THE CEREBELLAR ARTERIOVENOUS MALFORMATIONS

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The anatomic diversity of the cerebellar arteriovenous malformations requires a classification which is more surgically informative. The selection tools like the Spetzler-Martin grading system are best suited for the cerebral arteriovenous malformations, while they are quite inaccurate for the cerebellar AVMs. For this reason, in order to define the subtypes of cerebellar arteriovenous malformations that clarify the anatomy and the surgical management, Kim et al. (2015) had validated a supplementary grading system for the improvement of the preoperative risk prediction and for a more adequate selection of the patients for surgery. The predictive accuracy of the supplementary grading system had been superior to that of the Spetzler-Martin grading for the cerebellar arteriovenous malformations. It had also been proven that the supplementary grading system is better than the Spetzler-Martin system for the prediction of the outcomes after the resection of a cerebellar arteriovenous malformation. In a consecutive surgical series of 367 patients, 28 (7.62%) had cerebellar arteriovenous malformations. The subtypes of the cerebellar arteriovenous malformations had been as follows: hemispheric 9 (32.14%); vermian 7 (25%); tentorial 4 (14.28%); tonsillar 3 (10.71%); suboccipital 3 (10.71%); of the cerebellopontine angle 2 (7.14%). The patients with cerebellar arteriovenous malformations had presented more often with hemorrhage than the patients with cerebral AVMs, reason which justifies the use of a more aggressive treatment. The postoperative evolution of the 28 patients with cerebellar arteriovenous malformations had been very good and good, without being registered any important neurological deficits. The evolution of the patients with tonsillar and tentorial arteriovenous malformations had been the most favorable.

Keywords: Cerebellum, Arteriovenous malformation, Spetzler-Martin grading scale, Microsurgical resection, Aberrant angiogenic characteristics of arteriovenous malformations.

INTRODUCTION

The cerebellar arteriovenous malformations differ from the cerebral AVMs with respect to their hemorrhagic behavior, clinical presentation, and surgical outcomes. They make up < 15% of all the brain arteriovenous malformations (Arnaout et al., 2009).

One of the most devastating symptoms of the cerebral arteriovenous malformations is hemorrhage. The risk of this event is expressed in terms of a low annual risk of 2% to 4% per year over a patient’s lifetime (Brown et al., 1988; Ondra et al., 1990; Mast et al., 1997; Wedderburn et al., 2008; da Costa et al., 2009), although it had been shown that factors such as previous hemorrhage, age, location, and drainage patterns influence their natural history (Marks et al., 1990; Pollock et al., 1996; Stapf et al., 2006).

Peerless et al. (1996) had shown that the arteriovenous malformations which are located in a cerebellar hemisphere, where in the vermis predominate. Unfortunately, the arteriovenous malformations located in the brain stem are the next most common group.

The arteriovenous malformations located in the cerebellopontine angle, in the flocculonodular lobe, and in the fourth ventricle are less common, but they pose their own unique technical challenges (Peerless et al., 1996). Kopitnik et al. (2004) had categorized the arteriovenous malformations which are found in the posterior fossa as located in the cerebellar vermis, in the cerebellar hemisphere, in the cerebellar tonsil, in the superficial pial brainstem and in the deep parenchymal brainstem.
Rodriguez-Hernandez et al. (2012) had described the following 5 subtypes of cerebellar arteriovenous malformations: suboccipital, vermian, tonsillar, tentorial, and petrosal, which we are going to summarize below.

The **suboccipital** arteriovenous malformations are located on the posterior cerebellar surface facing the occipital bone, they being positioned below and between the transverse and the sigmoid sinuses.

The hemispheric portion is made up of the superior semilunar, the inferior semilunar, and the biventral lobules.

The **tentorial** arteriovenous malformations are located on the tentorial surface. The hemispheric part of the tentorial surface comprises the quadrangular, the simple, and the superior semilunar lobules.

The **petrosal arteriovenous malformations** are located on the petrosal surface, the anterior cerebellum that faces the posterior petrous bone. The petrosal surface is formed by the anterior surfaces of the quadrangular, the simple, the semilunar and the biventral lobules and the flocculus.

The **vermian** arteriovenous malformations are positioned on the midline, and they might be located either on the tentorial surface or on the suboccipital surface. The tentorial part of the vermian surface includes the culmen, the declive, and the folium. The suboccipital part of the vermian surface includes the tuber, the pyramid, the uvula, and the nodulus.

The **tonsillar** arteriovenous malformations are located in the tonsils, they being positioned on the inferomedial aspect of the cerebellar hemispheres that attach superolaterally to the cerebellum through the tonsillar peduncles.

The treatment of these lesions is limited to surgery, endovascular embolization, and stereotactic radiosurgery, especially in the cases which are deemed to have a too high risk for an invasive intervention.

Currently there are no specific medical therapies. Some neurosurgeons had expressed dissatisfaction with the Spetzler-Martin grading scale as a tool for the selection in view of the surgical treatment of the patients with cerebellar arteriovenous malformations because the deep nuclei are the only eloquent structures in the cerebellum, and the galenic venous drainage is not a good indicator of the depth of the cerebellar AVMs (Fine et al., 2007).

The arteriovenous malformations are treated through other modalities besides surgery, such as embolization and radiosurgery, as well as using multimodality strategies.

The classification schemes which are used to predict the surgical risk offer no information about the risks of these alternative therapies (Davies, 2012).

Inoue et al. (1995) had identified the size, the morphology, and the hemodynamics as the factors which are the most predictive for the radiosurgical outcome.

Pollock and Flickinger (2002) had developed a more quantitative classification scheme for radiosurgery.

The endovascular embolization had been for a long time a surgical adjunct rather than a curative therapy.

For this reason, there had been few authors who had attempted to develop a classification scheme for the endovascular treatment.

Vinuela et al. (1995) had developed a classification based on the dimensions of the arteriovenous malformation, the number of feeding arteries, and the presence of the pial or of the perforating feeding arteries.

Lawton et al. (2010) had envisioned a grading system which would supplement rather than replace the already entrenched Spetzler-Martin grading system.

However, the supplementary grading system for the brain arteriovenous malformations had been introduced in 2010 as a tool for the improvement of the preoperative risk prediction and for a more adequate selection of the patients for surgery.

### Table 1

Comparison between the Spetzler-Martin and the supplementary grading systems (Kim et al., 2015)

<table>
<thead>
<tr>
<th>The Spetzler-Martin grading scale</th>
<th>Points</th>
<th>The supplementary grading scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size (cm)</td>
<td>1</td>
<td>Age (years)</td>
</tr>
<tr>
<td>≤ 3</td>
<td>1</td>
<td>≤ 20</td>
</tr>
<tr>
<td>3–6</td>
<td>2</td>
<td>20–40</td>
</tr>
<tr>
<td>&gt; 6</td>
<td>3</td>
<td>&gt; 40</td>
</tr>
<tr>
<td>Venous drainage</td>
<td>Bleeding</td>
<td></td>
</tr>
<tr>
<td>Superficial</td>
<td>0</td>
<td>Yes</td>
</tr>
<tr>
<td>Deep</td>
<td>1</td>
<td>No</td>
</tr>
<tr>
<td>Eloquent Compactness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>0</td>
<td>Yes</td>
</tr>
<tr>
<td>Yes</td>
<td>1</td>
<td>No</td>
</tr>
<tr>
<td>Total</td>
<td>5</td>
<td></td>
</tr>
</tbody>
</table>
There had been assigned points for the following features of the arteriovenous malformations: the patient age, the bleeding or the hemorrhagic presentation and the AVM compactness, in a manner similar to the Spetzler-Martin grading system (Kim et al., 2015) (Table 1).

The pediatric patients (< 20 years of age) had been assigned 1 point; the adults (20–40 years of age) had been assigned 2 points; and the older patients (> 40 years of age) had been assigned 3 points.

The patients who present with unruptured arteriovenous malformations had been assigned 1 point, while those with ruptured AVMs had been assigned 0 points. The diffuse arteriovenous malformations had been assigned 1 point, while the compact AVMs, 0 points. These points are added together for the supplementary AVM grade which ranges from 1 to 5 (Kim et al., 2015).

This combination of 2 grading systems had proved to be a useful way of simplifying the complex treatment decisions and thus had become an integral part of the assessment process of the arteriovenous malformations. Kim et al. (2015) had demonstrated that the Spetzler-Martin score in association with the supplementary score have a greater predictive accuracy than the Spetzler-Martin grading system alone, and that this supplemented scoring is currently the best method for the estimation of the neurological outcomes after the surgical treatment of the arteriovenous malformations.

However, the cerebellar arteriovenous malformations are more likely than the cerebral AVMs to present with hemorrhage.

The hemorrhagic presentation of the patients with cerebellar arteriovenous malformations is not attributable to the differences in AVM size. The cerebellar arteriovenous malformations are more likely to have deep venous drainage than the cerebral AVMs, and less likely to involve eloquent structures.

The deep perforating blood supply in the cerebral arteriovenous malformations is twice as frequent as in the cerebellar AVMs (Rodriguez-Hernandez et al., 2012). The patients with cerebral AVMs tend to have more favourable outcomes than those of the cerebellar AVMs.

The tests performed by Rodriguez-Hernandez et al. (2012) had demonstrated that the supplementary grading scale for the arteriovenous malformations has a greater predictive accuracy than the Spetzler-Martin scale and that its accuracy is consistent with all the AVMs, including the cerebellar arteriovenous malformations.

The greatest predictive accuracy obtained through the use of the grading systems is achieved by combining the 2 scores in a supplemented Spetzler-Martin score (Rodriguez-Hernandez et al., 2012).

## PATIENTS AND METHODS

Out of the 367 patients with arteriovenous malformations who had been operated by me, 28 (7.6%) had this type of lesions located at the level of the cerebellum.

The patients with arteriovenous malformations located in the midbrain, in the pons and in the medulla oblongata had not been included.

The postoperative follow-up period had an average length of 8 years, with the maximum and the minimum durations of 21 years and 4 years, respectively.

### THE PATIENTS WITH CEREBELLAR ARTERIOVENOUS MALFORMATIONS

There had been 16 women (57%) and 12 men (43%) with an average age of 39 years (range, 16–76 years).

Twenty-one patients (75%) had presented with hemorrhage; 4 patients (14.28%) had presented with headaches or neurological deficits; while 3 patients (10.71%) had incidental, asymptomatic arteriovenous malformations.

The hemorrhagic presentation had been more frequent in women (85%) than in men (15%). None of the patients had presented with seizures.

The average size of the arteriovenous malformations had been of 2 cm (Table 2).

**Table 2**

<table>
<thead>
<tr>
<th>Age, years, average (range)</th>
<th>39 (16–76)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender, n%</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>12 (43)</td>
</tr>
<tr>
<td>Female</td>
<td>16 (57)</td>
</tr>
<tr>
<td>Presenting symptoms</td>
<td></td>
</tr>
<tr>
<td>Hemorrhage</td>
<td>21 (75)</td>
</tr>
<tr>
<td>Headaches or neurological deficits</td>
<td>15 (53.57)</td>
</tr>
<tr>
<td>Incidental</td>
<td>3 (10.71)</td>
</tr>
</tbody>
</table>
Table 2
(continued)

<table>
<thead>
<tr>
<th></th>
<th>Age, years, average (range) 39 (16–76)</th>
<th>Size of the arteriovenous malformation, cm</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0–3 cm</td>
<td>19 (67.85)</td>
</tr>
<tr>
<td></td>
<td>3–6</td>
<td>8 (28.57)</td>
</tr>
<tr>
<td></td>
<td>&gt; 6</td>
<td>1 (3.57)</td>
</tr>
<tr>
<td>The average AVM size, cm</td>
<td>2</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Venous drainage</th>
<th></th>
<th></th>
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</thead>
<tbody>
<tr>
<td>Superficial</td>
<td>12 (42.85)</td>
<td></td>
</tr>
<tr>
<td>Deep</td>
<td>16 (57.14)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Eloquence</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>No</td>
<td>19 (67.85)</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>9 (32.14)</td>
<td></td>
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<table>
<thead>
<tr>
<th>Compactness</th>
<th></th>
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<tbody>
<tr>
<td>Compact</td>
<td>22 (78.57)</td>
<td></td>
</tr>
<tr>
<td>Diffuse</td>
<td>6 (21.42)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Spetzler-Martin grade</th>
<th></th>
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</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>3 (10.71)</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>6 (21.42)</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>12 (42.85)</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>6 (21.42)</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>1 (3.57)</td>
<td></td>
</tr>
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</table>

We had ascertained that the vermian, the tentorial and the tonsillar arteriovenous malformations had been responsible for more than 90% of all the hemorrhages.

Five patients (17.85%) had intracranial aneurysms (3 feeding artery aneurysms and 2 intracranial aneurysms).

Four of the five patients with aneurysms had intracerebral hemorrhages caused by the rupture of the arteriovenous malformations.

Only 1 patient had subarachnoid hemorrhage which was the result of a ruptured aneurysm.

However, in 20 of the 21 patients who had developed hemorrhages in the posterior fossa, the bleeding had originated from a ruptured arteriovenous malformation.

IMAGING

In the absence of a known underlying etiology which could have caused the hemorrhagic incident, the non contrast head CT scan had been the typical initial imaging examination in all the 21 patients with cerebellar hemorrhages.

On this non contrast CT examination, prominent serpentine hyperattenuating structures, representing draining veins, components of the nidus or dilated arterial feeders had suggested the diagnosis of arteriovenous malformation.

However, while the large arteriovenous malformations could be identified easily, the smaller ones were difficult to detect.

The conventional MRI, the susceptibility-weighted imaging, the contrast enhanced time-resolved magnetic resonance angiography (MRA), the time-of-flight magnetic resonance angiography (TOF-MRA), the diffusion tensor imaging (DTI), and the MRI perfusion imaging are just several of the much more sophisticated noninvasive imaging methods which are used for the diagnosis of the arteriovenous malformations.

The digital subtraction angiography (DSA) remains the gold standard for the evaluation of the cerebrovascular diseases, which include the cerebral arteriovenous malformations. This examination had been performed in all the 28 cases we had treated.

The combination of the exceptional spatial (0.2 mm) and temporal resolution (up to 24 frames per second) is unmatched by any other imaging modality (Eddleman et al., 2009).

The digital subtraction angiography is optimal for the evaluation of the angioarchitecture of the arteriovenous malformations, including an improved detection of the associated intranidal aneurysms, of the obstruction of the venous outflow, of the arterial feeder anatomy, and of the venous drainage patterns (Mossa-Basha et al., 2012).

In the setting of a hemorrhage, the nidus might be compressed by the hematoma, thus limiting the detection of any small underlying lesions.

However, this applies to all imaging modalities, including the CT angiography, the magnetic resonance imaging/magnetic resonance angiography, and even the digital subtraction angiography.

In this kind of situations, the investigations should be repeated after 4 to 6 weeks from the moment of the detection of the hematoma.

We present below, in decreasing order of frequency, the locations of the 28 arteriovenous malformations I had operated: hemispheric – 9 (32.14%); vermian – 7 (25%); tentorial – 4 (14.28%); tonsillar – 3 (10.71%); suboccipital – 3 (10.71%); cerebellopontine angle – 2 (7.14%).

However, it is very difficult to establish an accurate delimitation of the referred to arteriovenous malformations because, especially
the ones with dimensions larger than 3 cm, involve more than one region.

However, the arteriovenous malformations which involve the cerebellar hemispheres and the vermis are the most commonly encountered ones because of the large anatomic area represented by these cerebellar structures.

By knowing the localization, we had been able to anticipate the arterial supply and the venous drainage. Therefore, the hemispheric arteriovenous malformations typically receive unilateral arterial supply from the superior cerebellar artery (SCA), the anterior inferior cerebellar artery (AICA), and the posterior inferior cerebellar artery (PICA), unless the lesions are very large, and in such cases the PICA, and rarely the SCA, might be represented bilaterally (Kopitnik et al., 2004; Danaila et al., 2010).

This can help to distinguish the vermic lesions from the hemispheric ones when the arteriovenous malformations have a median location.

When the arteriovenous malformations are located near the cerebellopontine angle cistern, or involve the lateral aspect of the ventricular wall and the middle cerebellar peduncle, they receive a significant arterial supply from the branches of the anterior inferior cerebellar artery.

With the superiorly located hemispheric arteriovenous malformations, the superior cerebellar artery is the dominant arterial supply source (Kopitnik et al., 2004; Danaila et al., 2010). The arteriovenous malformations which are located superiorly within the cerebellar hemisphere typically drain into the petrosal vein laterally and into the galenic venous system superiorly (Kopitnik et al., 2004).

The vermian arteriovenous malformations receive their arterial supply from distal branches of the superior cerebellar artery and of the posterior inferior cerebellar arteries bilaterally. If a vermian arteriovenous malformation has a larger size, or if it reaches the vicinity of the roof of the fourth ventricle, it is typically found a deep arterial feeding from the anterior inferior cerebellar artery.

The deep branches of the anterior inferior cerebellar artery enter the foramen of Luschka, provide blood to the lateral part of the roof of the fourth ventricle, and constitute the deep arterial supply for the larger vermian arteriovenous malformations. The arteriovenous malformations located in the superior vermis, which is composed of folium, declive, culmen, central lobule and lingula, receive blood especially from the superior cerebellar artery.

The arteriovenous malformations located underneath the horizontal fissure, and which involve the tuber, the pyramid, the uvula and the nodulus, receive arterial blood especially from the posterior inferior cerebellar artery.

The arterial blood supply of the vermian arteriovenous malformations is bilateral in most of the cases. The venous drainage of the vermian arteriovenous malformations goes into the galenic venous system through the superior vermian vein and through bridging veins, as well as into the tentorium through the precentral cerebellar vein (Kopitnik et al., 2004).

Due to the fact that the cerebellar tonsils are small, there are rare instances when we can find arteriovenous malformations at their level. However, when there are occurrences of this type of lesions, the tonsillar arteriovenous malformations are usually fed with arterial blood from the posterior inferior cerebellar artery. When the lesions also involve the cerebellar hemispheres, they can have some blood supply from the anterior inferior cerebellar artery. The venous drainage is usually into the inferior vermian veins, or laterally into the sigmoid sinus (Kopitnik et al., 2004).

THE SURGICAL MANAGEMENT

The reaching of the decision to operate the 28 patients with cerebellar arteriovenous malformations had been based on the lesion-specific factors (dimensions, location and angioarchitecture), as well as on the patient-specific factors (age, medical condition and neurological status).

In relation to these major determinants, 4 (14.28%) patients with large hematomas which were compressing the fourth ventricle and the brainstem had been subjected to emergency surgical treatment (Fig. 1).

In order to save the life of the patient, in three of the cases it had been performed initially only the evacuation of the hematoma. In a single patient it had been performed the resection of the hematoma together with that of a small arteriovenous malformation of the inferior vermis.
In 4 patients (14.28%) it had been performed the embolization with polyvinyl alcohol particles, n-butyl cyanoacrylate glue, and Onix, followed by the surgical intervention.

Out of the 28 operated patients, in 5 (17.86%) of them there had been found complex arteriovenous malformations with intranidal aneurysms. In three of these, the arterial blood supply of the arteriovenous malformation had been provided by the posterior inferior cerebellar artery, while in the other two the arterial blood had come from the superior cerebellar artery. In all 5 of them the aneurysms had been located intranidally, and in one patient aged 34 years old, who had been operated for a right cerebellar arteriovenous malformation, the diameter of the aneurysm had been of 25 mm.

In all 5 patients it had been performed the surgical treatment of the aneurysm, as well as that of the arteriovenous malformation.

It should be mentioned that on the arterial feeders of the cerebellar arteriovenous malformations there are frequently found associated aneurysms, especially when the arterial blood supply is provided by the posterior inferior cerebellar artery.

The arteriovenous malformations located at the level of the cerebellar hemispheres receive arterial blood from the superior cerebellar artery and the posterior inferior cerebellar artery, as well as from their tributary arteries.

This arterial blood supply is often unilateral.

The venous drainage is achieved through the lateral hemispheric veins, as well as through the vermian veins.
The surgical approaches I had most frequently used for the microsurgical resection of the arteriovenous malformations of the cerebellar hemispheres had been the suboccipital approach and the retromastoid one.

For the very large arteriovenous malformations (Fig. 2) of the cerebellar hemispheres I had used combinations of the following approaches: the far lateral (for the inferolateral exposure), the extended retrosigmoid (for the superolateral exposure) and the torcular (for the superior exposure).

These are familiar to all neurosurgeons, and they provide a good exposure of the arteriovenous malformations located on the surface of the cerebellar hemispheres, of the vermis, of the cerebellar amygdalae, of the cerebellopontine angle, of the medulla oblongata and of the floor of the fourth ventricle.

The vermian arteriovenous malformations had been located in the vermis or in the adjacent medial portion of the cerebellar hemisphere. These lesions had been approached through a wide suboccipital craniectomy or craniotomy which exposed the midline from C1 to the torcular. In all 7 (25%) cases with this type of arteriovenous malformations the AVMs had been excised in their entirety, and the postoperative evolution had been good and very good.

In four of the patients, the arteriovenous malformation had been located in the anterosuperior region of the vermis (Fig. 3), so that the nidus and its feeding arteries could be controlled adequately.

In the other 3 patients, the arteriovenous malformations had been located in the inferior portion of the vermis (Fig. 4), cu feeding arteries originated from the posterior cerebellar artery. The postoperative evolution of these cases had also been good and very good.
The tentorial arteriovenous malformations (Fig. 5) located on the cerebellar surface of the tentorium and vascularized by branches of the superior cerebellar artery, had been accessed using an infratentorial supracerebellar approach. Whenever the malformation was extended inferiorly, the craniotomy had to be wider in order to also expose the suboccipital surface of the cerebellum.

When the craniotomy exposes the torcular herophili and the transverse sinus, the dural flap elevates the sinuses and widens this plane.

Subsequently, the tentorial surface of the cerebellum is accessed tangentially through infratentorial supracerebellar dissection (Danaila et al., 2010; Rodriguez-Hernandez et al., 2012).

The tentorial arteriovenous malformations are noneloquent, except those which are located anteriorly near the midbrain and the trochlear nerve (Rodriguez-Hernandez et al., 2012).

The tonsillar arteriovenous malformations (Fig. 6) had been resected through standard midline suboccipital craniotomies.
For the surgery of the **suboccipital arteriovenous malformations** located on the suboccipital surface of the cerebellum and vascularized by branches of the posterior inferior cerebellar artery, I had used an approach through a wide suboccipital craniotomy on the midline, with the elevation of the posterior arch of the atlas (Fig. 7).

However, the most commonly used approach is the lateral suboccipital craniotomy.

For the **cerebellopontine arteriovenous malformations** (Fig. 8), I had used either a retromastoid suboccipital approach through extended retrosigmoid craniotomies, or a retromastoid suboccipital one with the exposure of the foramen magnum, of the transverse sinus and of the sigmoid sinus.
Fig. 8. a. The lateral vertebral angiography performed before the operation of a cerebellopontine angle arteriovenous malformation; b. The lateral vertebral angiography after the operation. The postoperative evolution had been very good (surgeon: Leon Danaila).

However, the complete resection of the arteriovenous malformation had been achieved in 27 patients, the results being supported by angiographic confirmation (a surgical obliteration rate of 96.42%).

The resection of all the 27 arteriovenous malformations had been performed in a single-stage surgical treatment.

The patient in whom it had been left in place a small residual malformation component had been subjected to stereotactic radiosurgery.

THE PATIENT OUTCOMES

Generally, the postoperative evolution of the 28 patients with cerebellar arteriovenous malformations operated by me had been very good and good. The neurological deficits which had developed immediately after the surgical intervention in 6 (21.43%) patients had been related to gait and posture disorders, as well as to difficulties in the maintenance of the standing position and the persistence of dysmetria. The referred to deficits had been considered as transient, because they had resolved completely during the follow up period which had an average length of 8 years, with the maximum and the minimum durations of 21 years and 4 years, respectively.

The good and very good results we had obtained are the consequence of the use of very clear criteria for the selection of the patients, of the choosing of the optimal moment for the surgical intervention, and especially of my operative experience in the field of vascular neurosurgery.

The neurological deficits which had been present before the surgical intervention had manifested through astasia-abasia in 4 (14.29%) patients, neocerebellar syndrome in 6 (21.43%), dysarthria in 2 (7.14%) cases, and nystagmus in 3 (10.71%) patients.

All of them had been accompanied by headaches.

The natural history of the untreated arteriovenous malformations includes combined rates of major morbidity and mortality of 2.7% per year (Ondra et al., 1990).

The clinical outcomes are dependent on the site, the type and the size of the malformation (Santos et al., 2009; Reishofer et al., 2012).

Therefore, the cerebellar arteriovenous malformations are vascular lesions with a direct, abnormal connection between arteries and veins. The connection between the feeding arteries and the draining veins is referred to as a nidus, which is the nest of vessels without any interposition of brain parenchyma (Fleetwood and Steinberg, 2002; Danaila, 2002; Danaila, et al., 2010; Di Ieva et al., 2014).

The nidus is a complex vascular network formed by a tangle of coiled and tortuously enlarged vessels (Essing et al., 1996; Reishofer et al., 2012; Danaila, 2012).

DISCUSSION

Out of the 28 cerebellar arteriovenous malformations operated by us, the hemorrhage had been present before the surgical intervention in 21 patients (75%). It had been more frequent in women than in men.

According to Oliveira et al. (1998), Lawton et al. (2005), Munoz et al. (2007), Da Costa et al. (2009), Danaila et al. (2010) and Rodriguez-Hernandez et al. (2012), the hemorrhages cause preoperative neurological deficits.

The explanation for the increased bleeding from the cerebellar arteriovenous malformations is still unclear (Fleetwood and Steinberg, 2002; Stapf et al., 2006).
The common clinical manifestations of the cerebellar arteriovenous malformations had included hemorrhage, headache and progressive neurological deficits.

One important difference between the cerebellar and the cerebral arteriovenous malformations had been that the cerebellar AVMs did not present with seizures.

The main cause of mortality and of persistent morbidity is represented by hemorrhage, which is present in 30% to 89% of the patients with cerebral arteriovenous malformations (Stefani et al., 2002; Danaila, 2012).

The deep venous drainage is associated with an increased risk of rupture, and it had been found to be significantly increased in our cerebellar arteriovenous malformations.

In the patient presented in Fig. 2 we had big problems in terms of hemostasis and the loss of a very large amount of blood. In this case we had purposefully left behind several perinidal blood vessels in order not to compromise the neurological condition. For this reason, we had intervened in his case using the Gamma Knife. Nevertheless, the postoperative evolution had been excellent.

However, the increased hemorrhagic behavior of the cerebellar arteriovenous malformations supports a more aggressive management approach. (Batjer and Samson, 1986; de Oliveira et al., 1998; Halim et al., 2004; da Costa et al., 2009; Arnaout et al., 2009; Danaila et al., 2010; Rodriguez-Hernandez et al., 2012).

For this reason, Sanchez-Mejia et al. (2009) and Rodriguez-Hernandez (2012) recommend the resection and the radiosurgical treatment of the ruptured or unruptured cerebellar arteriovenous malformations, even for the patients with high Spetzler-Martin grade AVMs.

Alternatively, Lawton (2003) and Lawton et al. (2010) do not recommend this treatment for the patients with unruptured arteriovenous malformations, particularly for those with high grade AVMs and with epilepsy.

**Aberrant angiogenic characteristics of the arteriovenous malformations.** Generally, the causes which lead to their formation are represented by angiogenesis abnormalities.

Jabbour et al. (2009) had showed that the endothelial cells derived from the cerebral arteriovenous malformations produce angiogenic factors, such as the vascular endothelial growth factor (VEGF) and the endothelin-1 (ET-1), which are not normally produced by the quiescent brain vasculature.

Furthermore, these endothelial cells are highly activated and exhibit functional abnormalities in terms of migration and tubule formation which are consistent with the cell status in active angiogenesis (Jabbour et al., 2009).

Jabbour et al. (2009) had harvested tissues from 6 patients who had undergone the surgical excision of their arteriovenous malformations.

Control endothelial cells had been derived from the surgical specimens of 3 patients who had undergone temporal lobectomies for the treatment of epilepsy associated with mesial temporal sclerosis.

The brain endothelial cells isolated at the level of the arteriovenous malformation and the control brain endothelial cells had been evaluated immunohistochemically for the expression of the endothelial cell markers CD31 and von Willebrand factor, as well as of the angiogenic factors including the vascular endothelial growth factor A, the interleukin-8 and the endothelin-1.

The vascular endothelial growth factor receptors 1 and 2 had also been evaluated using immunohistochemistry techniques. The functional assays had evaluated the cell proliferation, the cytokine production, the tubule formation and the cell migration using the modified Boyden chamber method.

The endothelial cells derived from the cerebral arteriovenous malformations are highly activated cells that overexpress proangiogenic growth factors and exhibit abnormal functions, which are consistent with the profile of highly activated endothelial cells.

The vascular endothelial growth factor is a critical mediator of angiogenesis (Carmeliet, 2005). This growth factor stimulates different functions of the endothelial cells, including differentiation, survival, proliferation, and migration (Drake et al., 2000; Zadeh and Guha, 2003).

**The grading of the arteriovenous malformations.** Rodriguez-Hernandez et al., (2012) had showed that the Spetzler-Martin grading system seems to be less satisfactory than the supplementary grading system for the prediction of the patient outcomes after the resection of the cerebellar arteriovenous malformations.

The current range of grading systems is generally useful, but imperfect and evolving. For
this reason, Kim et al. (2015) had demonstrated the predictive accuracy of the supplemented Spetzler-Martin system in a large multicenter cohort, thus validating the use of this evaluation to establish the operability of the arteriovenous malformations. Kim et al. (2015) offered this supplementary grading system as just another tool to guide the process of analyzing some of the critical factors that influence the patient outcomes, in order to help the neurosurgeons to make more rational choices when weighing the known risk of spontaneous AVM rupture against the risks of intervention (Kim et al., 2015).

The supplemented grading system is currently the best method of predicting the neurological outcomes after the surgical treatment of an arteriovenous malformation, and Kim et al. (2015) had recommended it as a starting point in the evaluation of the AVM operability.

The cerebellar arteriovenous malformation types differ depending on their location. The goals of the treatment of an arteriovenous malformation include the alleviation of the mass effect and of the intracranial hypertension associated with the rupture, the prevention of future rupturing, the alleviation of the arterial steal phenomenon and the improvement of the associated epilepsy.

In the following pages I shall present several considerations about the 6 types of cerebellar arteriovenous malformations (hemispheric, vermian, tentorial, tonsillar, suboccipital and of the cerebellopontine angle) I had operated.

Although the arteriovenous malformations of the cerebellar hemispheres are the most surgically accessible ones, they usually bleed excessively during the resection procedure because of the fact that they have arterial afferences from almost all the arteries of the posterior fossa.

Therefore, the hemispheric cerebellar arteriovenous malformations are supplied by distal cortical branches from the superior cerebellar artery superiorly, from the posterior inferior cerebellar artery inferiorly and from the anterior inferior cerebellar artery laterally. The cortical feeders are occluded using a circumscribing incision around the arteriovenous malformation.

The deep perforating arteries are encountered along the deep plane of the nidus. The draining veins include the superior cerebellar veins, which drain the superior region of the cerebellum and empty into the straight sinus, the great cerebral vein of Galen and the transverse sinus and the superior petrosal sinus, as well as the inferior cerebellar veins that drain the inferior aspect of the cerebellum and empty into the occipital sinus, the superior petrosal sinus and the transverse sinus.

In some cases, the drainage veins travel into the contralateral hemisphere, or anteriorly into the great cerebral vein of Galen.

According to Kopitnik et al. (2004) and Danaila et al. (2010), the optimal approach for the surgical resection of the arteriovenous malformations which involve the cerebellar hemispheres depends on the size of the malformation, on the need to expose the cerebellopontine angle cistern, and on the preference of the surgeon.

We place most of the patients in the supine or lateral position for the resection of the arteriovenous malformations which involve the lateral areas of the cerebellar hemispheres.

After the exposure of a cerebellar hemispheric arteriovenous malformation, the microsurgical dissection is focused on the identification and the sectioning of the main arterial feeding vessels of the AVM originated from the superior cerebellar artery, the anterior inferior cerebellar artery and the posterior inferior cerebellar artery.

The dissection of the superior cerebellar artery branches can be difficult because they are often located in the vicinity of the venous structures of the arteriovenous malformation and are found during the dissection of the superior pial margin of the lesion (Kopitnik et al., 2004; Danaila et al., 2010).

The arterial feeders from the anterior inferior cerebellar artery which supply the arteriovenous malformation are identified in the cerebellopontine angle cistern, as the branches travel over the flocculus or enter the foramen of Luschka in order to supply the roof of the fourth ventricle. The branches from the posterior inferior cerebellar artery are identified adjacent to the cerebellar tonsil, and are followed distally to their entry into the arteriovenous malformation.

After the main arterial feeders are coagulated with the bipolar forceps adjacent to the nidus, the lesion is circumferentially dissected and elevated out in the direction of the venous drainage into the petrosal or the galenic systems (Kopitnik et al., 2004).

The vermian arteriovenous malformations are located on the midline, and they can be located either on the tentorial surface or on the hemispheric surface of the vermis.
The cerebellar arteriovenous malformations

The vermis is subdivided into a superior and an inferior portion. The superior portion is visible between the two hemispheres, while its inferior portion is buried between the two hemispheres. The inferior vermis forms the posterior cortical surface within the posterior cerebellar incisure, which also contains the falx cerebelli (Rodriguez-Hernandez et al., 2012). The tentorial part of the vermian surface includes the culmen, the declive and the folium, while the inferior vermis includes the tuber, the pyramid, the uvula and the nodulus.

According to Rodriguez-Hernandez et al. (2012), the small arteriovenous malformations located at the apex of the vermis can be approached with the patient in the sitting position. The superior vermian arteriovenous malformations often have a bilateral arterial blood supply through feeding arteries from the superior cerebellar arteries, while the inferior AVMs can be supplied by branches from both posterior inferior cerebellar arteries. The superior vermian arteriovenous malformations are 9 times more frequent than the inferior ones (90% and 10% respectively) (Rodriguez-Hernandez et al., 2012).

The lesions of the cerebellar vermis are best resected using a midline exposure. I had used, like many other neurosurgeons, a midline incision from above the inion up to the spinous processes of the C3-C4 vertebrae.

For the caudal lesions of the vermis, the bony opening is extended inferiorly to include the opening of the foramen magnum.

For the caudally positioned arteriovenous malformations, we had removed the foramen magnum and the posterior arch of the first cervical vertebra. For the arteriovenous malformations which involve the superior aspect of the vermis we, like Kopitnik et al. (2004), had exposed the transverse sinus and the torcular herophili.

After this, we had performed a wide stellate durotomy to maximize the exposure, in order to allow the opening of the arachnoid overlying the cisterna magna and the evacuation of the cerebrospinal fluid from the fourth ventricle to provide brain relaxation.

Subsequently, we had performed the microdissection of the posterior inferior cerebellar artery up to its entrance into the malformation. The feeding vessel from the posterior inferior cerebellar artery had been coagulated using the bipolar forceps as near to the nidus as possible. After the elimination of the main PICA feeding source, we had begun the circumferential dissection of the pial margin of the arteriovenous malformation in an inferior to superior direction. This had eliminated the superficial feeders from the anterior inferior cerebellar artery coming from the cerebellopontine angle cistern (Kopitnik et al., 2004). Attention had been paid to the feeders from the superior cerebellar artery because they are often closely associated with the superiorly directed venous drainage of the arteriovenous malformations (Kopitnik et al., 2004; Danaila et al., 2010).

Whenever there is encountered the persistence of the turbidity of the malformation after the transectioning of the arterial feeders from the posterior inferior cerebellar artery and the superior cerebellar artery, this is due to the deep arterial feeders from the superior cerebellar artery or the anterior inferior cerebral artery. When the vermian arteriovenous malformation is located in the proximity of the superior medullary velum or of the ceiling of the fourth ventricle, there should be coagulated and sectioned the deep branches of the anterior inferior cerebral artery. Sometimes it is necessary to open the ceiling of the fourth ventricle because the vermian arteriovenous malformations might also involve the ventricular surface.

After the bipolar coagulation of all the arterial pedicles, and subsequently that of the drainage veins, the nidus can be removed.

The tentorial arteriovenous malformations are exposed using a supracerebellar approach, after the removal of the arachnoid adhesions between the posterior border of the tentorial surface and the cerebellum.

The drainage veins on the tentorial surface are initially preserved. Then it is performed the coagulation of the arterial pedicles from the distal portion of the superior cerebellar artery using the bipolar forceps. The drainage veins on the tentorial surface are coagulated in the last stage of the operation.

According to Rodriguez-Hernandez et al. (2012), the venous drainage is typically through the superior veins which travel superficially towards the tentorium / tentorial sinus, the transverse sinus or the torcular herophili, or it has a deep trajectory towards the great cerebral vein of Galen.

The posterior and the inferior borders of the arteriovenous malformation are rendered evident after the incision of the cerebellar cortex and after
the evacuation of the eventually present hemispheric hematoma.

The tonsillar arteriovenous malformations are found in a relatively small region within the posterior fossa, and they are typically less frequent and the lesions are limited in size.

The tonsils of the cerebellum are much larger than the other lobules in the posterior lobe.

Under the action of an increased intracranial pressure, the cerebellar tonsils might protrude into the foramen magnum (cerebellar tonsillar herniation) and cause the compression of the medulla oblongata, which results in cardiovascular and respiratory depression.

These arteriovenous malformations of the cerebellar tonsils are supplied with arterial blood by the ipsilateral posterior inferior cerebellar artery.

The tonsillo-hemispheric lesions receive their arterial blood supply from both posterior inferior cerebellar arteries and from the anterior inferior cerebellar artery.

The venous drainage empties through the tonsillar veins into the inferior vermian system, as well as into the occipital, the superior petrosal and the sigmoid sinuses.

The cerebellar tonsil resection is relatively straightforward and uncomplicated, through a C-shape skin incision and a standard paramedian suboccipital craniotomy.

After the opening of the dura mater and the evacuation of the cerebrospinal fluid (CSF) from the cisterna magna, the subarachnoid dissection includes the widening of the vallecula to separate the tonsils, the opening of the cerebellomedullary fissure, and the splitting of the tonsillobiventral fissure.

Subsequently, it is performed the neurosurgical dissection in the cerebellopontine angle cistern in order to locate and control the posterior inferior cerebellar artery as it courses around the medulla.

The upward retraction of the inferior pole of the tonsillar arteriovenous malformation exposes the posterior inferior cerebellar artery as it approaches the lesion.

The PICA is then coagulated and sectioned as it enters the nidus of the arteriovenous malformation.

The distal en passage arteries are preserved.

The complete removal of the arteriovenous malformation removal can be accomplished by the simple resection or amputation of the involved cerebellar tonsil (Kopitnik et al., 2004; Danaila et al., 2010). The tonsillar arteriovenous malformations are located usually in non-eloquent areas, but they are adjacent to the medulla oblongata and the glossopharyngeal, the vagus, and the spinal accessory nerves (Danaila et al., 2010; Rodriguez-Hernandez et al., 2012).

The suboccipital arteriovenous malformations are most often approached through a lateral suboccipital craniotomy. For the large lesions there are used combined approaches, with the far-lateral approach adding to the inferolateral exposure, as well as the extended retrosigmoid approach, and the torcular approach.

They are located in the inferior portion of the major fissure. The minor fissures on the suboccipital surface include the horizontal fissure between the superior and the inferior semilunar lobules, the prebiventral fissure located between the inferior semilunar and the biventral lobules, and the tonsilllobiventral fissure.

The arteriovenous malformations of the cerebellopontine angle, which are located on the petrosal surface of the cerebellum, are exposed using a craniotomy centered about 1 cm behind the mastoid process, followed by the exposure of the sigmoid sinus from the transverse-sigmoid junction to the jugular bulb.

After the durotomy, the attention should be directed to the brain stem auditory responses that will act as an indication of the impact of the cerebellar retraction on the hearing function, which can be lost easily by direct medial retraction.

A special attention should also be paid to the subarachnoid dissection because of the fact that the referred to plane greatly facilitates the subsequent resection of the arteriovenous malformation and the preservation of the neural structures such as the trigeminal nerve (superiorly) and the ninth, tenth and eleventh cranial nerves (inferiorly).

With the involvement of the flocculonodular lobe and the closely adjacent ninth, tenth, and eleventh cranial nerves, the surgeon should acknowledge the fact that the resection in this area could lead to significant postoperative gait deficits, this being a function of the lower cranial nerves (Peerless et al., 1996).

A wide lateral bony exposure is necessary in order to gain proximal control of the vertebral artery and of the posterior inferior cerebellar artery, and to allow the precise identification and protection of both the cranial nerves and the vascular branches to the brain stem (Peerless et al., 1996).
The main feeder of these cerebellopontine arteriovenous malformations is the anterior inferior cerebellar artery, which is accompanied in some instances by feeders from the superior cerebellar artery and the posterior inferior cerebellar artery.

In the hemorrhagic cases it must be performed an incision in the cerebellar cortex lateral to the arteriovenous malformation, the excision of the respective lobule and the evacuation of the hematomas.

After the resection of all the arterial feeders, the draining veins (the anterior hemispheric veins, the vein of the cerebellopontine fissure which empties into the petrosal vein, and the superior petrosal sinus) should be preserved with great care and be the last ones to be coagulated.

However, the resection of such malformations should be performed only by neurosurgeons who, besides a vast experience, also have a great talent.

CONCLUSIONS

The population-based studies had revealed that between 35% and 50% of the patients with arteriovenous malformations initially present with hemorrhage, while the annual risk of further hemorrhagic episodes might vary from 1% to 18% (Brown et al., 1996; Mast et al., 1997; Hillman, 2001; Stapf et al., 2002; ApSimon et al., 2002; Al-Shahi et al., 2003; Stapf et al., 2003).

The cerebellar arteriovenous malformations are much more likely to bleed than the cerebral AVMs, reason for which they should be treated more aggressively.

Kim et al. (2015) had demonstrated the predictive accuracy of the supplemented Spetzler-Martin grading system in a large, multicenter cohort study, thus validating the use of the supplemented Spetzler-Martin scores.

Jabbour et al. (2009) had concluded that the endothelial cells derived from the cerebral arteriovenous malformations are highly activated cells which overexpress proangiogenic growth factors and exhibit abnormal functions, which are consistent with the profile of highly activated endothelial cells.

REFERENCES


