Angiographically occult arteriovenous malformations of the brain

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SUMMARY Six patients with cerebral arteriovenous malformations which did not show any pathological circulation at angiography are described. Computed tomogram appearances of such lesions may be difficult to distinguish from tumours. The need for surgical exploration in localised high attenuation lesions of uncertain nature is stressed, and the literature is reviewed.

Since the first description of a vascular malformation of the brain by Luschka in 1854, the presentation and progression of these congenital anomalies, and their surgical treatment, have been under continuous study. The first surgical exposure of such a lesion was performed by Giordano in 1890, but logical treatment depends on angiographic evaluation of the malformation, and modern surgery has evolved from the classical work of Bergstrand *et al.* (1936).

In 1948 Olivecrona and Riives published a detailed report of 60 patients, in which they stated that angiograms always disclosed the presence of an arteriovenous malformation (AVM), and implied that this was the ultimate diagnostic test. However, Crawford and Russell (1956) challenged this statement by describing AVMs which appeared as avascular masses in two patients. In all, 27 AVMs without angiographically demonstrable circulation have been described up to the present (Table 1). We present a further six cases, recently investigated, all of which showed focal high attenuation lesions on computed tomography (CAT).

Case studies

The relevant clinical abnormalities and the findings on special studies are summarised in Table 2. There were four women and two men aged between 24 and 55 years (mean 40 yr). Two had temporal lobe seizures, two Jacksonian fits and a minor degree of hemiparesis, and one severe headaches alone. The sixth patient presented acutely with an intracerebral haematoma.

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Table 1 Details of previously reported cases of arteriovenous malformation of the brain without angiographically demonstrable circulation

Reference	Year	Age (yr)	Sex	Angiographic appearance
Crawford and Russell	1956	32 11	M F	Mass Mass
Paterson and McKissock	1956	14 27	M F	Normal Normal (calcification seen)
Castaigne <i>et al.</i>	1961	31	F	Mass
Papathodorou <i>et al.</i>	1961	25	M	Normal
Kamrin and Buchsbaum	1965	15 46	F F	Normal Mass
Hammon	1969	7	M	Mass
Porter and Bull	1969	21	M	Normal
Salibi	1969	20	M	Mass
Scott <i>et al.</i>	1973	1½	M	Mass
Kendall and Claveria	1976	47	F	Mass
Kramer and Wing	1977	*	*	Seven cases: mass Two cases: normal One case: calcification and arterial dilatation
Terbrugge <i>et al.</i>	1977	*	*	Normal
Golden and Kramer	1978	36 32 20	F F F	Normal Normal Mass

*Details not given by author.

Angiography failed to show any abnormality in cases 2 and 4. Small avascular swellings were shown in cases 3 and 5, and typical vessel displacements were caused by the right posterotemporal haematoma in case 3. A posterolateral choroidal artery was rather prominent in case 1 (Fig. 1), but no pathological vessels were outlined in any of the angiograms. There was focal increase in the uptake of ^{99m}Tc in case 6, but gamma scans

Table 2 Summary of clinical abnormalities and findings in six additional cases of angiographically occult arteriovenous malformation of the brain

Case	Sex	Presenting symptom	Dominant hand	Skull radiograph	EEG	γ -scan	Angio	Pneumo-encephalography	CAT scan	Operative findings	Histology
1 FC (24 yr)	F	Headache Loss of memory and concentration	Left	Nodular calcification in right lateral ventricle	Amplitude greater on right	Normal	Normal (Fig. 1)	Mass in body of right lateral ventricle	Fig. 3 Enhanced with intravenous contrast medium†	No haematoma	Large vascular spaces, lined with collagen and a single layer of endothelial cells, containing thrombus.
2 AE (32 yr)	F	Temporal lobe epilepsy	Right	Normal	Right temporal * slowing on over breathing	*	Normal	*	Fig. 4 Did not enhance	No haematoma	Tangle of vessels, which mainly resemble veins containing thrombus.
3 DH (46 yr)	M	Fall three weeks earlier, impaired conscious level, and left hemiparesis for six days	Right	Left temporal fracture	*	*	Avascular temporal lobe mass	*	Fig. 5 Enhanced†	Haematoma	Small and medium sized vessels with adjacent blood clot.
4 RH (38 yr)	M	Left focal fits and hemiparesis	Left	Normal	Normal	Normal	Normal	Left ventricular enlargement Right shift of septum of pellucidum	Fig. 6 Enhanced†	Cyst No haematoma	Abnormal vessels with a thin fibrous wall and endothelial lining, containing thrombus
5 AM (47 yr)	F	Temporal lobe epilepsy	Right	Normal	Right temporal slow waves	Normal	Temporal lobe mass	*	Fig. 7 Enhanced†	No haematoma	Veins of varying calibre, containing thrombus.
6 GT (55 yr)	F	Right focal fits and hemiparesis	Left	Normal	Left para-sagittal theta waves	Left frontal parietal high uptake	Left hemi-sphere swelling	*	Fig. 8 Enhanced†	Cyst No haematoma	Vessels varying from capillaries to large spaces. Extensive thrombosis and some organisation

* Investigation not done.

† Contrast medium—meglumine iothalamate

(a)

(b)

Fig. 1 Case 1. (a) Vertebral angiogram: lateral projection, magnified arterial phase. The choroidal arteries (→) are prominent, but not abnormal. No pathological circulation is shown. Note the nodular calcification between arrowheads in the region of the body of the lateral ventricle. (b) Subtraction of slightly later phase. There is good filling of all the arteries up to the peripheral branches. No abnormal circulation is shown.

(a)

(b)

Fig. 2 Case 1. Pneumoencephalogram, (a) lateral brow up, (b) brow down. A mass is partially outlined in the body and trigonal region of the right lateral ventricle (roof indicated by arrows) and the calcified nodules (>) are localised within it.

were normal in cases 1, 4, and 5. Electroencephalography showed a minor abnormality on the side of the lesion in four cases, was normal in one, and was not performed in one. A plain skull radiograph was abnormal in case 1, showing multinodular calcification, and a pneumoencephalogram (Fig. 2) confirmed that this was within a mass extending into the right lateral ventricle.

A high attenuation lesion was delineated by plain CAT in every case. In case 1 it encroached into the lateral ventricle and contained nodular

Fig. 3 Case 1. (a) Plain CAT. There is an irregular high attenuation (25–56 EMI units) partly calcified lesion occupying most of the body and trigone of the right lateral ventricle, and partially obstructing it, without further mass effect. (b) After intravenous contrast medium. There is marked enhancement in the lesion. The left choroid plexus is shown normally enhanced. A thrombosed AVM was removed from the right lateral ventricle.

calcification (Fig. 3). In case 3 (Fig. 5) who had bled three weeks previously, a typical intracerebral haematoma was shown. Computed tomography was repeated in all cases after intravenous contrast medium, and enhancement occurred in five of them. In two cases, part of the enhanced shadow had a curvilinear form; in case 3 it extended well outside, and also into the haematoma (Fig. 5). Mass effect was present in case 3, and to a minor extent in cases 5 and 6; there was atrophy in case 4 and intracerebral cystic change in case 6.

Histology confirmed the AVM in every case. With the exception of case 3, the vessels of the AVM were occluded by thrombus, which was presumably the cause of non-filling at the time of angiography.

Fig. 4 Case 2. Plain CAT. There is a high attenuation (38 EMI units) lesion in the posterior part of the right temporal lobe. There is only minimal mass effect and there was no enhancement after intravenous contrast medium. There was good filling of the posterior cerebral artery on right carotid angiography, which showed no abnormality. Histology of a biopsy specimen showed a thrombosed AVM.

Results of surgery

In case 1, concentration and memory were improved by operation although there was some post-operative paresis affecting the left arm. In case 2 a biopsy only was performed, with no effect on the temporal lobe fits. In case 3 there was no neurological deficit after surgery, and the patient returned to work as a boiler engineer.

In case 4 the focal fits ceased after the operation. The left hemiparesis improved, and the patient returned to his work as a restaurant manager.

In case 5 the temporal lobe fits were abolished by the temporal lobectomy, and in case 6 the right focal fits did not recur postoperatively, and the right hemiparesis continues to improve.

Discussion

A malformation may not be shown at angiography for technical reasons which can be avoided by modern techniques. Rapid circulation through arteriovenous shunts is easily recorded on rapid serial films. An AVM situated at the junction of two major arterial territories may not be filled by selective injection of one of them, but catheter angiography facilitates examination of all potential feeding arteries. Lesions near the midline, which often fill from the contralateral anterior cerebral artery, as observed by Olivecrona and Riives (1948), and by Paterson and McKissock (1956), are always subjected to bilateral angiography.

Fig. 5 Case 3. (a) Plain CAT. There is a haematoma in the posterior part of the right temporal lobe causing compression of the lateral ventricle and slight midline shift. (b and c) After intravenous contrast medium. There is enhancement around the haematoma with serpiginous components extending towards the convexity. The appearances strongly suggest an angiomatous malformation adjacent to the haematoma. The AVM did not fill at angiography but its presence was confirmed at operation and by histological examination of sections of the wall of the haematoma cavity.

Fig. 6 Case 4. (a and b) Plain CAT. There is an irregular high attenuation lesion (25-30 EMI units) without mass effect in the right frontal parietal region above the lateral ventricle. (c and d) After intravenous contrast medium. Slight curvilinear enhancement is shown. Angiography was normal. A gliotic and cystic mass containing thrombosed angioma was removed at surgery.

Fig. 7 Case 5. (a and b) Plain CAT: two adjacent cuts. A large irregular and mottled area of high attenuation is shown in the anterior and medial parts of the right temporal lobe. It enhanced only slightly after intravenous contrast medium. Carotid and vertebral angiography showed a small anterior temporal mass, but no abnormal circulation. At operation a completely thrombosed AVM was excised.

Fig. 8 Case 6. (a and b) Plain CAT. There is a high attenuation lesion in the posterior part of the left frontal lobe, with a cyst of water attenuation anterior to it, causing slight mass effect. (c and d) After intravenous contrast medium. There is some enhancement of the high attenuation lesion. No filling at angiography. The cyst communicated with the lateral ventricle near the foramen of Monro, and contained an extensively thrombosed angioma in its posterior wall.

An AVM, particularly if very small, may be destroyed by haemorrhage or may fail to outline because of compression by adjacent clot, as was evident in case 3. It is known that malformations can thrombose spontaneously and apparently disappear on angiography (Conforti, 1971). Partial spontaneous thrombosis was first documented in 1949 by Norlén, and if extensive, will diminish the flow and prevent the vessels filling with sufficient contrast medium to appear radiopaque. Kramer and Wing (1977) suggested that CAT could detect these small concentrations of contrast medium, but extravasation of contrast medium into damaged tissues also contributes to the enhancement shown by a proportion of these lesions. The occluding thrombus may organise and form connective tissue (Paterson and McKissock, 1956), as occurred in our case 6.

Plain skull radiographs show abnormal calcification related to an AVM in 15% (Houdart and Le Besnerais, 1963) to 29.5% (Rumbaugh and Potts, 1966) of cases. It can be in the walls of the abnormal vessels or in the adjacent brain. Although it is detected by CAT when present at lower concentrations, its structure is better defined on plain radiography. A curvilinear form, suggesting blood vessels, is most typical of AVM, but a nodular form is more frequent and is helpful when related to a subarachnoid or intracerebral haemorrhage (Hayward, 1976).

Despite the enthusiastic report of Tinterov (1969), isotope scanning has consistently shown only large or superficial AVMs; about 40% of those under 20 mm in diameter are not revealed (Planiol and Akerman, 1965; Waltimo *et al.*, 1973) even in optimum conditions. The more specific features of isotope scanning in AVMs reflect the increased blood flow and would clearly be absent in the subgroup under discussion. Electroencephalography usually provides supporting evidence of an organic lesion, but is nonspecific.

Computed tomography revealed a lesion in all cases and often suggested its nature. Kendall and Claveria (1976) found diagnostic CAT appearances in 66% of patients with AVMs. Typically, an AVM is of slightly increased attenuation, corresponding to that of circulating blood, which is proportional to the haemoglobin level and equivalent to 28 EMI units with a haemoglobin of 15 g/dl. It may be of greater attenuation when clotting or calcification has occurred within or adjacent to the lesion, or if a haematoma is present. These secondary changes may be the most prominent features of the plain CAT.

Enhancement is usually considerable and enlarged vessels constituting the AVM may be more

evident, appearing tubular or vermiform when they lie in the plane of the tomographic section, and rounded or mottled when at right angles to it. Feeding arteries and draining veins may be recognised and provide useful evidence of the nature of an atypical lesion. An AVM may be suspected in the presence of an intracerebral haematoma when any of these features are present. Enhancement related to a spontaneous haemorrhage is usually confined to the immediate surrounds, and rarely evident less than six days after the bleed (Kendall and Radü, 1978). Earlier or more extensive enhancement should suggest an underlying angioma or tumour.

Kramer and Wing (1977), and more recently Golden and Kramer (1978), reported similarities in the CAT appearances of angiographically avascular AVMs and tumours. Cases 2 and 6 in our series could not be recognised as AVMs before surgery and, although the diagnosis was suspected in the other cases, surgery was necessary for the confirmation and treatment of the lesion.

Histological examination of each operative specimen demonstrated an AVM with vessels of varying calibre. Their walls tended to be degenerate and fibrosed, with dystrophic calcification and no elastic lamina. The surrounding brain was oedematous, showed gliosis, and contained haemosiderin and an infiltrate of mononuclear cells. Preoperative thrombosis was evident within the lumina in all except case 3, and was organising in case 6. Surgical exposure is usually necessary to confirm the diagnosis and can be followed by excision in suitable cases. Excision was achieved in four of our cases, a formal temporal lobectomy being carried out in case 5 where the lesion lay medially.

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