Association between posterior fossa arteriovenous malformations and prenidial aneurysm rupture: potential impact on management

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Object. In patients with posterior fossa arteriovenous malformations (AVMs) who present with hemorrhage, feeding artery aneurysms are often the source of bleeding. The aim of this study was to evaluate the relative proportions of cases of infra- and supratentorial AVMs in which patients presented with prenidial aneurysm rupture. The management and outcome of 9 cases of posterior fossa AVMs associated with prenidial aneurysm rupture are presented.

Methods. The authors retrospectively reviewed 233 consecutive AVM cases involving patients treated at their institution between April 2001 and August 2012. Patients with a prenidial aneurysm as the cause of the hemorrhage were identified. The frequencies of prenidial aneurysm-related ruptures were compared in cases of supra- and infratentorial AVMs. Management and clinical outcome (modified Rankin Scale [mRS] score) of patients with posterior fossa AVMs treated for ruptured prenidial aneurysms were recorded.

Results. Of 233 AVMs, 25 (11%) were in the posterior fossa, and in 22 (88%) of these cases, the patients presented with hemorrhage, including 9 patients (41%) who presented with hemorrhage due to prenidial aneurysm rupture. Of 208 patients with supratentorial AVMs, 107 (51%) presented with hemorrhage, including 5 patients (4.7%) in whom the hemorrhage was associated with a prenidial aneurysm ($p < 0.01$). All 9 patients with posterior fossa AVMs and prenidial aneurysm rupture were treated with early embolization of the offending aneurysm. There was no early rebleeding or clinical complication related to this approach. At the end of follow-up (mean 46.8 months), 2 patients had an mRS score of 0, 1 had a score of 1, 3 had a score of 2, 2 patients were dead (mRS score of 6), and 1 patient was lost to follow-up.

Conclusions. Posterior fossa AVM hemorrhages are frequently associated with prenidial arterial aneurysms. Urgent endovascular treatment of the aneurysm was effective in this case series.
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KEY WORDS • arteriovenous malformation • cerebral AVM • embolization •
prenidial aneurysm • posterior fossa AVM

RUPTURED cerebral arteriovenous malformations (AVMs) are associated with significant morbidity and mortality.^{1,3,4} Up to 18% of patients with bleeding AVMs harbor associated arterial aneurysms, approximately half of which occur on feeding arteries (prenidial aneurysms).⁵ Numerous reports have suggested a correlation between AVM-related aneurysms and hemorrhage risk.^{9–11} Some authors have previously noticed that prenidial aneurysm ruptures were frequent when AVMs were located in the posterior fossa.^{2,8,12} We sought to confirm, in a series of cases involving patients with AVMs who were treated at our institution, that patients with posterior fossa AVMs frequently present with hemorrhage associated with prenidial aneurysm ruptures. We also aimed to review the outcome for patients presenting with such ruptures who were treated by endovascular means.

Abbreviations used in this paper: AVM = arteriovenous malformation; mRS = modified Rankin Scale; PICA = posterior inferior cerebellar artery.

Methods

We conducted a retrospective chart and imaging review of all consecutive cases of AVMs in which patients were treated by means of endovascular techniques in our institution between April 2001 and August 2012. This study was approved by our local ethics committee. Patients were included if their neuroradiology reports (angiography and CT) mentioned hemorrhage and the presence of a prenidial aneurysm. Neuroimaging studies were reviewed by a senior neuroradiologist (D.R.). By definition, hemorrhage secondary to the rupture of a prenidial aneurysm was diagnosed when the hematoma and/or the epicenter of subarachnoid hemorrhage was clearly remote from the AVM nidus but in the vicinity of the aneurysm (Fig. 1). Cases in which hemorrhage could be related to an aneurysm located at the circle of Willis or at usual locations in the posterior circulation were excluded. Using this definition, we recorded posterior fossa AVMs that bled from prenidial aneurysms and looked for cases in which

FIG. 1. **A:** CT scan showing inferior vermis hematoma with blood inside the fourth ventricle. **B:** CT angiogram (sagittal view) showing AVM feeding artery aneurysm in the epicenter of a hematoma (long arrow) with the AVM nidus (short arrow) clearly remote from the hematoma. **C:** Left vertebral artery angiogram, lateral view, confirming the CT angiography findings. **D:** Superselective injection of the PICA branch harboring the aneurysm. **E:** Left vertebral injection after glue embolization of the aneurysm and parent vessel.

the patients had a similar presentation but had supratentorial AVMs. Proportions were compared using the Z-test. Finally, we report the outcome (modified Rankin Scale [mRS] score) for patients with posterior fossa AVMs presenting with prenidial aneurysm rupture and treated with endovascular embolization.

Results

Presentation and Incidence of Prenidial Aneurysm Rupture

We identified 233 cases of AVMs involving patients treated between April 2001 and August 2012 in our insti-

tution. The patients' mean age was 44 years (range 11–80 years). Of the 233 AVMs, 208 were supratentorial (89%), and 25 (11%) were located in the posterior fossa. Hemorrhage occurred in 107 (51.4%) of the supratentorial AVMs compared with 22 (88%) of the 25 posterior fossa AVMs. In 9 (41%) of the 22 cases of posterior fossa AVMs, the patients presented with a hemorrhage related to a prenidial aneurysm, whereas only 5 (4.7%) of 107 patients with supratentorial AVMs presented with a prenidial aneurysm hemorrhage ($p < 0.01$). These data are summarized in Table 1.

Management and Outcome

The patients' age, AVM and aneurysm characteristics, treatment, and mRS scores are summarized in Table 2. In all 9 cases of hemorrhage due to prenidial aneurysm rupture, the aneurysm presumed responsible for the hemorrhage was treated by means of an endovascular approach on an emergency basis with either coils (5 aneurysms) or glue (4 cases). In most cases (7), the aneurysm was occluded along with its parent artery. One patient (Case 7) died of the initial hemorrhage. In 2 cases, parent vessel occlusion caused a small vermian infarct, with only transient symptoms, if any. There was no permanent neurological complication related to the treatment of prenidial aneurysms. In the 8 surviving patients, 3 unruptured prenidial aneurysms (2 in Case 9, 1 in Case 2) and 1 unruptured posterior inferior cerebellar artery (PICA) aneurysm (in Case 4) were treated electively after the acute phase. Curative treatment of the AVM itself, proven by angiography, was performed on an elective basis in 3 cases (all with embolization followed by resection). One patient (Case 9) was treated the following month, and the other 2 (Cases 4 and 6) were treated 5 months and 20 months, respectively, after the initial hemorrhage, when clinical recovery was optimal. In the remaining 6 patients, the AVM nidus was left untreated: 1 patient (Case 7) died of initial hemorrhage; 1 patient, who was 76 years old at presentation (Case 3), was lost to follow-up; and 4 patients declined definitive treatment of the AVM. One of these patients who declined definitive AVM treatment died 4 years later from AVM rupture (Case 5). The mean follow-up period of the 6 surviving and followed patients was 46.8 months (range 19–74 months). At the end of follow-up, 6 of 9 patients had a good outcome. The mRS score was 0 in 2 cases, 1 in 1 case, 2 in 3 cases, 6 in 2 cases, and 1 patient was lost to follow-up.

TABLE 1: Proportion of bleeding AVMs with prenidial aneurysm rupture

AVM Location	Total No. of AVMs	AVMs w/ Hemorrhagic Presentation	Hemorrhage Due to Prenidial Aneurysm Rupture*
supratentorial	208	107 (51.4%)	5 (4.7%)
infratentorial	25	22 (88%)	9 (41%)

* Percentages calculated based on number of AVMs with hemorrhagic presentation.

Management of prenidial aneurysm rupture in posterior fossa AVMs

TABLE 2: AVM characteristics, management, and clinical outcomes in patients with posterior fossa AVM and prenidial aneurysm rupture*

Case No.	Pt Age (yrs)	AVM Size (cm)	AVM Location	Aneurysm Size (mm)	Aneurysm Location	Treatment	Complication	AVM Nidus Treatment	Outcome
1	58	3.0	superficial right hemisphere	10	right PICA	PVO coils	infarct, asymptomatic	no	43 mos, mRS 0
2	57	2.5	superficial right hemisphere	4	right PICA	PVO glue	infarct, asymptomatic	no	51 mos, mRS 1
3	76	1.5	superior vermis	10	right SCA	PVO coils	none	no	lost to follow-up
4	48	2.5	superficial right hemisphere	6	left SCA	selective coils	none	yes	23 mos, mRS 2
5	58	1.7	superior vermis	5	left SCA	PVO coils	none	no	hemorrhage 4 yrs later, mRS 6
6	21	2.4	superior vermis	3	right SCA	PVO glue	none	yes	19 mos, mRS 2
7	62	1.3	superior vermis	13	left SCA	selective coils	none	no	mRS 6, acute phase
8	63	1.7	superior vermis	5	left PICA	PVO glue	none	no	74 mos, mRS 0
9	45	3.1	superior vermis	multiple aneurysms	left PICA	PVO glue	none	yes	71 mos, mRS 2

* mRS = modified Rankin Scale score; PICA = posterior inferior cerebellar artery; pt = patient; PVO = parent vessel occlusion; SCA = superior cerebellar artery.

Discussion

We present a retrospective study, showing that patients with posterior fossa AVMs frequently presented with prenidial aneurysm rupture. The occurrence of hemorrhagic presentation with the hematoma or hemorrhage center close to a prenidial arterial aneurysm and remote from the AVM nidus was more frequent in patients with posterior fossa AVMs than in those with supratentorial AVMs (41% vs 4.7%) in our patient population.

Westphal and Grzyska¹² also reported that feeding vessel aneurysms occur more frequently in association with infratentorial than supratentorial AVMs. In the series reported by Schmidt et al.,⁸ 30% of posterior fossa AVMs versus 11% of supratentorial AVMs were associated with aneurysms (on feeding vessels within the nidus). The main difference concerned the feeding artery aneurysms (25% vs 5%). Associated aneurysms were the source of bleeding in 10.5% of cases of infratentorial AVMs but only 1.7% of cases of supratentorial AVMs.⁸ In the series of posterior fossa AVMs reported by da Costa et al.,² the presence of associated aneurysms was associated with worse outcomes.

We acknowledge multiple weaknesses in this study. This is not a natural history study, since only treated patients who had at least 1 endovascular treatment were registered and reviewed. Patients were often referred from other centers, and reasons for referrals and indications for treatments have undoubtedly biased numbers. A systematic imaging protocol was not followed, and not all patients presenting with hemorrhage had an early angiogram to specifically look for aneurysms. We found an almost 10-fold difference between supratentorial and infratentorial AVMs, with respect to the presence of ruptured prenidial aneurysms (41% vs 4.7%). This difference may be related to an apparent increased “fragility” that has been evoked in hemorrhagic dissections and nonmycotic distal aneurysms.^{6,7}

Knowledge of this tendency in posterior fossa AVMs may be useful for the early management of these cases given that the associated aneurysms are often easier to treat definitively than the AVMs themselves. This may be especially true should the AVM be associated with a higher treatment risk due to its location size or patient’s factors.

The frequent association of prenidial aneurysms in bleeding posterior fossa AVMs suggests that they should be systematically searched for. We suggest that urgent treatment of the offending aneurysm with embolization may be indicated to prevent early rebleeding, with treatment of the AVM itself being deferred if desirable.

Conclusions

In patients with posterior fossa AVMs presenting with hemorrhage, the hemorrhage is frequently associated with prenidial arterial aneurysms. Our series and others suggest that this association could be more frequent in patients with posterior fossa AVMs than in those with supratentorial AVMs. Urgent endovascular treatment, most frequently by parent vessel occlusion, was effective in this small series.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Roy, Weill. Acquisition of data: Roy, Kouznetsov. Analysis and interpretation of data: Roy, Kouznetsov, Weill, Ghostine. Drafting the article: Roy, Gentric. Critically revising the article: Ghostine, Raymond. Administrative/technical/material support: Gentric.

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