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Management of ruptured posterior fossa arteriovenous malformations

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ABSTRACT

Objective: Posterior fossa arteriovenous malformations (pAVMs) are rare and because of their location at or close to vital structures, their treatment remains challenging despite overall improvements in the management of cerebrovascular lesions. We reviewed our recent series of ruptured pfAVMs in search of guiding principles in the management of these complex lesions.

Methods: This is a retrospective series of consecutive patients admitted for a ruptured pfAVM from 2002 to 2013. We analyzed clinical and radiological data, as well as initial and definitive management. Outcome was assessed using the modified Rankin Scale (mRS) at 6 months.

Results: The study included 34 patients (19 women and 15 men). Upon admission, 79% of patients presented with an intra-cerebellar hematoma, 42% of which required urgent drainage. Hydrocephaly was also present in 82% of patients, 56% of which required emergency ventriculostomy. There was an aneurysm associated with the AVM in 47% of cases. In 38% of the cases, the aneurysm was the source of the hemorrhage. Only 68% of patients were amenable to undergo treatment of the AVM: 24% exclusively by surgery, 9% by embolization, 3% by radiosurgery, and 32% using combined means. Five patients died within the first week: one as a direct result of the severity of the hemorrhage, and the other four due to re-bleeding before treatment. Outcome was favorable (mRS 0–2) in 71% of patients.

Conclusion: Patients with a ruptured pfAVM are often comatose upon admission, requiring emergency live-saving surgical treatment. An associated aneurysm is often the source of bleeding which if dealt with immediately, offers time to plan the most appropriate strategies to eliminate the AVM. Nevertheless, early re-bleeding is frequent, and a cause of concern as it often leads to death. Despite the gravity of the clinical condition upon admission, outcome is favorable for those amenable to treatment.

1. Introduction

Posterior fossa arteriovenous malformations (pfAVMs) are rare, representing only 5–15% of all AVMs, with an incidence in the population of 2 per 10,000 [1–5]. In contrast with supra-tentorial AVMs, pfAVMs present with hemorrhage much more frequently, varying between 72% and 92% depending on the series [1,2,6–8]. A few studies have suggested that the posterior fossa location is an independent risk factor for rupture [4,9,10]. Because of the presence of vital structures in this confined area, they are life-threatening [1–3,11]. Despite of the availability and improvements in multimodal treatments, their management remains challenging [6,7].

The objective of this study is to review our recent series of ruptured pfAVMs in search of guiding principles in the management of these complex lesions.

2. Materials and methods

We reviewed retrospectively a consecutive series of patients admitted for a ruptured pfAVM in our neurosurgical department, between March 2002 and August 2013. Clinical and radiological data were analyzed as well as treatment modalities. AVMs were classified according to the Spetzler-Martin scale [12]. They were considered eloquent if they involved the deep cerebellar nuclei, the brainstem or the cerebellar peduncles. Venous drainage was defined as deep, when at least one vein drained into the deep

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venous system. Associated aneurysms were classified as intranidal, prenidal and remote according to Redekop et al. [13] Outcome was assessed using the modified Rankin Scale (mRS) at 6 months.

3. Results

3.1. Patients

Between March 2002 and August 2013, 34 patients (19 women and 15 men), with a mean age of 46 years (range from 6 to 73 years), were admitted for a ruptured pfAVM.

3.2. Clinical presentation

Eighteen patients (53%) were admitted with impaired consciousness: eight had a glasgow coma scale (GCS) between 13 and 9 and 10, a score inferior to 9, thus requiring intubation. Nine of the 24 patients not requiring intubation on admission, presented with neurological deficits and two with trigeminal neuralgia (Table 1). Initial CT scan revealed a hemorrhage in all cases: a cerebellar hematoma in 27 patients (79%), a subarachnoid hemorrhage (SAH) in 27 patients (79%), an intra-ventricular hemorrhage (IVH) in 18 patients (53%), and a hydrocephaly in 28 patients (82%). As for the 10 intubated patients, 9 presented with a hematoma and all of them with hydrocephaly (Table 1).

3.3. AVM features

An AVM was located in a cerebellar hemisphere in 17 cases, in the vermis in 8 cases, in both these locations in 2 cases, and in the brainstem in 7.

The Spetzler-Martin grade was as follows: 11 patients were classified as grade I, 14 patients were grade II, and 9 grade III. No AVM graded IV or V was observed. A deep venous drainage was present in 14 of the 34 AVMs (Tables 2 and 5).

Table 1
Clinical and radiological initial presentation.

Initial presentation	Total, $n = 34$	Intubate, $n = 10$
Initial GCS		
15–14	16	–
13–9	8	–
8–3	10	10
Neurological deficits	9	
Ataxia	8	
Dysmetry	3	
Dysarthria	1	
Diplopia	4	
Trigeminal neuralgia	2	
Hemorrhage	34	10
SAH	27 (79%)	7
IVH	18 (53%)	7
Hematoma	27 (79%)	9
Hydrocephaly	28 (82%)	10

Abbreviations: GCS, glasgow coma scale; SAH, subarachnoid hemorrhage; IVH, intraventricular hemorrhage.

Table 2
AVM features: location and Spetzler-Martin grade.

Location/Spetzler-Martin grade	I	II	III	IV/V
CH	7	9	1	
V	4	3	1	
CHV	0	1	1	
Brainstem	0	1	6	
Total	11	14	9	0

Abbreviations: CH, cerebellar hemispheres; V, vermis; CHV, cerebellar hemisphere and vermis.

Table 3
Management in emergency.

	Total, $n = 34$	Intubate, $n = 10$
Hematoma	27 (79%)	9
Evacuation	14 (42%)	7
Aneurysm	16 (47%)	9
Embolization	12 (35%)	7
Hydrocephaly	28 (82%)	10
Ventricular drain	19 (56%)	10

Conventional angiogram performed on all 34 patients revealed an aneurysm in 16 (47%) of them, 9 of whom had multiple aneurysms, for a total of 32 aneurysms. The distribution of these aneurysms was as follows: 24 pre-nidals, 5 intra-nidals, and 3 aneurysms unrelated to the AVM. Based on the location of the hemorrhage as seen on the CT-scan it was possible to conclude that the source of the bleeding was the aneurysm in 13 cases (38%) and the AVM for all other cases. Of the eighteen patients who did not have an associated aneurysm, 3 (16%) had a GOS of less than 9, whereas of the 16 patients with an aneurysm, 7 (47%) had a GOS of less than 9. Of the 10 patients who were intubated upon admission, 7 (70%) had an aneurysm responsible for the bleeding.

3.4. Management

Because of the life-threatening clinical condition upon admission, 14 patients (42%) required an urgent evacuation of a cerebellar hematoma (Table 3). Drainage of cerebrospinal fluid (CSF) was necessary in 19 patients (56%), including in all those who were comatose. In cases (13) where the source of hemorrhage was a ruptured aneurysm, aneurysms in 12 patients (35%) were embolized and one was clipped while evacuating a hematoma. The mean time between admission and embolization was 16 h (range from 1 h to 39 h). As for the AVMs (Table 4), 23 patients (68%) were amenable to treatment: 8 patients (24%) underwent surgery alone, 3 patients (9%) underwent embolization alone, one patient (3%) received radiosurgery alone, and 11 patients (32%) were treated with a combination of these modalities. Of these 23 treated patients, in 21, treatment resulted in complete exclusion of the AVM. A residual AVM was seen in two patients, one who had been treated by radiosurgery alone, and one who had undergone embolization followed by radiosurgery. In six patients no attempt was made to treat the AVM: in 5 of them the AVMs were located in the brainstem, and 1 patient with a grade II AVM chose not to be treated.

Case examples from our series are illustrated in Figs. 1 and 2.

3.5. Complications

In this series, 5 patients (15%) re-bled after initial hemorrhage. In four of them, the re-bleeding occurred soon after admission, before initial treatment, all of them between the second and the third day. For these 4 patients, the reason for re-bleeding was an associated aneurysm in 2 cases and the AVM itself in two others (Table 5). Concerning the 2 AVMs that re-bled, both were located in the cerebellar hemisphere, one with deep venous drainage (grade II) and the other without (grade I). All four patients died as a result of re-bleeding. As for the fifth patient, re-bleeding occurred months after the initial treatment, while waiting for definitive treatment. The reason for delaying definitive treatment was the severity of the patient's neurological status after the rupture of the AVM. This one patient who re-bled improved gradually (mRS 3). Of the 19 patients who required an external ventricular drain, one bled in the tract while inserting the drain. This patient had a favorable outcome (mRS 2). Two other patients contracted meningitis resulting from the drain, and were successfully treated. Six patients required a ventriculo-peritonal shunt (VPS). Two patients experienced a stroke: one