Letters

What is a “cryptic” arteriovenous malformation?

Sir: Small arteriovenous malformations (AVMs) as a cause of sudden intracerebral haematoma was first recognised by Margolis et al. Crawford and Russell applied the term “cryptic” to these AVMs, defining them as lesions which are clinically silent, measuring less than 2 cm to 3 cm in diameter, and therefore difficult to find at necropsy. This pathological definition has been adapted for clinical usage and is used interchangeably with “microan giomas” and “small AVMs.” However, more recent reports on “cryptic” AVMs deal with AVMs much smaller than originally defined, that is, less than 1 cm in diameter and often a few mm in size. Terao et al further define “cryptic” AVMs as those “not detectable by routine angiography”, and also state that there is no proper definition for this entity. Thus there is no uniformity in the usage of the term “cryptic”. A patient with a “cryptic” AVM recently managed by us prompted us to consider the continued usage of this term.

A 29 year old female presented with a history of sudden onset of headache and vomiting 3 days prior to admission. On examination the patient was awake and alert but complained of severe headache. She had no deficits other than a right homonymous hemianopia. There was no neck rigidity and Kernig’s sign was negative. CT showed a large haematoma in the left parietal lobe with evidence of mass effect. No contrast enhancing lesion was seen. Left carotid angiogram showed a retrost Sylvian mass lesion and in addition, a small AVM in the posterior parietal region fed by one of the terminal branches of the left middle cerebral artery and there was a single draining vein (fig).

The patient underwent emergency left parietal craniotomy with complete evacuation of the haematoma. A small AVM, 5 mm in diameter, was seen on the posterior wall of the haematoma cavity close to the surface. After coagulating and cutting the feeding artery and draining vein, the AVM was excised. The patient made an uneventful recovery and at the time of discharge had no deficits other than a persistent hemianopia.

Most reports of “cryptic” AVMs include AVMs less than 2–3 cm in size. Non-visualisation at angiography is not a criterion for their inclusion in this group. However, there are others who believe that these lesions should be angiographically occult. This confusion has arisen because a term which was initially used to define a pathological entity is being used for clinical description. The AVM in our patient would qualify as a “cryptic” AVM if only the criteria of size is applied to it. But it was visible on the angiogram and is therefore not angiographically occult. If size be the only criteria, then many of the “cryptic” AVMs would be visualised with present day angiographic techniques and so “cryptic” (meaning “hidden” or “ocult”) would be inappropriate. Moreover the size of AVMs currently being called “cryptic” is less than that originally defined.

Both the criteria of size and being angiographically occult can be used to define these lesions. But many of the angiographically occult vascular lesions are visible on a CT scan and so they are not radiologically occult. Also these angiographically occult vascular lesions consist of all types of vascular malformations including cavernous angiomas.

We feel that “cryptic” AVM is a term which should no longer be used in clinical reports as its usage lacks uniformity. Further, with better angiographic techniques, and imaging like the high resolution CT scan and magnetic resonance, no AVM will be really “hidden” unless it has been destroyed by the haemorrhage or compressed by the hemoma.

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References

Transmissible agent in the amyotrophic form of Creutzfeldt-Jakob disease

Sir: We wish to record further data on a patient with the amyotrophic form of Creutzfeldt-Jakob disease which we described in 1971. A squirrel monkey was inoculated with brain from the patient on 17 May 1971 at the National Institutes of Health, Bethesda, Maryland, USA. The transmission of the Creutzfeldt-Jakob agent was reported as negative in 1983 but the inoculated monkey died on 10 August 1984 and the histopathological changes found in the brain were those of Creutzfeldt-Jakob disease (Rogers-Johnston, personal communication 1986). The conclusion drawn from negative transmission experiments using central nervous system tissue from this and other patients was that most cases of dementia associated with early amyotrophy are more closely related to classic amyotrophic lateral sclerosis than to transmissible Creutzfeldt-Jakob disease and do not deserve the label “amyotrophic Creutzfeldt-Jakob disease”. Indeed, the existence of an amyotrophic form of Creutzfeldt-Jakob disease is regarded by some authors as unproven.