

## SHORT REPORT

## Spontaneous thrombosis of an arteriovenous malformation

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### Abstract

**A patient with a spontaneously thrombosed arteriovenous malformation (AVM) presented with epilepsy. The CT and MRI appearances were of an intrinsic cerebral neoplasm with extensive surrounding vasogenic cerebral oedema and a mass effect. Histopathology confirmed a large thrombosed AVM. The natural history of AVMs and spontaneous thrombosis are reviewed.**

(*J Neurol Neurosurg Psychiatry* 1994;57:1410-1412)

Arteriovenous malformations (AVMs) are well known for their serious manifestations of haemorrhage, epilepsy, and progressive neurological deficits. Spontaneous thrombosis of AVMs is a rare but recognised event in the natural history of these lesions,<sup>1</sup> which typically occurs silently and is considered an excellent outcome protecting the patient from more serious events. We describe the spontaneous thrombosis of an AVM that probably resulted in the lesion becoming symptomatic with epilepsy. The uncharacteristic radiological appearance of a mass effect and extensive surrounding vasogenic oedema did not suggest the correct diagnosis and hence angiography was not considered. Possible reasons for the presence of vasogenic oedema are discussed. The important principle of obtaining adequate tissue for histopathological assessment in intracranial mass lesions is emphasised.

### Case history

A 34 year old left handed male financial advisor with an otherwise unremarkable medical history presented with a nine month history of typical complex partial epilepsy. He described seizures of about five minutes duration, stereotyped in nature, beginning with increased salivation, followed by a metallic taste, a strong pungent smell, and lastly a period of lack of awareness. Three to four episodes occurred each month, with some reduction in frequency with therapeutic levels of carbamazepine. Neurological and general physical examinations were normal.

### Investigations

Routine haematology, biochemistry, and chest radiograph were normal. An ECG showed persistent left frontotemporal abnor-

malities consistent with a focus in this region. On the non-contrast CT head scan there was an ill defined area of low attenuation in the left temporal lobe, extending superiorly and posteriorly into the inferior parietal lobe with some effacement of cortical sulci over the left hemisphere and slight compression of the left lateral ventricle. A contrast study showed a small area of enhancement in the temporal lobe portion of the lesion.

An MRI study confirmed the mass effect with enlargement of the left temporal lobe, effacement of the cortical sulci, and upward displacement of the sylvian fissure. There was extensive oedema of the white matter of the left temporal and lower parietal lobes. A central area of low signal was thought to represent the lesion's centre (fig 1). There were no flow void phenomena and no abnormal vessels. The radiological appearances were of an intrinsic cerebral neoplasm, most likely of glial type.

### Management

On the basis of the clinical history and radiological investigations an intrinsic glial neoplasm was considered most likely. A craniotomy to obtain histological diagnosis and to debulk the lesion was considered appropriate as the left side likely represented the patient's non-dominant temporal lobe.

### OPERATION

A left temporal craniotomy was performed under general anaesthesia. The dura was opened to show a slightly swollen but otherwise normal temporal lobe. As intended pre-operatively, a cortical incision was made 5 cm from the temporal pole and a modified temporal lobectomy performed preserving the superior temporal gyrus and medial structures, removing a firm dark lesion, 4 cm in diameter, which macroscopically seemed composed of thrombosed vessels. There was no intraoperative haemorrhage and the surrounding white matter seemed macroscopically oedematous.

His immediate postoperative course was unremarkable. He was discharged on the sixth day after operation, without neurological deficit, and with the operative wound well healed.

### HISTOPATHOLOGY

The microscopic appearance was characteristic of an AVM with numerous blood vessels of different sizes; some contiguous and others

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Received 9 March 1994  
and in revised form  
16 May 1994.  
Accepted 24 May 1994

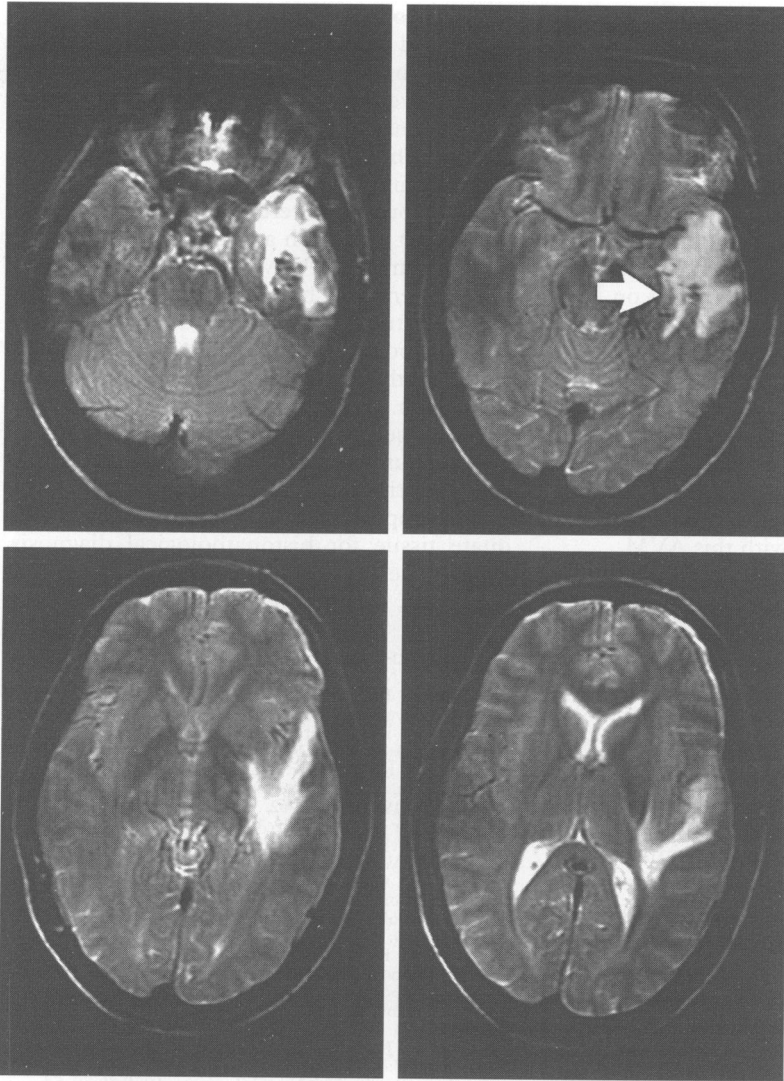


Figure 1 Axial T2 MR images in which the vasogenic oedema of white matter is extensive. The lesion's centre is seen low in the temporal lobe (top right arrow). No signal voids are present to suggest abnormal patent vessels.

separated by either normal or gliosed cerebral cortex (fig 2). Many of the smaller blood vessels had an irregular, sometimes duplicated, internal elastic lamina indicating an arterial wall structure whereas other vessels resembled venous channels. The larger venous channels and occasional arteries contained organised thrombi in which striking recanalisation had occurred. Focal haemosiderin pigment deposition indicated previous microscopic haemorrhage and the surrounding white matter was oedematous.

Figure 2 Histological specimen from operation, showing numerous blood vessels of different sizes, some with organised thrombi and others recanalised.



#### FOLLOW UP

In the two years since operation he has been seizure free, including the past year without medication. He remains without neurological deficit and has returned to his previous occupation. Follow up MRI showed the absence of any remaining abnormality or mass effect.

#### Discussion

Consideration of the natural history of AVMs is extremely important when making reasoned decisions regarding their management. They are theoretically present at birth, yet mostly become symptomatic during the second to fourth decades,<sup>1-3</sup> hence they are not static entities, but are dynamic and change with time.<sup>4</sup> Progressive enlargement of feeding arteries and veins, enlargement of the nidus, increased shunting, and, rarely, regression or spontaneous thrombosis occur.<sup>4,5</sup> Spontaneous thrombosis and regression of an AVM is generally considered a favourable event, which occurs silently, subsequently protecting the patient from intracranial haemorrhage.<sup>1</sup> This rare event of spontaneous thrombosis occurred in the case we describe, but during this event the patient became symptomatic. Symptomatic complete thrombosis of an AVM with no previous history of haemorrhage is uncommon; only 32 cases could be found in a review of the literature by Warren *et al.*<sup>6</sup>

Wilkins<sup>1</sup> in a comprehensive review of 1500 patients, determined that an unruptured AVM carries an annual risk of haemorrhage of 2-3%, with an annual risk of death of 1%. Haemorrhage, seizures, and headaches, individually or in combination, are the most common manifestations. The phenomenon of apparent spontaneous closure, he concluded, occurred very rarely and was not a real consideration in the outlook of an untreated lesion. No mention was made of the incidence of spontaneous thrombosis by Crawford *et al.*,<sup>2</sup> reporting a mean 10 year follow up of 217 patients with unoperated AVMs.

The fact that AVMs change size with time has been documented in several angiographic series.<sup>7-9</sup> Most AVMs increased in size, a minority remained the same, and they regressed only rarely. Those lesions that reduced in size or completely regressed were small or fed by a single or a few feeding arteries. In our case, it was impossible to determine the number of feeding arteries but the lesion was not less than 4 cm in diameter.

A CT without contrast enhancement shows calcification in up to 25% of patients harbouring an AVM. A mass effect is rarely seen. Hypodensity of the surrounding brain parenchyma may be seen, indicating atrophy or gliosis caused by vascular steal or previous haemorrhage. With contrast there is pronounced enhancement of the nidus, feeding arteries, and draining veins.<sup>10</sup> The CT appearance of previously reported thrombosed AVMs is of a mass with increased density and usually showing slight enhancement

with contrast. A mass effect and surrounding oedema were not reported.<sup>6</sup> The CT studies, with and without contrast, in this reported case did not show any calcification. There was a pronounced mass effect, very little contrast enhancement, and the hypodensity shown was not suggestive of atrophy.

Signal void due to high flow through the nidus, feeding arteries, and draining veins which is the typical finding of AVMs on MRI was absent in this case. A mass effect on MRI and CT is not characteristic of AVMs.<sup>11</sup> The extensive area and increased signal within the associated temporoparietal lobe white matter on the T2 weighted MRI sequence is suggestive of extensive vasogenic oedema<sup>12</sup> and is supported by the histopathology findings. The reason for the occurrence of this is not clear. There was no suggestion of underlying neoplasm in association with this AVM.

The phenomenon of normal perfusion breakthrough after the surgical resection of large AVMs occurs as blood is suddenly shunted through neighbouring channels that had previously been under perfused because of the steal effect of the AVM.<sup>13,14</sup> A local disturbance of the blood-brain barrier leads to cerebral swelling from vasogenic oedema or haemorrhage. The redistribution of blood flow that followed the spontaneous thrombosis of this AVM may have had a similar effect suggesting this phenomenon as a possible mechanism for the formation of the vasogenic oedema. Many thrombosed vessels were recanalising with new vascular endothelium, which does not possess the "tight" junctions of vascular endothelium typical of cerebral blood vessels and forming the blood brain barrier. Leaking of fluid from these new vessels may also contribute to the vasogenic oedema.

Seizures are a common first manifestation of AVMs.<sup>1,2</sup> Neuronal damage or irritation from ischaemia, compression by the enlarging malformation, progressive reactive gliosis, or iron pigment deposited after haemorrhage<sup>15</sup> are suggested as pathophysiological mechanisms of seizure initiation. The presence of iron pigment in this case suggests previous occult haemorrhage, but irritation of the surrounding neurons by the mass effect and vasogenic oedema may also have contributed to initiation of seizures.

Many hypotheses have been suggested as the mechanism for spontaneous thrombosis of AVMs including progressive arteriosclerosis and occlusion of feeding vessels,<sup>16</sup> occluding emboli,<sup>17</sup> episodes of stagnant or slow flow,<sup>18</sup> intravascular turbulence from the irregular tortuous vessels,<sup>19</sup> and the hypercoagulable state in hormonally active females.<sup>20</sup> The relation between thrombosed AVMs and angiographically occult AVMs was reviewed by Ebeling,<sup>21</sup> who suggested that thrombosed AVMs may haemorrhage from recanalising vessels. The management of asymptomatic spontaneously thrombosed AVMs remains uncertain as does their true natural history and exact relation with angiographically occult AVMs.<sup>22</sup>

## Conclusion

When and why this AVM thrombosed and its relation to the onset of the seizures and to their precipitation remains uncertain. Suggestions and possibilities have been discussed. The CT and MRI appearances were of an intrinsic cerebral neoplasm with extensive surrounding cerebral oedema but the findings at operation and histopathology were of a thrombosed AVM. The phenomenon of normal perfusion breakthrough subsequent to the redistribution of blood flow on the AVM's spontaneous thrombosis is a possible cause of the vasogenic oedema surrounding this lesion. The recanalisation of the thrombosed vessels with channels that have vascular endothelium without tight junctions is a possible contributory factor.

The important principle of obtaining adequate tissue for histopathological diagnosis, even if the most advanced neuroradiology is employed, is emphasised.

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