

Coincident Giant Cavernous Angioma and Large Middle Cerebral Artery Aneurysm

Tom D. Newton, M.D., Saravanan Munusamy, M.D., and Roger Laitt, M.D.

Cavernous angiomas although relatively common lesions rarely reach a large size. They have a well documented association with AVMs, capillary telangiectases and venous angiomas but are not specifically associated with intracerebral aneurysms. We present a case of what we believe to be the 4th largest reported giant cavernous angioma to present in adulthood. This cavernous angioma also happened to be associated with a large intracerebral aneurysm, an association not previously reported. The sometimes confusing nomenclature of cavernous angiomas and other similar vascular malformations is also discussed.

Introduction

Cavernous angiomas although relatively common lesions rarely reach a large size. They have a well documented association with AVMs, capillary telangiectases and venous angiomas but are not specifically associated with intracerebral aneurysms. We present a case of what we believe to be the 4th largest reported giant cavernous angioma to present in adulthood.

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Abbreviations: AVM, arteriovenous malformation; CT, computed tomography; MRI, magnetic resonance imaging

Tom D. Newton, M.D. (Email: tomnewton30@doctors.org.uk), Saravanan Munusamy, M.D., and Roger Laitt, M.D., are in the Department of Neuroradiology, Greater Manchester Centre for Neurosciences, Hope Hospital, Salford, Manchester, UK.

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Case Report

A 24-year-old woman presented with a 2-year history of headache, recent temporal lobe seizure and papilloedema. CT demonstrated a "tumour" in the temporoparietal region which biopsy showed to be a possible haemangioma but clinically and radiologically was a malignant lesion, likely a glioma. The patient was regularly seen by a neurologist who treated her conservatively with anticonvulsants and dexamethasone. Repeat CT in 1983 reportedly showed a slight increase in tumour bulk. A CT scan in 1985 showed calcification and the lesion was thought to be an oligodendroglioma. Evaluation of an MRI scan in 1990 led to a diagnosis of a benign haemangioma or teratoma. Repeat MRI scan in 1995 gave a definitive diagnosis of a cavernous angioma and described a "nodule" measuring 10mm arising from its inferior surface. By 1996 her headaches and fits were well controlled so she was discharged from the care of the neurologist.

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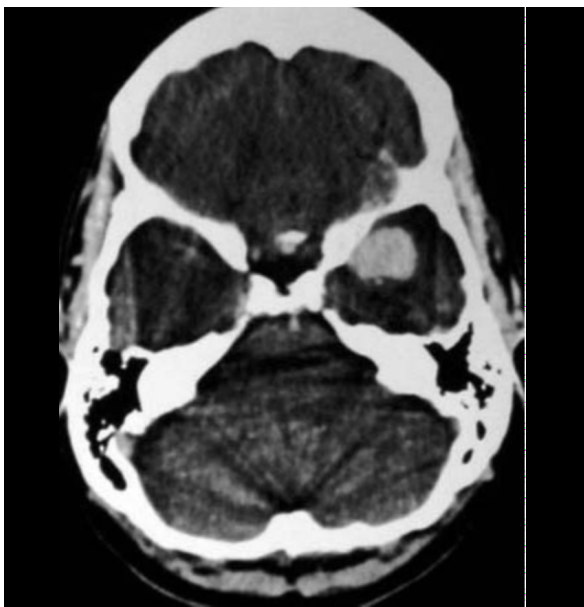


Figure 1A. 43-year-old woman with worsening left-sided headaches. Post-contrast CT scan shows left MCA aneurysm.

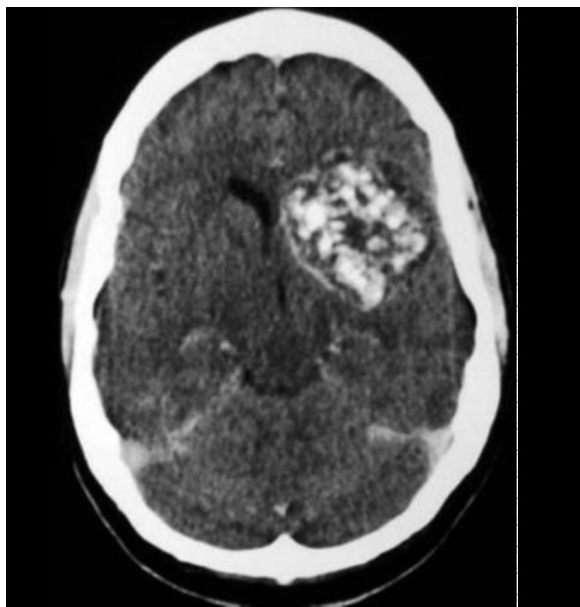


Figure 1B. Post-contrast CT scan shows left cavernous angioma.

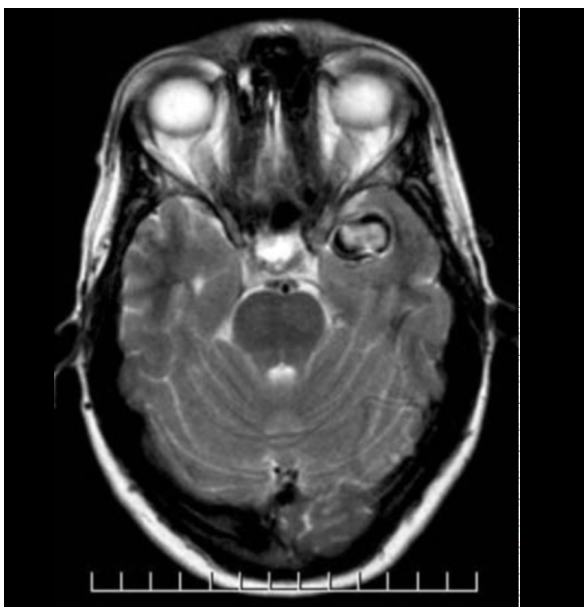


Figure 2A. Axial T2W images demonstrate MCA aneurysm which was originally mistaken for a "tumour nodule."

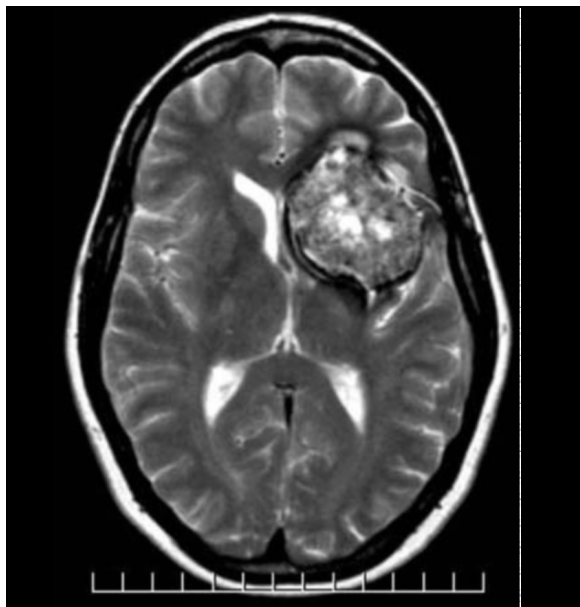


Figure 2B. Axial T2W images demonstrate typical features of a cavernous angioma with a mixed signal core, complete haemosiderin rim, and blooming susceptibility effect.

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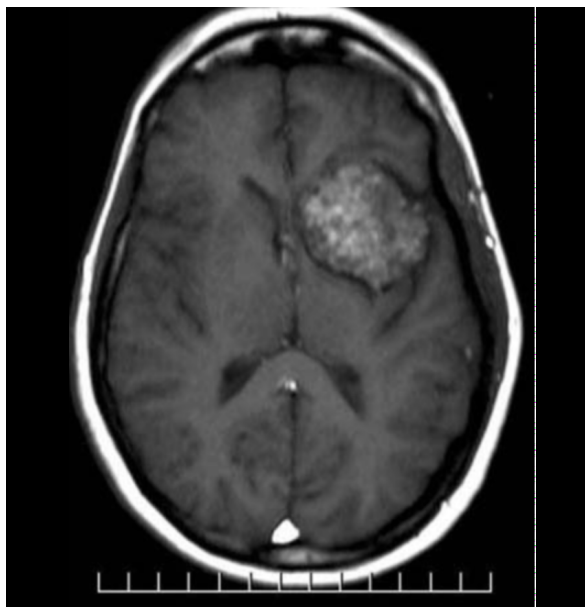


Figure 3A. Axial T1W pre-contrast (gradient echo) MRI. Note the marked heterogeneity within the cavernous angioma, the high signal change which is consistent with blood products.

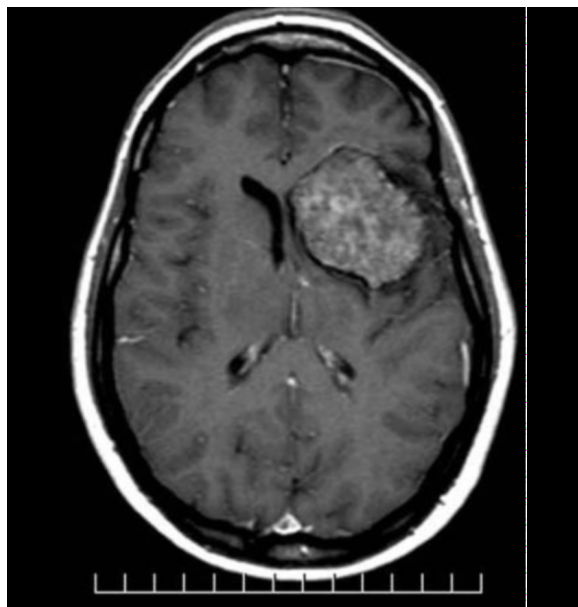


Figure 3B. Axial T1W post-contrast (spin-echo) MRI. No contrast enhancement is demonstrated.

Nineteen years later, at age 43, the patient re-presented with a 6-month history of worsening left-sided headaches although her fits were still well controlled. The CT and MRI showed a 20 mm middle cerebral artery (MCA) bifurcation aneurysm immediately below the cavernous angioma which measured 50 mm (see Figures 1 to 4 and legends for description), findings which were supported by a catheter angiogram (see Figure 5). It was estimated that the risk of bleeding from the aneurysm was 1-2% per year. The prospective surgical option, which would necessitate removal of the cavernous angioma for access to the aneurysm, carried an estimated risk of 15% significant morbidity and mortality and as such, the lady opted for conservative management. Unfortunately, the aneurysm did eventually bleed just 7 months later, requiring her aneurysm to be clipped. The event was not fatal but she was left with moderate neurological sequelae.

Discussion

Cavernous angiomas, also known as cavernomas

or cavernous malformations, are very common lesions with an incidence of 0.5% [1]. Although one fourth of cavernous angiomas occur in children, cavernous angiomas are usually detected in the third to fifth decade and are much more likely to be multiple in familial cases [2]. The favoured imaging modality is MRI. The lesions are characteristically inhomogeneous, with a “popcorn-like” appearance surrounded by a dark rim. Besides a minor contribution by slowly flowing blood containing compartments and possible intralésional calcification, the appearances are mostly explained by the presence of hemoglobin breakdown products within and around the lesion. Methemoglobin, which has paramagnetic properties and hence exhibits hyperintense signal on both T1- and T2-weighted images, is located centrally and is surrounded by a hypointense rim of intracellular hemosiderin [3,4]. The lesions show minimal or no enhancement and exhibit susceptibility “blooming” effects on T2 and T2* sequences. These MRI findings are considered diagnostic and accepted as a definitive diagnosis where histopathology is not available [5]. CT findings are less distinctive. CT will be negative in 30 to 50% especially with smaller lesions. If they are to present clinically either due to bleeding or mass effect,

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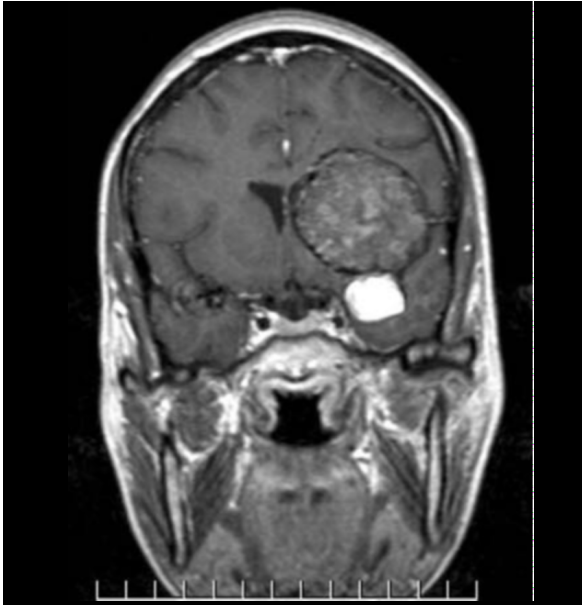


Figure 4. T1W post-contrast coronal MRI clearly demonstrating the non-enhancing cavernoma and the immediately subjacent enhancing aneurysm.

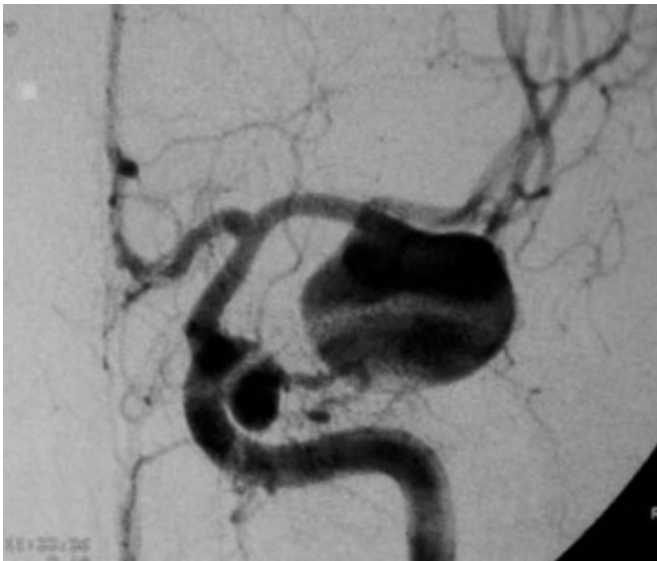


Figure 5. Selective left internal carotid artery angiogram clearly depicting the large left middle cerebral artery aneurysm. The cavernous angioma is angiographically occult.

approximately half will have headache, half will present with seizure and half will have focal neurological deficit [6-8]. After diagnosis the haemorrhage rate is in the region of 3% per patient-year [9] (haemorrhage being defined as symptomatic presence of extralesional blood on MRI scan).

When searching the literature it has become apparent that often no distinction is made between a cavernous angioma and a cavernous haemangioma yet there are important radiological and pathological distinctions between the two entities [10,11]. This lack of distinction appears to arise partly because of confusion in the literature regarding the classification of vascular malformations. Also pathologists are said to often be at a disadvantage due to receiving suboptimal/incomplete specimens [12] and having a lack of information in terms of radiological correlative study and clinical context whilst formulating their reports. This has made it difficult to determine the true incidence of giant cavernous angiomas with several previous reports of giant “cavernous angiomas” actually being cavernous haemangiomas [13-15].

Cavernous haemangiomas (true angioproliferative tumours) are tumours that can be found in both dural and extradural localizations. They are characterized by angiogenic cellular proliferation. They are typically identified on conventional angiography and demonstrate a faint homogeneous capillary blush. Cavernous angiomas on the other hand, are non-tumoral (i.e. non-proliferative) vascular lesions and are the only true venous malformations of the central nervous system. They increase in size because of intracavernous and local intracerebral self-limiting haemorrhagic events [16]. They are not identified on routine angiography because of slow flow, i.e. are angiographically occult lesions [17].

Cavernous angiomas have a known association with other vascular malformations (AVMs, capillary telangiectases and venous angiomas) with the coexistence of two or more of them within the same patient being well documented [18-23]. This is especially so in young children where in most cases rather than the malformation being a pure cavernous angioma, there will be complex/overlapping vascular malformations with pseudoan-

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giomatous vessels, capillary telangiectasia and/or venous ectasia being found in the parenchyma surrounding the cavernoma [24]. This is thought to account for the fact that these “cavernomas” tend to be much larger than those found in adulthood, as they are in fact lesions with mixed pathology, albeit with a cavernous component. This also accounts for their protean radiological appearance, in contrast to the consistent appearances of the more benign adult cavernous angiomas [25-28].

It is exceptionally rare to see a cavernous angioma over 2.5cm presenting in the adult population. There is no known association between cavernous angiomas and intracranial aneurysms. After an extensive literature research we believe this to be the joint 4th largest reported giant cavernous angioma to present in adulthood [29-32] (the largest being 7.5cm by reported Siddique on 2001) and it also happens to be immediately associated with a large intracranial aneurysm, an association never previously been reported in the literature.

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