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Posterior fossa arteriovenous malformations: Significance of higher incidence of bleeding and hydrocephalus

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ABSTRACT

Objective: Hydrocephalus associated with different types of intracranial arteriovenous malformations (AVMs) has been scarcely studied. In the present report we investigate this association with posterior fossa AVMs (pfAVMs). We hypothesized that there is an increased risk of hydrocephalus and required permanent cerebrospinal fluid (CSF) shunt in patients with pfAVMs that may be linked to the increased risk of bleeding of these lesions. We also review the factors associated with this increased risk of hemorrhagic presentation and we assess how it affects management strategies and functional outcomes in these patients.
Methods: Out of a prospective registry of 374 patients with brain AVMs diagnosed in our center from 1993 to 2013, 60 (16%) had a pfAVM. We described these patients' demographics, their AVM characteristics, clinical presentation, and hydrocephalus incidence and compared the results with those of the supratentorial AVM (spAVM) patients recorded during the same period.
Results: Out of the 60 patients with pfAVMs, 10 (16.7%) presented AVMs located in the brainstem. Hemorrhagic presentation (49/60; 82%) was significantly higher in pfAVMs than in spAVMs (122/314; 38.8%; $p < 0.05$). Hydrocephalus was a common complication in pfAVM patients who had a statistically significant higher need for both temporary external ventricular drain (EVD) (6.7 vs. 20%; $p < 0.05$) and permanent CSF shunts (3.5 vs. 20%; $p < 0.05$). The initial mortality was high (12/60; 20.3%) and half of these patients died before any treatment option could be offered. However, out of those who survived, 70% (42/60) had already shown good clinical outcome at the 6-month follow-up.
Conclusions: Hemorrhagic presentation and hydrocephalus have a higher incidence in pfAVM patients, which initially results in more neurological deficits and an elevated mortality even before receiving any treatment. However, a large number of survivors present good functional outcomes at early follow-up, justifying an aggressive management strategy with microsurgery as the first treatment option in most cases, and radiosurgery as an alternative, especially in brainstem AVMs.

1. Introduction

Posterior fossa arteriovenous malformations (pfAVMs) represent 7 to 15% of all intracranial AVMs. This special subgroup presents with hemorrhage in over 80% of cases, involves different neurological complications when compared with supratentorial counterparts, and presents specific treatment challenges. PfAVM patients face an initial high mortality rate. However, if they survive they show unexpectedly good functional outcomes taking into account the neurological devastation most patients present with.

The clinical course of pfAVMs appears to be more aggressive than in supratentorial AVMs (spAVMs). Patients with pfAVMs are more likely to present with hemorrhage and experience devastating consequences with significant morbidity and mortality. Although hematomas associated with infratentorial AVMs are smaller than those associated with supratentorial AVMs, they are

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badly tolerated and produce a poorer neurological presentation [1]. Such findings warrant an aggressive surgical posture with pfAVMs both before and after rupture.

Since Olivecrona performed the first pfAVM surgery in 1932 [2], surgical approaches and microsurgical techniques for this pathology have significantly improved. Today, surgical removal remains the first treatment choice in most pfAVM cases. However, deep location of the nidus, limited surgical exposure, and the close relationship with eloquent neural structures make this type of surgery challenging and risky [3]. Similar to what happens with basal ganglia AVMs, these surgical challenges in pfAVM have prompted the search for alternative treatments such as radiosurgery, embolization, or even observation.

PfAVMs seem to have more related arterial aneurysms, which are associated with an increased risk of poor functional neurological outcome after AVM rupture. The presence of an associated aneurysm may change treatment strategy. In a ruptured AVM the immediate rehemorrhage risk is low and a definitive treatment could be deferred. In contrast, when a ruptured aneurysm is associated with the AVM, urgent treatment is mandatory to reduce the risk of rebleeding.

Another frequent complication associated with pfAVMs is hydrocephalus. However, this relationship has not yet been studied extensively. Reports about its incidence are scarce and the association of hydrocephalus with different AVM presentations and locations remains unclear.

The goal of this report is to describe our experience with pfAVMs, focusing on multimodality treatment options and main causes of morbimortality, including the initial bleed and subsequent hydrocephalus. We hypothesized that regardless of whether rupture had occurred, hydrocephalus is more frequent in pfAVMs than in supratentorial AVMs. Finally, we review our patients' outcomes and the influence the aforementioned complications had on those outcomes.

2. Clinical material and methods

The study design, a retrospective review of a prospectively collected database, was conducted in compliance with medical ethical guidelines and forcible patient privacy regulations.

2.1. Patients

Over a 20-year period up to December 2013, 374 patients with brain AVMs were managed at our institution and prospectively registered in an internal database. Overall, 60 patients (16%) presented a pfAVM located in the brainstem in 10 cases (2.7%) and in the cerebellum in 50 cases (13.6%). These 60 patients were included in the present study and their medical records, radiographic studies, and clinical evaluations were thoroughly reviewed.

2.2. Clinical presentation

According to their clinical presentation, pfAVMs were recorded as ruptured or unruptured. Ruptured AVMs were further classified into four groups according to the bleeding pattern observed in the initial diagnostic CT. Group 1 included all pfAVMs presenting with intraparenchymal hematoma (IPH); Group 2 included patients presenting only with intraventricular hemorrhage (IVH); Group 3 included patients presenting with IPH and IVH; and Group 4 included pfAVMs presenting with subarachnoid hemorrhage (SAH) alone.

2.3. AVM characteristics

The grading systems and anatomical classifications initially used for our prospective registry were the classic Spetzler–Martin system and the pfAVMs subtypes proposed by Batjer and Samson [4]. Upon diagnosis, all available imaging studies were systematically reviewed in order to grade the AVMs as I, II, III, IV, or V according to the Spetzler–Martin classification [5], while anatomical location was classified as vermian, cerebellar hemispheric, tonsillar, ponto-cerebellar angle, or brain stem.

2.4. Clinical outcome evaluation

Neurological outcome was assessed using the Glasgow outcome scale (GOS) as defined by Jennett and Bond [6]. The assessments were performed at diagnosis, during the first visit, or upon admission to our institution as well as at the 6-month follow-up during a clinical visit. Good outcome was defined as a final GOS score of 4 or 5. Poor outcome was defined as a final GOS of less than 4. Improvement was defined as an increased GOS score and deterioration was defined as a decreased GOS score.

2.5. Statistical analysis

Data were analyzed and summarized using the SPSS for Mac program (Version 20, SPSS, Inc., New York, USA). Characteristics of posterior fossa and supratentorial AVMs were evaluated using descriptive statistics, *t*-test for continuous variables, and chi-square tests for categorical variables. Non-parametric data were evaluated with the Mann–Whitney test for continuous variables and the Fisher exact test was used for categorical variables. Statistical significance was assigned to p values that were less than or equal to 0.05.

3. Results

3.1. Patients

PfAVM patients included in the study consisted of 20 women (33%) and 40 men (67%) with a mean age of 41.6 years (range, 1–79). We did not find any statistically significant differences with regard to gender distribution or mean age when compared to data from the supratentorial AVM group.

Most pfAVM patients (49/60; 81.7%) presented with a hemorrhagic event. Compared to spAVM patients registered in our database, pfAVM patients had more frequent hemorrhagic presentations (81.7 vs. 38.8%; $p < 0.05$). Of the 49 patients who presented with hemorrhage, the first clinical manifestation of the hemorrhagic event in the majority of cases was loss of consciousness (21/49; 42%) or severe headache (19/49; 38.8%). Among the patients with an unruptured AVM, most (7/11; 63.6%) were diagnosed incidentally (Table 1).

3.2. AVM characteristics

The median Spetzler–Martin grade obtained was 2. Of the 60 pfAVM patients, 15 (25%) harbored an SM grade I AVM; 16 (26%) had a grade II AVM, 24 (40%) had a grade III AVM; five (8%) had a grade IV AVM, and five more (8%) harbored a grade V AVM. All 10 brainstem AVMs were classified as SM grade III or greater and all had deep venous drainage (Table 2).

Twelve patients (20.3%) presented one or more associated aneurysms in one of the AVM afferent arteries. These patients were equally distributed among SM grade I (5/12; 41.7%), grade II (3/12; 25%), and grade III (4/12; 33.3%). No flow-related aneurysms associated with higher SM grade AVMs (grades IV and V) were detected.