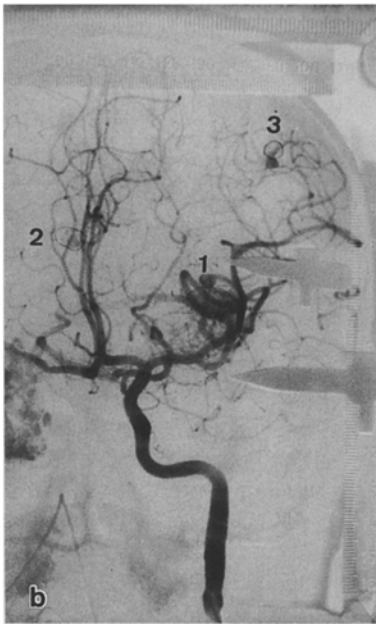


Fig. 1. **a** Left carotid angiogram (lateral view) showing three small arteriovenous malformations (outlined).
b Right vertebral angiogram showing large temporal and small parieto-occipital malformations.
c Right carotid angiogram showing four AVMs, one of which is partly shown in **b**

Fig. 2. **a** Right and **b** left carotid angiogram (AP-view) showing the malformations

The incidence of multiple AVMs is extremely low. Cerebral angiography carries a risk, albeit low, of serious complications. It is therefore not mandatory to perform complete cerebral angiography as a screening procedure to rule out multiple lesions in patients with a known AVM but without any clinical or radiological evidence of additional lesions. However, all possible feeders, including the external cerebral arteries, should of course be injected for complete angiographic mapping of an AVM, so that four vessel cerebral angiography may be necessary. MRI and MR angiography (MRA) are becoming more and more important in the complete mapping of an AVM, and MRA will probably replace conventional angiography in many of these patients.

Due to the extremely low incidence of multiple AVMs no material large enough for statistical analysis exists. We have, however, no reason to suppose that multiple AVMs behave differently from a single AVM when irradiated. A large series of single AVMs irradiated at Karolinska Hospital [10] showed complete obliteration in 87% at 3 year follow up. Therefore, the changes of complete obliteration of all six AVMs irradiated in this patient within a three year period could be estimated at approximately 43%. Undue radiation effects are a function of the size of the irradiated volume, the dose delivered and the patient's individual sensitivity to radiation. The very steep dose gradient of the gamma knife makes the cumulative dose to the brain very small [24, 25], and the risk of undue radiation effects should, for each AVM, be unaltered compared to the risk incurred when irradiating single AVMs.

The risk of haemorrhage from intracranial arteriovenous malformations has been estimated to 2–4 per cent per year per lesion [1, 26, 27]. Since the second CT showed evidence of previous bleeding the patient must be considered

lucky not to have suffered significant intracranial haemorrhage prior to treatment. Stereotaxic irradiation usually causes obliteration of the AVM after 2–3 years [10], and the bleeding risk is believed to be substantially unchanged until complete obliteration is achieved. The patient is therefore still at risk. Several of the AVMs could be operated upon or treated by interventional neuroradiology. The risk would then be highly dependent on individual skills but the risk of bleeding would be eliminated immediately by total extirpation.

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