Posterior fossa arteriovenous malformations

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Arteriovenous malformations (AVMs) of the posterior fossa are complex neurovascular lesions that are less common than their supratentorial counterparts, accounting for <15% of all AVMs. The majority of patients with these lesions present with intracranial hemorrhage, a factor that has been consistently shown to increase one’s risk for subsequent bleeding. Studies have additionally shown a posterior fossa or deep AVM location to portend a more aggressive natural history. The authors reviewed the literature on posterior fossa AVMs, finding their annual rupture rates to be as high as 11.6%, an important factor that underscores the importance of aggressive treatment of lesions amenable to intervention as therapeutic options and results continue to improve. (DOI: 10.3171/2009.2.FOCUS0914)

Key Words • arteriovenous malformation • infratentorial location • posterior fossa • natural history

Although infratentorial AVMs comprise only 7–15% of intracranial AVMs,1,2,8,10,26,38 accumulating data have demonstrated an independent association of infratentorial AVM location and hemorrhagic presentation.19,26,38 This is alarming in light of the considerably greater morbidity and mortality associated with posterior fossa AVM rupture.12,18 Fortunately, with accumulating surgical experience and the cultivation of multimodality AVM therapy, therapeutic success continues to improve.18,21,25 This is particularly crucial in light of the growing body of modern literature pertaining to AVMs in which authors have detailed the aggressive natural history of these lesions.12,15,18,24,40

Posterior Fossa AVMs

Clinical Presentation

Unlike their supratentorial counterparts, infratentorial AVMs are more likely to present with hemorrhage and rarely present with seizures.2,4,8,11,18,26,27,29,34,36 With 6 of the larger surgical series combined, 207 (84%) of 246 patients harboring infratentorial AVMs presented with hemorrhage (Table 1). In the original Cooperative Study,27 7 (27%) of 26 patients died as a result of their presenting hemorrhage. Fults and Kelly22 noted that less than half of their patients with posterior fossa AVMs survived an initial hemorrhage, and hemorrhagic mortality rates of up to 66.7% have been reported for posterior fossa AVM rupture.34

In addition to the aforementioned surgical series, numerous recent observational studies have also demonstrated the greater rate of hemorrhagic presentation among patients with these lesions,11,17,28,32,39 with only a few exceptions.9,20,22 In a multivariate analysis targeted at elucidating the relationship between infratentorial location and hemorrhagic presentation, Khaw et al.19 demonstrated that infratentorial AVMs were almost twice as likely to present with hemorrhage as supratentorial AVMs. Similarly, in the Toronto Study group32 21 (60%) of 35 patients with infratentorial AVMs presented with hemorrhage, compared with 125 (35%) of 355 patients with supratentorial AVMs and 88 (29%) of 299 patients with superficial AVMs. Kader et al.37 reported similar findings in which 36 (86%) of 42 patients with infratentorial AVMs presented with hemorrhage compared with 221 (54%) of 407 with supratentorial lesions.

Progressive neurological deficits (including those secondary to mass effect, ischemia, and hydrocephalus) were the second most common mode of presentation, seen in up to 28% of patients (Table 1). With respect to

Abbreviations used in this paper: AVM = arteriovenous malformation; mRS = modified Rankin Scale.
asymptomatic posterior fossa AVMs, these reports are likely to underestimate the true prevalence as individuals in this subset may not present for clinical evaluation. However, the incidence of asymptomatic posterior fossa AVMs may rise in the future with the increasing use of advanced neuroimaging modalities for nonspecific symptomatology.

### Risk Factors for Hemorrhage

Numerous studies on posterior fossa AVMs have demonstrated a correlation between the incidence of hemorrhage and an increased risk of subsequent rupture (Table 2) with few exceptions. This phenomenon is perhaps most dramatically illustrated in the report by Mast et al., in which annual hemorrhage rates of lesions that had previously bled were as high as 17.8%. The authors concluded that hemorrhagic presentation was the most important feature influencing subsequent hemorrhage rates. This was reinforced by Stapf et al. who, in a multivariate model, reported annual rehemorrhage rates ranging from 4.5 to 34.3%, depending on lesion location and deep venous drainage. In their prospective analysis of 678 patients, da Costa et al. found that hemorrhage on presentation was an independent predictor of future bleeding.

Interestingly, a proclivity toward hemorrhage clustering is underscored by the 6–15% annual rehemorrhage rates reported across the 1st year after hemorrhage in several studies. Furthermore, recent studies of Halim et al., Yamada et al., da Costa et al., and Hernesniemi et al. have shown relatively elevated annual rehemorrhage rates of 5–6% up to 5 years after an initial hemorrhagic presentation.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Posterior Fossa AVMs</th>
<th>Hemorrhagic Presentation (%)</th>
<th>Other Presentation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perret et al., 1996</td>
<td>32</td>
<td>26 (81)</td>
<td>4 (13) symptomatic, 2 (6) incidental</td>
</tr>
<tr>
<td>Batjer et al., 1986</td>
<td>32</td>
<td>23 (72)</td>
<td>9 (28) progressive deficits</td>
</tr>
<tr>
<td>Drake et al., 1986</td>
<td>66</td>
<td>61 (92)</td>
<td>3 (5) headache, 2 (3) focal deficit</td>
</tr>
<tr>
<td>Solomon et al., 1986</td>
<td>12</td>
<td>11 (92)</td>
<td>1 (8) “gradual onset of symptoms”</td>
</tr>
<tr>
<td>Symon et al., 1995</td>
<td>28</td>
<td>24 (85.7)</td>
<td>4 (14) progressive deficits, 2 (7) occipital/retroauricular pain, 1 (4) headache only</td>
</tr>
<tr>
<td>Kelly et al., 2008**</td>
<td>76</td>
<td>62 (82)</td>
<td>9 (19) cranial nerve palsy, 8 (17) headache, 1 (2) hemiparesis, 11 (23) ataxia</td>
</tr>
</tbody>
</table>

* Modes of presentation depicted for 48 patients with Spetzler-Martin Grade III–V AVMs.
Posterior fossa AVMs

venous drainage seen with posterior fossa AVMs may also contribute by increasing the pressure gradient in the AVM nidus. Vinuela et al.37 have described the possibility of hemodynamic changes resulting from the convergence of draining veins into the vein of Galen and the straight sinus as contributory factors for increased rupture rates. Finally, Willinsky et al.39 have proposed that venous “kinking” at the level of the tentorium, producing a state of venous outflow stenosis which has been previously associated with increased rupture risk,3,6,14 may also be a contributing factor.

Other factors that may modify the risk of rupture of these lesions have been studied, but there is currently little agreement in the literature regarding the significance of their contribution. For instance, whereas several authors have noted small nidus size as a risk factor for rupture,17,20 others have found that larger AVMs tended to bleed more frequently.15,24,33 Other factors thought to be associated with rupture risk include age,5,11,30,40 sex,23,40 and venous drainage patterns20,22,24 for which the data were similarly inconsistent.

### Treatment and Outcomes

Accumulating experience with the treatment of posterior fossa AVMs and the incorporation of multimodality approaches, including radiosurgery, endovascular therapy, and improved microsurgical techniques, have significantly contributed to continuously improving outcomes (Table 4).

Their classic series detailing surgical outcomes in the treatment of posterior fossa AVMs, Drake et al.8 reported a 92% obliteration rate with 71% of patients having an “excellent to good outcome.” Batjer and Samson2 reported a 100% obliteration rate with 80% of patients experiencing “excellent to good” outcomes in their experience with 30 posterior fossa AVMs. Kelly et al.18 have also described their experience with treating AVMs located in the posterior fossa, although the analysis included only the 48 lesions that were Spetzler-Martin Grades III–V and the rate of angiographic obliteration was 52%, whereas 81% of patients had “excellent to good” outcomes. Finally, in their series of 98 posterior fossa AVMs,
da Costa et al.7 reported “good” mRS outcomes in 67% of cases whereas in 28% of the cases patients suffered from “poor” outcomes but also presented that way, implying that the final outcome was related to the mRS score on initial presentation.

Conclusions

Infratentorial AVMs are complex neurovascular lesions that, as opposed to their supratentorial counterparts, pose an increased risk for hemorrhagic presentation as well as increased morbidity and mortality due to their presence in the narrow confines of the posterior fossa and in proximity to many critical structures including the brainstem, cranial nerves, and small perforator vessels. An intimate understanding of the natural behavior of these lesions is paramount to allow one to develop a timely management plan. Although the risks of treatment are considerable, outcomes are reassuring. It is clear that intracranial AVMs comprise a heterogeneous group of vascular lesions with variable natural history; posterior fossa AVMs merit multimodality intervention when feasible in most cases because of their higher risk of rupture and higher potential for morbidity and mortality.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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**TABLE 3: Studies reviewing infratentorial and/or deep AVM location as a risk factor for hemorrhage**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients, No. Infratentorial, Study Type</th>
<th>Mean Follow-Up</th>
<th>Location as a Risk Factor (deep &amp;/or infratentorial)</th>
<th>Annual Infratentorial Hemorrhage Rates</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fults et al., 1984</td>
<td>131, 10 infratentorial, retrospective</td>
<td>8.7 yrs</td>
<td>yes (infratentorial)</td>
<td></td>
</tr>
<tr>
<td>Pollock et al., 1996</td>
<td>315, 97 deep,* retrospective</td>
<td>10,939 patient-ys</td>
<td>yes (deep on univariate)</td>
<td></td>
</tr>
<tr>
<td>Mine et al., 2000</td>
<td>55, 16 deep,† retrospective</td>
<td>10.5 yrs</td>
<td>yes (deep/infratentorial)</td>
<td>5 yrs: 7.5%; 10 yrs: 5.0%</td>
</tr>
<tr>
<td>Stefani et al., 2002</td>
<td>390, 35 infratentorial, prospective</td>
<td>10.5 yrs</td>
<td>yes (deep/infratentorial)</td>
<td></td>
</tr>
<tr>
<td>Stapf et al., 2006</td>
<td>622, 74 infratentorial, prospective</td>
<td>3.1 yrs</td>
<td>no (infratentorial)</td>
<td></td>
</tr>
<tr>
<td>Yamada et al., 2007</td>
<td>305, 105 deep,‡ mixed</td>
<td>892 patient-ys</td>
<td>yes (deep/infratentorial)</td>
<td>if prior hem: 11.4%; if no hem: 4.4%</td>
</tr>
<tr>
<td>da Costa et al., 2008</td>
<td>678, 85 deep,§ prospective</td>
<td>2.9 yrs</td>
<td>no (infratentorial)</td>
<td></td>
</tr>
<tr>
<td>Hernesniemi et al., 2008</td>
<td>238, 18 infratentorial retrospective</td>
<td>13.5 yrs</td>
<td>yes (infratentorial)</td>
<td>overall: 6.7%; 5 yrs: 11.6%</td>
</tr>
</tbody>
</table>

* Thalamus, basal ganglia, corpus callosum, or brainstem.
† Thalamus, basal ganglia, brain stem or cerebellum.
‡ Insular, thalamus, basal ganglia, corpus callosum, brain stem, or cerebellum.
§ Deep white matter tracts, basal ganglia and thalamus, peri-ventricular regions, or posterior fossa.

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