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Multiple cerebral arteriovenous malformations (AVMs)

Review of our experience from 203 patients with cerebral vascular lesions

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Summary. From our series of 203 patients with cerebral vascular lesions, 18 (9%) could be included in the multiple arteriovenous malformation category. There were five patients with Rendu-Osler-Weber, one with Wyburn-Mason syndromes and two with concurrent arteriovenous malformations. The remaining ten patients (4%) had multiple brain arteriovenous malformations. Careful angiography with magnification is necessary to try to diagnose multiple brain AVMs, since these sometimes become apparent only after embolization of a larger dominant AVM. The incidence of multiple brain arteriovenous malformations is likely to have been underestimated due to the failure to recognize microarteriovenous malformations associated with larger arteriovenous malformations.

Key words: Multiple arteriovenous malformations – Central nervous system – Cerebral angiography

Considering the congenital nature of cerebral vascular malformations, it is surprising that there are few reports of multiple arteriovenous malformations (AVMs) in the central nervous system (CNS). The literature has a limited number of reports on multiple brain AVMs [1–7], on the association of cerebral and spinal angiomas [8–10], on retinocephalic vascular malformations [11–13], and on angiomatoses with CNS involvement [14].

Methods

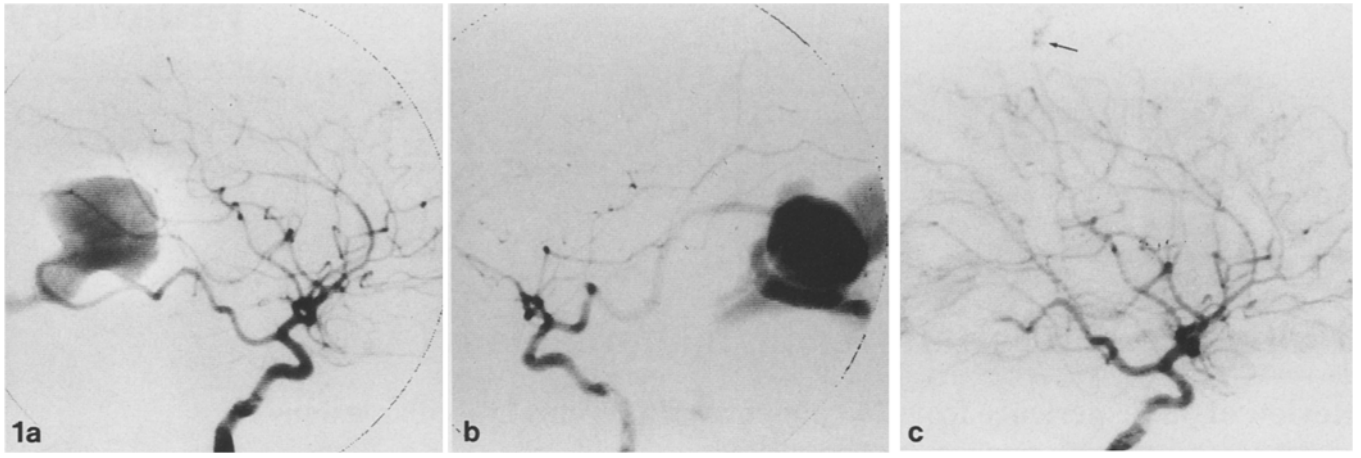
We reviewed the charts of 229 patients with cerebral vascular lesions diagnosed and/or treated at the Hopital Bicetre and the Toronto Western Hospital between 1981 and 1986. Twenty-six patients were discarded due to incomplete charts or inadequate angiography, leaving 203 patients. Patients with Sturge-Weber and cavernomas were also excluded from consideration, since these are venous malformations. For clarity, we separate those patients with multiple cerebral AVMs from those with a brain AVM associated with an AVM outside the CNS.

Results

Eighteen of the 203 patients fell into the multiple malformation group. The distribution of these multiple AVMs is presented in Table 1. Table 2 presents the clinical data and topography for each patient. The average age of these 18 patients was 29, ranging from 1 to 70 years. There were ten females and eight males. The group with multiple brain AVMs presented with one or more of the commonly recognized clinical features: neurological deficit, hemorrhage, headache and seizure. In the group with associated AVMs, the brain AVM was incidental in one. In the Rendu-Osler-Weber (ROW) group, two of the five patients had no symptoms related to their brain lesion. In the three youngest patients in the series, the main lesions were arteriovenous fistulae (AVF). One of these patients (patient 10) presented with aneurysmal dilatation of the vein of Galen and 2 separate vascular shunts. A second (patient 5) had bilateral occipital AVF (Fig. 1) with marked venous ectasias and a small parietal AVM. The third (patient 5) had an aneurysmal malformation of the vein of

Table 1. Distribution of our multiple arteriovenous malformations ($n = 18$)

Rendu-Osler-Weber	5
Wyburn Mason	1
Multiple Nidus	10
Associated systemic AVMs	2



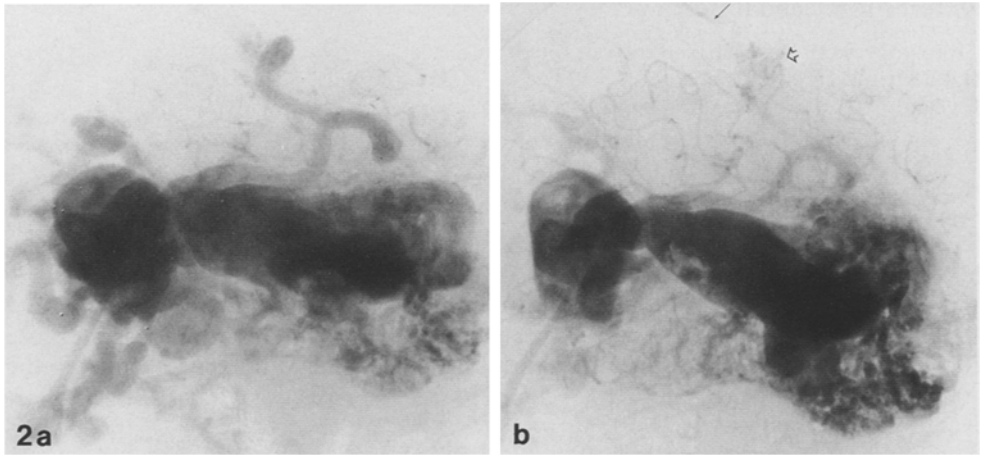


Fig. 1. Lateral right (**a**) and left (**b**) internal carotid angiograms in a patient with ROW show bilateral occipital AVF. Post embolization right internal carotid angiogram (**c**) shows micro-AVM *(arrow)*

Fig. 2 a, b. Patient with a large temporal AVM. Pre (**a**) and post-embolization (**b**) internal carotid angiograms show that an associated micro-AVM *(open arrow)* with its early draining vein *(closed arrow)* is better visualized after embolization

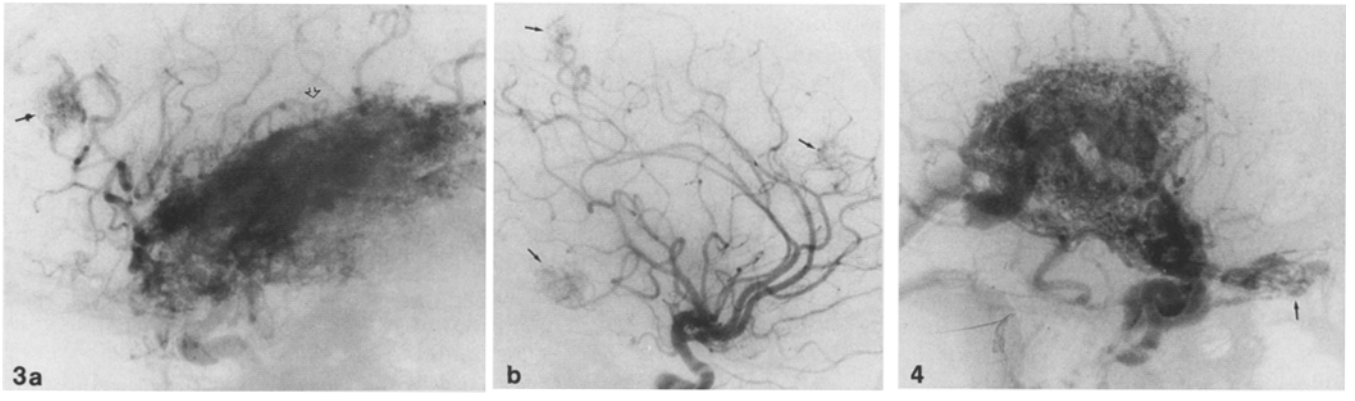


Fig. 3. Lateral left (**a**) and right (**b**) internal carotid angiograms show a large insular AVM *(open arrow)* with multiple superficial micro-AVMs *(closed arrows)*

Fig. 4. Lateral internal carotid angiogram shows a deep-seated AVM which extends into the orbit *(arrow)*

Galen with bilateral AVF at the level of the thalamostriate veins.

Three patients had small AVMs which we call micro-AVMs (patients 5, 6, 7). These were cortical or superficial with a feeding artery of normal size, a nidus less than 1 cm and an early draining vein of normal size [15]. In patient 7 the micro-AVM was not visualized after embolization of the larger AVM. In patient 6 one of the micro-AVMs was visualized after embolization of the largest AVM (Fig. 2). Patient 12 had an ipsilateral bone dysplasia associated with separate orbital, maxillofacial, mesencephalic and thalamic AVMs, all on the same side. Most patients in the multiple nidus group had either deep or cortical lesions, not both. Only patient 8 had separate cortical and deep AVMs. Patient 14 had a cortical AVM with a separate dural AVM. Apart from the ROW group, two patients had skin telangiectasias which were clinically distinct from the skin lesions of ROW. One of these patients had a family history of vascular lesions.

Table 2. Clinical summary

	Patient	Sex/Age	Clinical	Type	No. of Nidus (CNS)	Associated AVMs
ROW	1	F/28	Seizure	Cortical	2	
	2	M/60	Headache, seizure	Cortical	1	
	3	M/56	–	Cortical	1	
	4	F/56	–	Deep	1	
	5	F/7	Hemorrhage	Cortical	3	
Multiple Nidus	6	F/14	Seizure	Cortical	5	Skin telangiectasia
	7	M/33	Headache, deficit, hemorrhage, seizure	Cortical	2	
	8	F/26	Deficit	Cortical deep	2	
	9	F/25	Deficit	Deep	2	
	10	M/12	Hemorrhage, hydrocephalus, cardiac failure	Deep	2	Skin telangiectasia
	11	M/70	Hemorrhage	Cortical	2	
	12	M/12	Hemorrhage (oral), bone dysplasia	Deep	2	Orbit, maxillofacial
	13	F/30	Deficit	Cortical	2	
	14	F/33	Hemorrhage, headache, deficit	Cortical dural	2	
	15	M/1	Hydrocephalus	Deep	2	
Associated AVMs	16	M/34	–	Deep	1	Facial
	17	F/29	Hemorrhage	Deep	1	Liver hemangioma
Wyburn-Mason	18	F/13	Deficit, headache	Deep	1	Retinocephalic

Discussion

The Rendu-Osler-Weber syndrome is characterized by multiple telangiectatic lesions of the face and body, giving rise to recurrent epistaxes, melena, hematemesis, genital bleeding and other hemorrhagic complications associated with secondary anemia. It is a congenital malformation inherited as an autosomal dominant with good penetrance but variable expressivity. Central nervous system involvement is common [16–19]. Roman found vascular malformations of the brain in 28% and spinal AVMs in 8% of 215 patients with ROW [18]. Reddy reported one patient with 3 separate cerebral AVMs and a family history of ROW [3]. In our series, five of the 18 patients had ROW with large AVMs, micro-AVMs and/or arteriovenous fistulae of the CNS (Fig. 1).

To our knowledge, no new AVM has been seen to develop in the brain of any patient with ROW. Thus, in ROW the brain AVMs conform to the usual congenital types. In contradistinction, the telangiectasias of ROW are acquired lesions which appear to result from hyperactive angiogenetic factors.

The present clinical definition of ROW (hereditary hemorrhagic telangiectasia) is too self-limiting. It should be broadened to include familial diseases of multiple AVMs and hyperactive angiogenetic factors (telangiectasias). In this light, two other patients in our series could have been included in this expanded ROW group. One of these patients (patient 10) had a vein of Galen ectasia with skin telangiectasias and a family history of AVMs. In addition, patient 6 had 5 cerebral AVMs and skin telangiectasias (Fig. 3). Since other reports of familial cerebral AVMs [20–23] may fit into this group, diligent search for skin telangiectasias should be included in the evaluation of all patients with cerebral AVMs.

The Wyburn-Mason syndrome is an arteriovenous malformation of one or both sides of the midbrain with ipsilateral or bilateral arteriovenous malformation or other vascular anomalies of the retina and multiple cutaneous nevi. Theron reviewed 25 such cases and referred to this entity as unilateral retinocephalic vascular malformations [12]. Eight of the 25 cases had cutaneous vascular nevi. All but three cases involved the optic nerves. The jaws, nose and mouth were frequently involved. When the brain is involved, the malformation closely follows the optic tracts and optic radiations. This series included one patient with Wyburn-Mason disease (case 18) (Fig. 4). In addition, patient 12 could be considered a variant of Wyburn-Mason, since he did have unilateral orbital and brain involvement. However, the AVMs did not follow the optic tracts and the lesions were discreet, unlike the characteristic Wyburn-Mason.

The association of cerebral AVMs with systemic vascular malformations is well known [2, 11, 14]. Hanieh reported a case in which 2 cerebral AVMs were associated with 3 bluish, pulsatile swellings in the left hand, thought to be vascular malformations [2]. Smith reported a case of congenital multiple angiomatosis with skin and brain involvement [14]. Tamaki reported a case with an AVM involving the scalp as well as dura, retina, cerebrum and posterior fossa. This may belong to the unilateral retinocephalic group [11]. In our series, patient 16 had a facial AVM and an asymptomatic hypothalamic AVM, while patient 17 had a posterior fossa AVM and a liver hemangioma. Since we could not find a previous similar report of concurrent cerebral and hepatic malformations, that association may be fortuitous.

The association of a dural AVM and a brain AVM occurred in one of our patients (patient 14). The lateral sinuses were patent, so we could not implicate a thrombosis of a sinus as a cause of the dural shunt [24, 25]. Schlacter's report on multiple AVMs included a patient with a dural AVM of the tentorium and a so-called "cryptic" malformation in the right hemisphere [4].

The concept that small arteriovenous shunts form a subgroup of arteriovenous malformations must be challenged. Three of the ten patients in our group with multiple cerebral AVMs had small or micro-AVMs. The

question raised is whether these are true AVMs or shunts that open in response to the changes induced by the larger AVM. We believe that visualization of small shunts following embolization of large AVMs was likely due to reduction of the steal phenomena through the larger AVM and thus to better visualization of the adjacent territories. In patient 7 the microarteriovenous shunt could not be seen after embolization of the larger AVM. This is surprising. However, we could suggest that the embolization either induced a thrombosis of this lesion (if it was an AVM) or that the ischemia-related shunt spontaneously disappeared in conjunction with the embolization. These concepts as well as the question of growth of these small AVMs must be explored further. Micro-AVMs must be carefully searched for on high quality angiography, since they are occult to CT and MRI [15].

The diagnosis of multiple brain AVMs can sometimes be difficult. Since early venous return related to ischaemia does not occur in the deep system, deep arteriovenous shunts associated with a separate brain AVM certainly represent multiple brain AVMs [26]. Similarly, contralateral arteriovenous shunts obviously represent multiple brain AVMs. Thus only the superficial arteriovenous shunts that are ipsilateral to the brain AVM remain controversial.

Patients with multiple brain AVMs have niduses that are separated by normal brain tissue. In some cases it is difficult to distinguish between a single nidus with multiple angiographic compartments and multiple niduses. With cortical-ventricular AVMs the superficial and deep components often have their own arteries and veins, yet, anatomically, these are single lesions. If we are not sure whether the nidus is single or multiple, we consider the AVM to be a single unit.

Three of our patients had multiple AVF draining into the deep venous system. The deep venous system, unlike the superficial, converges at the vein of Galen. We consider multiple AVF to represent multiple AVMs when they have separate draining veins which also collect normal veins before converging. In two of these cases the AVF were bilateral and thus clearly separate AVMs. In the third patient, one fistula was at the foramen of Monro and the other was on the same side in the wall of the vein of Galen. The importance of this grouping between multiple and isolated AVFs relates to treatment. If multiple, treatment of one of the AVF by embolization leaves the second AVF open. If single, treatment of one of the shunts by embolization (blocking both the shunt and the exit veins) will be an effective treatment of the malformation even if other shunts exist into this single compartment.

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