Clinics in Diagnostic Imaging (56)

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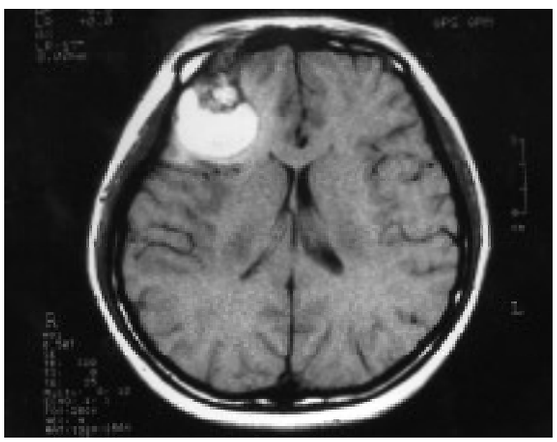

Fig. 1a Axial T1-weighted MR image.

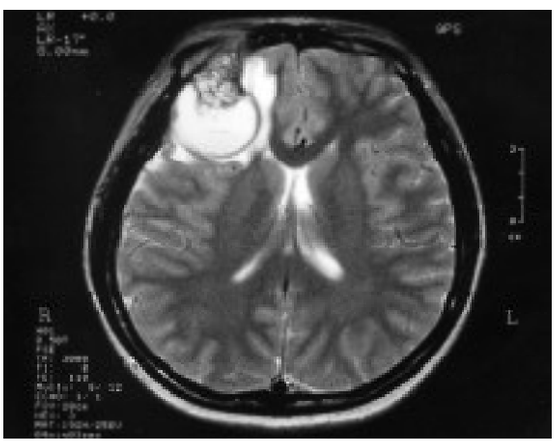

Fig. 1b Axial T2-weighted MR image.

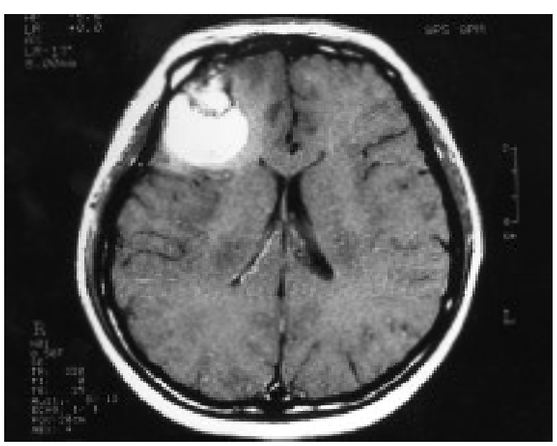

Fig. 1c Axial enhanced T1-weighted MR image.

CASE PRESENTATION

A 28-year-old woman was seen in the emergency department with a complaint of seizure about 1 hour before admission. Clinically, the seizure was of a generalised tonic type with post- ictal amnesia, and the seizure duration was approximately 1 minute. Routine haematological laboratory studies were normal. The blood pressure was 130/80. On physical examination, the patient exhibited a drowsy mental state. There were no meningeal signs nor was there any focal neurological abnormality. Emergency magnetic resonance (MR) imaging was performed (Figs. 1a-c). What do the MR images of the brain show? What is the diagnosis?

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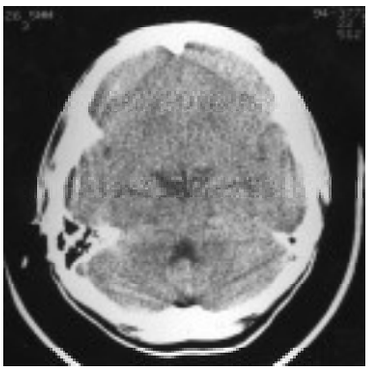

Fig. 2 Previous unenhanced CT scan of same patient as Fig. 1 shows a small area of high signal attenuation (arrows) in the subcortical area of the right frontal lobe.

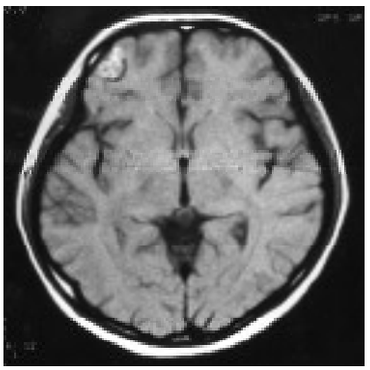

Fig. 3a Previous axial T1-weighted MR image of same patient as Figs. 1-2 shows a subtle right frontal lobe heterogeneous signal intensity focus with surrounding low signal rim.

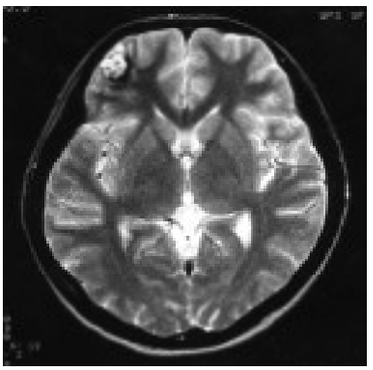

Fig. 3b Previous axial T2-weighted MR image of same patient as Figs. 1-3a shows again the heterogeneous signal intensity lesion with a surrounding low signal rim. No surrounding parenchymal oedema is noted.

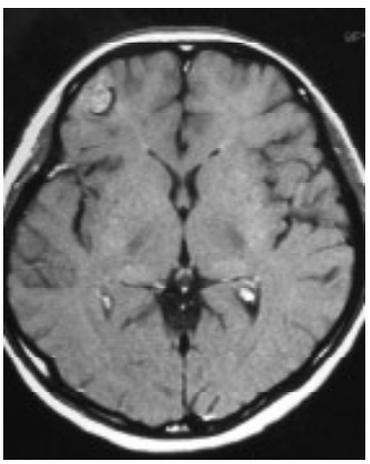

Fig. 3c Previous axial enhanced T1-weighted MR image of same patient as Figs. 1-3b shows no significant enhancement.

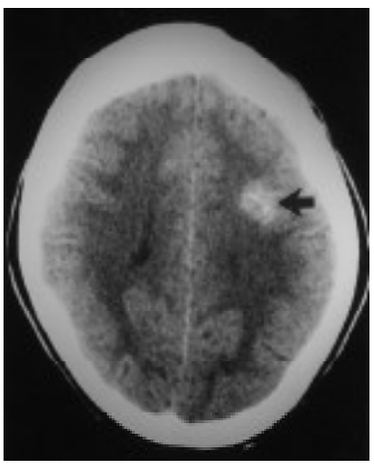

Fig. 4a Unenhanced CT scan of the brain in a 26-year-old woman presenting with a 2 year history of generalised seizures shows a focal hyperdense lesion in the left parietal lobe with focal calcification (arrow) within the lesion. There was no mass effect or surrounding oedema.

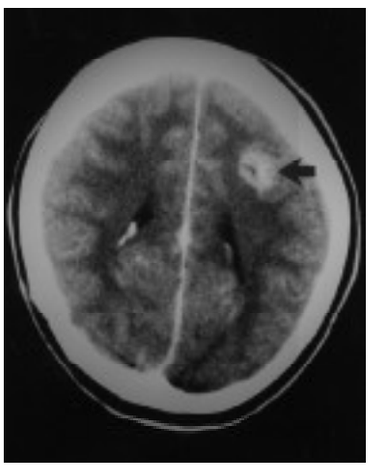

Fig. 4b Enhanced CT scan in the same patient as Fig. 4a shows enhancement of the lesion with a small low density focus of gliosis within the lesion.

IMAGE INTERPRETATION

MR imaging was performed with a 0.5T system. The unenhanced T1-weighted image of the brain (Fig. 1a) shows a 3.6cm size, round, homogeneous high signal intensity lesion with a focal, eccentric heterogeneous focus in the right frontal lobe. The heterogeneous focus shows an irregular low signal intensity rim.

The T2-weighted image (Fig. 1b) shows the larger rounded portion of the lesion to be of homogeneous high signal intensity. The smaller eccentric focus shows mottled, heterogeneous intensity. Surrounding parenchyma oedema is noted.

After IV gadolinium administration (Fig. 1c), the larger rounded portion of the lesion does not enhance, but the eccentric focus shows minimal enhancement centrally.

A review of the previous CT scan (Fig. 2) shows a subtle area of hyperdensity in the right frontal lobe (arrow). A previous MR examination (Figs. 3a-c) shows a heterogeneous, mixed signal right frontal focus with a surrounding low signal rim on both enhanced and unenhanced T1- and T2- weighted images. These findings are compatible with cryptic vascular malformation (CVM) (or cavernous angioma) with recent haemorrhage.

DIAGNOSIS

Cavernous vascular malformation with intracerebral haematoma.

CLINICAL COURSE

The patient subsequently underwent angiography (not shown) which revealed no abnormal vasculature. At surgery, the vascular mass was removed in en-bloc. Crowding of thin- walled dilated vessels, fresh and organised chronic haematomas, and surrounding hemosiderin deposition were noted microscopically. The pathological diagnosis was a partially- thrombosed and haemorrhagia cavernous vascular malformation.

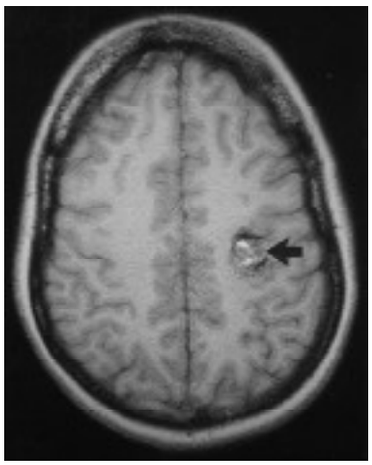

Fig. 5a Patient with CVM. Axial fast spin echo T1- weighted MR image shows high signal intensity methaemoglobin within the lesion and a heterogeneous "mulberry" appearance (arrow).

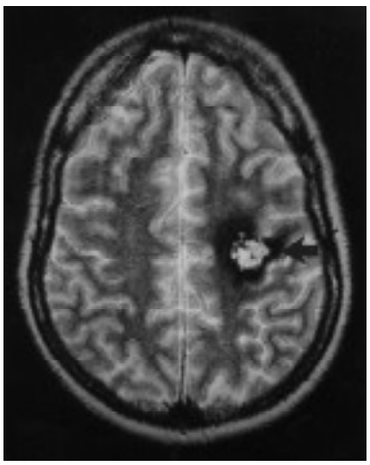

Fig. 5b Same patient as Fig 5a. Axial grqadient echo T2- weighted MR image shows a prominent low signal intensity rim of haemosiderin (arrows), typical of cavernous venous malformation.

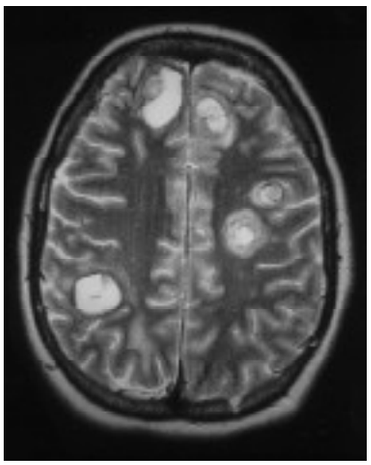

Fig. 6 Axial fast spin echo T2- weighted MR image of a patient with haemorrhagic metastases of the brain due to melanoma shows multiple lesions which have mixed methaemoglobin and haemosiderin blood products. These may mimic multifocal carvenous malformations.

DISCUSSION

Cryptic or occult cerebral vascular malformations are a group of vascular lesions that are not visualised at standard angiography. The term "cryptic" (i.e., hidden) vascular malformation was first used by Crawford and Russel in 1956. They were referring to small, clinically "latent" lesions, some of which were angiographically occult. Subsequently, many authors described other lesions as "occult" or "cryptic" vascular malformations. There was no wide agreement on terminology nor was consensus reached on the precise pathology or pathophysiology of these lesions. However, at present, the terms occult and cryptic are more or less synonymous[1] and generally refer to vascular malformations that have in common angiographic invisibility and a distinct appearance on MR images[2]. These lesions are unlike the fast-flow arteriovenous malformations (AVMs) that are angiographically apparent. Clinically, these lesions cause a wide range of signs and symptoms, including transient neurological deficits, seizures, and headaches. Haemorrhage or thrombosis of a vascular malformation is often not clinically suspected.

Although recent improvements in angiographic techniques have allowed some of these lesions to be depicted[3], angiography usually shows no abnormal vessels. For this reason, both CT and MR imaging are usually required in the detection of these lesions. Before the development of CT, the diagnosis of the CVM was usually made at surgery or at autopsy. But with the advent of CT, it became apparent that CVM was not simply a small AVM[3]. However the diagnosis of the CVMs may be difficult to make in some instances with CT alone. On CT, they are noted to be small isodense or hyperdense masses with little mass effect, often containing punctate calcifications, and exhibiting mild contrast enhancement (Figs. 4a-b). The differential diagnosis on CT includes calcified low-grade cerebral neoplasms (eg. astrocytoma, oligodendroglioma), chronic granulomatous infection (eg. tuberculoma), hamartoma, or nonspecific chronic haemorrhage[3,4].

The advent of MR imaging ushered in a new era in the diagnosis of CVMs[1,5]. They are now easily recognised on MR images and no longer are "cryptic" or "occult" with regard to presurgical diagnostic imaging techniques. The findings on unenhanced MR imaging represent calcifications and/or paramagnetic/ferromagnetic effects[6]. Common typical features of CVMs[5] include a heterogeneous core with multiple foci of hyperintensity on short TR/TE and sidual sinusoids within the lesion, while peripheral rim enhancement indicates leakage of contrast agent into the gliotic capsule. There are few or are no reports of these lesions occurring without associated haemorrhage or thrombosis, unlike the other categories of vascular malformations[1]. CVMs usually have well-defined margins. The supratentorial lesions tend to be subcortical or periventricular[6]. Oedema may be encountered when acute bleeding or thrombosis is present[7].

The primary differential diagnosis based on MR imaging studies is haemorrhagic neoplasm (Fig. 6). The combination of MR findings with the clinical and positive familial histories (familial predilection) provides sufficient conclusive evidence in most cases for the diagnosis of CVM to be made. Consequently, high field MR imaging may obviate the need for diagnostic biopsy of these lesions[6,7], particularly with regard to CVMs in critical anatomic locations[6,7]. In addition, MR imaging

may reveal additional lesions (ie. A tendency towards multiplicity) not seen on CT, and MR imaging may be useful in the sequential follow-up of these lesions over time[7]. Pathologically, CVMs include all varieties of vascular malformations: thrombosed arteriovenous malformations, capillary telangiectasias, venous angiomas, and cavernous angiomas[3,4]. However, the Tomlinson et al[10], in a review of the MR appearance and histopathology of 25 CVMs, concluded that histopathologically, "cavernous lesions were the commonest form of occult vascular malformation". They also emphasised that most of the cavernous angiomas contained thrombosis, a feature atypically seen in AVMs[10]. Many so-called thrombosed AVMs might in fact actually be mixed cavernous angiomas that have associated telangiectasias or arteriovenous shunts, the presence of which has led to the prior classification of these lesions as AVMs. Thus, most, if not all, CVMs are probably actually cavernous angiomas.

The pathogenesis of CVM is still unknown. In some families, cavernous angiomas arise as a result of genetic mutation. Wilson proposed that nonspecific petechial haemorrhage leads to the development of a CVM[2]. Dillon suggested that elevated venous pressure consequent to venous outflow obstruction within the territory of a venous malformation may play a key role in the development of sporadic cavernous angiomas by provoking a cascade of events, including ischaemia and haemorrhage, the release of angiogenic factors, and the subsequent development of a cavernous angioma[1]. Observations supporting this theory include their close association with venous malformations, the development of cavernous angiomas from preexisting venous malformations, and the evidence of increased pressure within venous malformations. This concept explains the pathological diversity that has been reported, as well as the coexistence of multiple (ie. mixed) vascular malformations in a single specimen[1].

The natural course of untreated cavernous angiomas has been reviewed by several authors. Repeated, often occult, haemorrhagic or thrombotic episodes are common[7]. However, the risk of haemorrhage from a CVM is only 0.25% to 0.7% per year[1]. This incidence appears to be more common in cases of infratentorial lesions. Kondziolka et al[8] found that in patients with prior clinical haemorrhage, the annual rate of re-haemorrhage rose to 4.5%. However, MR imaging by itself cannot be used to predict the bleeding potential of CVMs[7]. The bleeding or thrombosis may occur within the lesion itself or in its immediate vicinity. Massive haemorrhage sometimes ruptures into the adjacent parenchyma beyond the peripheral border of gliosis of the CVM. The expanding haematoma may tear adjacent arterioles, thereby inducing increased bleeding. The haematoma may eventually liquefy and harbour xanthochromic fluid[7].

The options for treatment of cavernous angiomas depend in large part on the natural course of the individual lesion, as well as its location and its surgical accessibility[1]. Therapeutic strategies include the conservative observation of patients with asymptomatic or surgically - inaccessible lesions, surgical excision of symptomatic and readily accessible lesions, and radiosurgery for progressively symptomatic but surgically inaccessible lesions[1,2,7].

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ABSTRACT

A 28-year-old woman presented following a seizure. Physical and neurological examinations showed no significant abnormality except a drowsy mental state. Routine laboratory studies were normal. Emergency MR imaging showed a rounded, high signal intensity lesion with a focal eccentric heterogeneous signal focus in the right frontal lobe. The heterogeneous focus revealed an irregular low signal rim on T1- and T2-weighted images. Mild surrounding parenchymal oedema was noted. The eccentric focus showed some central enhancement. The diagnosis of cavernous vascular malformation with intracerebral haematoma was confirmed at surgery. The combination of typical MR imaging findings, with clinical and family histories consistent with cavernous angiomata provides sufficient conclusive evidence.

Keywords: cavernous vascular malformation, cerebral angioma, cerebral vascular, malformation, computed tomography, magnetic resonance imaging