What is a "cryptic" arteriovenous malformation?

Sir: Small arteriovenous malformations (AVMs) as a cause of spontaneous intracerebral haematomas was first recognised by Margolis *et al.*[1] Crawford and Russell[2] 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 "microangiomas"[3] and "small AVMs".[4] However, more recent reports[5,6] 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*[5] 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 retrosylvian 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.

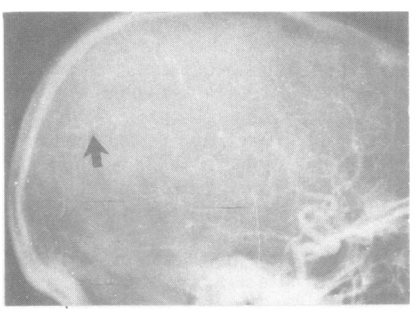

Fig *Lateral view of left carotid angiogram showing a small AVM (arrow) with feeding artery and draining vein.*

Most reports[4,7,8] 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[5] 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 "occult") 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[5] and so they are not radiologically occult. Also these angiographically occult vascular lesions consist of all types of vascular malformations including cavernous angiomas.[9]

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 hematoma.

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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.[1] 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[2] but the inoculated monkey died on 10 August 1984 and the histopathological changes found in the brain were those of Creutzfeldt-Jakob disease (Rodgers-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".[2] Indeed, the existence of an amyotrophic form of Creutzfeldt-Jakob disease is regarded by some authors as unproven.[3]

The neuropathological abnormalities in the spinal cord in this case have been agreed by all observers, but Salazar *et al*[2] considered that non-specific, age-related, spongiform change can be confused with specific Creutzfeldt-Jakob changes and assigned this patient to such a category in their 1983 publication. In our opinion, the cortical spongiform change observed in this case is specific though of mild degree. The pathogenesis of the neuropathology in the amyotrophic form of Creutzfeldt-Jakob disease is probably similar to that observed in scrapie in which the infection can be specifically anatomically targeted, thus determining clinical and pathological abnormalities.[4] This anatomical variation may be the result of differing routes of entry of the agent to the CNS.

Various infectious agents have been postulated in Creutzfeldt-Jakob disease. Intracytoplasmic spiroplasma-like bodies have been described[5-7] in the axoplasm, primarily in presynaptic terminals in this disease. These loosely spiral structures measured up to 1000 nm in length and varied from 40–137·5 nm in width. Although we described[1] a number of unidentifiable structures including one which was tightly coiled, intranuclear, and which measured 570 nm in length by 45 nm in width, no spiroplasma-like shapes were seen. Also, Leach *et al*[8] were neither able to cultivate spiroplasmas from brains of 18 cases nor to detect antibody to seven strains of spiroplasma in sera from 15 patients. From all the available evidence it is unlikely that spiroplasmas are involved.

Since this patient's brain contained a transmissible agent which produced Creutzfeldt-Jakob disease in an inoculated squirrel monkey the combination of dementia and motor neuron disease should now be included in the transmissible Creutzfeldt-Jakob group.

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Horner's syndrome due to superior-mediastinal schwannoma

Sir: A benign schwannoma of the superior-mediastinal sympathetic chain was discovered in a 21 year old man seven years after the onset of Horner's syndrome and unilateral headaches. The patient was initially misdiagnosed as having Raeder's syndrome.

At the age of 14 he presented with a 2 year history of a left sided ptosis, anhydrosis, miosis (Horner's syndrome) and unilateral headaches. The headaches were localised over the left eye, occurred two to three times a week, were of variable severity, lasted several hours, were occasionally throbbing in nature and associated with blurring of vision. Detailed clinical examination, full blood count, WR and skull radiograph were all normal. In the absence of a demonstrable cause a diagnosis of idiopathic Raeder's syndrome[1] was made (the association of miosis, ptosis and unilateral peri-orbital headache). The fact that anhydrosis is not part of Raeder's syndrome was overlooked at this stage. The patient's headaches and associated symptoms resolved over a one month period on prophylactic migraine therapy (sanomigran 0·5 mg tds) but the Horner's syndrome persisted. After 6 months the medication was stopped without recurrence of headache.

Seven years later he was referred with a 6 month history of dysphagia and a 3 kg loss of weight. On examination he had a mass in the left thoracic inlet with displacement of trachea. The Horner's syndrome was still present on the left although he no longer suffered from headaches. On examination the irises were noted to be markedly different in colour. The miotic pupil failed to dilate with cocaine instillation. Plain radiographs and CT of the thoracic inlet confirmed a mass in the left superior mediastinum displacing the oesophagus, trachea and great vessels to the right.

After an incision biopsy had confirmed the presence of a benign schwannoma, this was excised via a transverse cervical incision dividing the sternocleidomastoid. The tumour was well encapsulated, 5 × 5 × 6 cm in size approximately and was easily enucleated. It appeared to arise from the sympathetic chain at the level of the T2/3 nerve root. After operation the patient's dysphagia resolved over 3 months although the left Horner's syndrome has predictably persisted on 2 year follow-up.

The finding of a schwannoma of the sympathetic chain explained both the original presentation of Horner's syndrome and the subsequent dysphagia. Parapharyngeal and particularly superior mediastinal schwannomas are very unusual. When they do occur in this site they arise from either the vagus or cervical sympathetic nerves.[2]

This patient was originally incorrectly diagnosed as Raeder's syndrome. This syndrome differs from Horner's syndrome in that the miosis and ptosis are associated with retro-orbital headache; in addition, there is no anhydrosis. Facial sweating is retained in Raeder's syndrome because the hypothetical interuption to the sympathetic pathways is distal to the bifurcation of the common carotid artery so that the distribution of the external carotid retains its sympathetic innervation.[3] The fact that this patient had anhydrosis and that the pupil failed to dilate with cocaine instillation[4] indicates that the functional sympathetic lesion was between the superior cervical ganglion and the carotid bifurcation.

Raeder[1] recognised the incomplete form of Horner's syndrome in 1924. Two types of Raeder's syndrome are recognised. *Type I* is associated with para-sellar nerve involvement (that is III, IV, and VI) and is usually due to a demonstrable cause. *Type II* does not involve cranial nerves, is commonly idiopathic but has once been shown to be caused by an aneurysm of the internal carotid artery.[5]

Heterochromia iridis can be congenital