TESIS DOCTORAL

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INDICADORES CLÍNICO-RADIOLÓGICOS DE VALOR PRONÓSTICO EN LA PATOLOGÍA VASCULAR CEREBRAL

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Intraoperative arteriovenous malformation: causes, management techniques, outcomes, and the effect of neurosurgeon experience.

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Intraoperative arteriovenous malformation rupture: causes, management techniques, outcomes, and the effect of neurosurgeon experience

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Object. Intraoperative rupture can transform an arteriovenous malformation (AVM) resection. Blood suffuses the field and visualization is lost; suction must clear the field and the hand holding the suction device is immobilized; the resection stalls while hemostasis is being reestablished; the cause and site of the bleeding may be unclear; bleeding may force technical errors and morbidity from chasing the source into eloquent white matter; and AVM bleeding can be so brisk that it overwhelms the neurosurgeon. The authors reviewed their experience with this dangerous complication to examine its causes, management, and outcomes.

Methods. From a cohort of 591 patients with AVMs treated surgically during a 15-year period, 32 patients (5%) experienced intraoperative AVM rupture. Their prospective data and medical records were reviewed.

Results. Intraoperative AVM rupture was not correlated with presenting hemorrhage, but had a slightly higher incidence infratentorially (7%) than supratentorially (5%). Rupture was due to arterial bleeding in 18 patients (56%), premature occlusion of a major draining vein in 10 (31%), and nidal penetration in 4 (13%). In 14 cases (44%), bleeding control was abandoned and the AVM was removed immediately ("commando resection"). The incidence of intraoperative rupture was highest during the initial 5-year period (9%) and dropped to 3% and 4% in the second and third 5-year periods, respectively. Ruptures due to premature venous occlusion and nidal penetration diminished with experience, whereas those due to arterial bleeding remained steady. Despite intraoperative rupture, 90% of AVMs were completely resected initially and all of them ultimately. Intraoperative rupture negatively impacted outcome, with significantly higher final modified Rankin Scale scores (mean 2.8) than in the overall cohort (mean 1.5; $p < 0.001$).

Conclusions. Intraoperative AVM rupture is an uncommon complication caused by pathological arterial anatomy and by technical mistakes in judging the dissection distance from the AVM margin and in mishandling or misinterpreting the draining veins. The decrease in intraoperative rupture rate over time suggests the existence of a learning curve. In contrast, intraoperative rupture due to arterial bleeding reflects the difficulty with dysplastic feeding vessels and deep perforator anatomy rather than neurosurgeon experience. The results demonstrate that intraoperative AVM rupture negatively impacts patient outcome, and that skills in managing this catastrophe are critical.
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Key Words • **arteriovenous malformation** • **intraoperative rupture** • **microsurgical resection** • **technique** • **learning curve**

Arteriovenous malformation (AVM) resection should be bloodless, with sharp dissection of subarachnoid spaces, precise occlusion of pial feeding vessels, clean dissection of parenchymal margins, and careful occlusion of deep supply along the ependymal plane.[1,2,6,9,14,15] The slightest bleeding is meticulously stopped to maintain absolute hemostasis before progressing to the next step. Ironically, AVM resections are some of the cleanest and driest dissections despite their highly charged hemodynamics. Nonetheless, some intraoperative bleeding is inevitable with AVM resection, no matter how meticulous the dissection, how skilled the neurosurgeon, or how cooperative the AVM. Cauterizing dysplastic arteries with high flow will stir up bleeding that ranges from a small trickle from a pesky perforator to a torrent from nidal rupture. The threat of intraoperative rupture is pervasive in these cases, and when it occurs, it can transform the operation. Blood suffuses the field and visualization is lost; suction must remain on the bleeding vessel to clear the field and the hand holding the suction device is immobilized; the resection stalls while hemostasis is being reestablished; the cause and site of the bleeding may be unclear; bleeding may force technical errors and morbidity from chasing the source into eloquent white matter; and AVM bleeding can be so brisk that it overwhelms the neurosurgeon and spirals out of control. Although AVM bleeding is unnerving, it must be met with a swift response to avert catastrophe.

Abbreviations used in this paper: AVM = arteriovenous malformation; mRS = modified Rankin Scale.

The technical response to intraoperative AVM rupture differs from aneurysm rupture, which progresses through an orderly sequence of tamponade, suction, proximal control with temporary clipping, distal control with temporary clipping, and permanent aneurysm clipping. There is no aneurysm dome to tamponade with AVM bleeding, and there are too many feeding arteries for proximal control. There is no temporary clipping or neck to close. The bleeding source must be pursued, exposed, and controlled. A small cottonoid is applied at or near the bleeding site and suctioned to dry the field. A jet of blood is traced back to the source, which may be arterial, venous, or nidal. Although there is an extensive literature about intraoperative aneurysm rupture, its management, and the effects on patient outcome, relatively little has been written about intraoperative AVM rupture.[5] Therefore, we reviewed our experience with this feared complication to examine these same factors.

Methods

Patient Population

The study was approved by the Committee on Human Research and conducted in compliance with Health Insurance Portability and Accountability Act regulations. Overall, 591 patients with AVMs were treated surgically by the senior author (M.T.L.) during a 15-year period between September 1997 and January 2013, including 297 female and 294 male patients with a mean age of 38 years (range 1–90 years). Operative reports and surgeon records were reviewed to identify 32 patients (5%) experiencing intraoperative AVM rupture. Their inpatient charts, imaging studies, outpatient clinic evaluations, and data from the prospectively maintained database of the vascular neurosurgery service were reviewed to analyze the patient demographics, AVM features, and surgical factors associated with such events.

Intraoperative AVM Rupture

Intraoperative AVM rupture was defined as torrential bleeding from the AVM, above and beyond what is encountered in typical resections, that was difficult to control. Ruptures were further categorized as follows: 1) bleeding from ruptured or escaped feeding arteries at the AVM margin; 2) bleeding from a microsurgical violation of the AVM margin with penetration of the nidus; and 3) distension and bursting of the nidus due to premature occlusion of venous outflow.

Postoperative Outcomes

Radiographic outcome was assessed with postoperative angiography performed in all patients after microsurgical resection. Patient outcome was assessed using modified Rankin Scale (mRS) scores. Neurological evaluations were performed by a neurologist, neurosurgeon, or research nurse during postoperative clinic visits. Good outcomes were defined as final mRS scores of 0–2, and poor outcomes were defined as a final mRS scores > 2. Improvement was defined as a decrease in mRS score, and deterioration was defined as an increase in mRS score.

Results

Patient Demographic Data

There was a male predominance among the 32 patients with intraoperative AVM rupture (21 male patients, 66%), and the mean age was higher than the age of the overall group (43 years, range 11–90 years). Patients with intraoperative AVM rupture were not more likely to present with hemorrhage (17 patients, 53%, with hemorrhagic presentation). Two patients had incompletely obliterated AVMs after radiosurgery, and 11 patients (34%) underwent preoperative embolization (Table 1). Five patients (16%) had flow-related aneurysms, but these were not the source of intraoperative rupture.

Characteristics of the AVMs

Intraoperative rupture was associated with AVMs in all locations (Table 1). Twenty-five of the intraoperative ruptures (78%) occurred with supratentorial AVMs and 7 (22%) occurred with infratentorial AVMs. However, intraoperative rupture had a slightly higher incidence infratentorially (7 [7%] of 95 AVMs) than supratentorially (25 [5%] of 496 AVMs), although this was not statistically significant ($p = 0.36$).

The majority of intraoperative ruptures occurred with AVMs of intermediate (44%) and high (31%) Spetzler-Martin grades[20] (Table 2). Size was an important point, with 22 AVMs (69%) measuring greater than 3 cm in diameter. Similarly, the majority of intraoperative ruptures occurred with AVMs of intermediate (19%) and high (66%) supplementary grades.[12] Of the factors incorporated into the supplementary grade,[3,4,8,12] diffuseness was an important point, with 20 patients (63%) having diffuse AVMs, which is approximately twice the overall incidence of diffuse AVMs.

Intraoperative AVM Rupture

Arterial bleeding was the most common type of intraoperative AVM rupture; it was observed in 18 patients (56%). Furthermore, the majority of these arterial bleeding events were associated with deep perforating feeding vessels traversing white matter tracts to supply the deep plane of the nidus; such vessels are often difficult to visualize and difficult to coagulate. Frank AVM rupture occurred in 10 patients (31%) after premature occlusion of a major draining vein. Inadvertent nidal penetration was the cause of intraoperative rupture in 4 patients (13%). In 18 cases (56%), intraoperative rupture was managed by controlling the bleeding or working through the bleeding to finish the resection quickly. In the remaining 14 cases (44%), control of bleeding was considered hopeless, as was the ability to work through the bleeding, and the dissection plan was changed to remove the AVM immediately. This so-called commando resection opened access to deep anatomy to control bleeding points or disconnect the final feeding vessels. Eleven AVMs (79%) requiring commando resection were greater than 3 cm in size and 11 were diffuse. Six (33%) of 18 cases of uncontrolled arterial bleeding required commando resection, whereas 6 (60%) of 10 cases of frank rupture after premature venous occlusion required commando resection.

TABLE 1: Characteristics of patients and AVMs, and registered cause of intraoperative rupture*

			AVM Grade		Previous Treatments		Cause of Intraop Rupture		
Case No.	Age (yrs), Sex	AVM Location	SM	Suppl	Radiosurgery	Embolization	Arterial	Nidal	Venous
1	41, F	parietal	4	4	yes	no	no	yes	no
2	25, F	frontal	3	3	no	no	no	no	yes
3	67, M	cerebellar	3	4	no	no	no	no	yes
4	40, M	frontal	1	2	no	no	no	no	yes
5	15, M	occipital	4	2	no	yes	no	no	yes
6	33, M	temporal	4	2	no	yes	yes	no	no
7	63, M	cerebellar	3	4	no	no	yes	no	no
8	46, M	occipital	3	4	no	yes	yes	no	no
9	46, F	occipital	2	4	no	no	yes	no	no
10	34, M	cerebellar	4	4	no	yes	yes	no	no
11	68, F	deep central	2	4	no	no	yes	no	no
12	31, F	frontal	2	2	no	yes	yes	no	no
13	75, M	brainstem	3	3	no	no	no	no	yes
14	11, M	cerebellar	3	3	no	no	yes	no	no
15	46, M	cerebellar	3	4	no	yes	yes	no	no
16	37, F	parietal	5	3	no	no	yes	no	no
17	69, M	parietal	3	4	no	no	no	no	yes
18	41, M	parietal	3	4	no	no	yes	no	no
19	40, F	frontal	3	4	yes	no	no	yes	no
20	23, M	frontal	4	3	no	no	yes	no	no
21	31, F	temporal	3	4	no	no	no	yes	no
22	26, M	occipital	5	4	no	yes	no	no	yes
23	46, F	temporal	2	4	no	no	yes	no	no
24	37, M	temporal	1	4	no	no	no	no	yes
25	45, M	temporal	3	3	no	yes	yes	no	no
26	41, M	parietal	4	4	no	no	no	yes	no
27	48, F	frontal	4	4	no	yes	no	no	yes
28	61, M	temporal	2	4	no	no	yes	no	no
29	65, M	cerebellar	3	4	no	no	yes	no	no
30	20, M	temporal	4	2	no	yes	yes	no	no
31	90, F	temporal	1	4	no	no	yes	no	no
32	36, M	parietal	3	4	no	yes	no	no	yes

* SM = Spetzler-Martin; Suppl = supplementary.

More than half of the intraoperative ruptures (17 cases, 53%) happened during the first 5 years of the senior author's experience. The incidence of intraoperative rupture was highest during this 5-year period (17 ruptures [9%] of 180 microsurgical resections), dropping to 3% (5 ruptures in 177 resections) in the second 5-year period and 4% (10 ruptures in 234 resections) in the third 5-year period. This decrease in rupture rate from the first period to subsequent periods was statistically significant ($p = 0.037$). Most of the ruptures caused by premature venous occlusion occurred during this initial 5-year experience (7 [70%] of 10 cases), and all 4 of the ruptures caused by nidal penetration occurred then, indicating that these technical mistakes may be corrected by experience. Intraoperative rupture due to arterial bleeding was more evenly distributed (6 of 18 during the initial 5-year period), indicating that the difficulty in managing deep perforating vessels is determined more by anatomy than surgeon experience. Commando resections were also more frequent during the initial 5-year period (9 [53%] of 17 cases).

Interestingly, both patients previously treated radiosurgically experienced rupture from nidal penetration, indicating the difficulty in determining the nidal margin in these altered AVMs.

Postoperative Outcomes

Despite intraoperative rupture, 29 (90%) of 32 AVMs were completely resected. In 3 patients (10%), residual nidus was seen on postoperative angiography, and these individuals required a second surgical stage to remove the AVM completely (100% complete resection rate). This incidence of residual AVM was higher than that in the

TABLE 2: Summary of AVM grades in 32 patients

AVM Grade	No. (%)
Spetzler-Martin	
1	3 (9.4)
2	5 (15.6)
3	14 (43.8)
4	8 (25.0)
5	2 (6.3)
Supplementary	
1	0 (0)
2	5 (15.6)
3	6 (18.8)
4	21 (65.6)
5	0 (0)

overall experience (22 [4%] of 591 patients), indicating that complete resection may be compromised when surgery is complicated by intraoperative rupture.

Four patients died in the perioperative period (surgical mortality 12.5%) and 4 more patients (12.5%) died at late follow-up (Table 3). Eighteen patients (56%) were improved or unchanged relative to their preoperative mRS score, and 6 patients (19%) were worse (mean follow-up duration 10 months). Of these 6 patients, 4 still experienced good outcomes (mRS ≥ 2). Outcomes improved in later 5-year periods compared with the first period. Still, intraoperative AVM rupture negatively impacted outcome, with significantly higher final mRS scores (mean 2.8 and median 3) than in the overall cohort (mean 1.5 and median 1; $p < 0.001$).

Discussion

Intraoperative AVM rupture is an uncommon complication, occurring with a 5% frequency that is slightly less than intraoperative aneurysm rupture (7%–15% in our experience[16,19]). Unlike aneurysms that are fragile when they present with subarachnoid hemorrhage and are more likely to rerupture during dissection, intraoperative AVM rupture was not correlated with hemorrhagic presentation.[11] It appears to be caused by technical mistakes in judging the dissection distance from the AVM margin and in mishandling or misinterpreting the draining veins. The drop in intraoperative rupture rate over time and in these two causes in particular suggests the existence of a learning curve, as with most challenging neurosurgical operations.[21] In contrast, the intraoperative rupture rate due to arterial bleeding was steady throughout the study period, indicating that the difficulty with dysplastic feeding and deep perforating vessels reflects nasty anatomical challenges rather than neurosurgeon experience. Our experience demonstrated that intraoperative AVM rupture negatively impacted patient outcome, and therefore it is important to describe insights and techniques for managing this catastrophe that might help others progress through their learning curves.

Intraoperative Rupture Due to Arterial Bleeding

Arterial bleeding originates from the parenchymal

TABLE 3: Summary of clinical outcomes in 32 patients with intraoperative AVM rupture

	Total		Arterial Bleeding		Nidal Penetration		Venous Occlusion	
Scores & Outcomes	No.	%	No.	%	No.	%	No.	%
preop mRS score								
0	4	12.5	2	6.2	1	3.1	1	3.1
1	8	25	5	15.6	1	3.1	2	6.2
2	5	15.6	3	9.4	0	0	2	6.2
3	4	12.5	2	6.2	0	0	2	6.2
4	8	25	4	12.5	2	6.2	2	6.2
5	3	9.4	2	6.2	0	0	1	3.1
6	0	0	0	0	0	0	0	0
final mRS score								
0	5	15.6	2	6.2	1	3.1	2	6.2
1	6	18.8	5	15.6	1	3.1	0	0
2	4	12.5	2	6.2	0	0	2	6.2
3	8	25	3	9.4	0	0	5	15.6
4	1	3.1	1	3.1	0	0	0	0
5	0	0	0	0	0	0	0	0
6	8	25	5	15.6	2	6.2	1	3.1
relative outcome								
improved	11	34.4	8	25	0	0	3	9.4
unchanged	7	21.9	2	6.2	2	6.2	3	9.4
worse	6	18.8	3	9.4	0	0	3	9.4
dead	8	25	5	15.6	2	6.2	1	3.1

side of the resection bed, not from the nidus itself. A cottonoid prevents suction from injuring the brain, and some retraction pressure can be applied to the cottonoid with the sucker tip, allowing the sucker to be a dynamic retractor in the hunt. Small bleeding vessels can be controlled with bipolar forceps and suction, suctioning right at the bleeding point or even drawing the artery into the barrel of the sucker. The sucker simultaneously clears the blood and immobilizes the bleeder, while the bipolar cauterizes the artery proximally. When cautery fails, microclips will close small perforating arteries and aneurysm clips will control large feeding arteries.[1,13] Thin, friable arteries become more manageable as they are exposed proximally. Therefore, arteries with intractable bleeding are chased proximally, unfortunately with deeper transgression into brain tissue that may jeopardize eloquent tracts or cortex.[5,13]

Arterial bleeders are points of hemorrhage that can be closed precisely. They should never be packed, covered with hemostatic agents, or left unattended because they will continue to bleed out of view, either intraparenchymally or intraventricularly. Concealed hemorrhages can also occur with arteries that reopen after they have been coagulated, cut, and retracted into brain parenchyma. These hemorrhages blossom into hematomas and fill ventricles without any apparent bleeding in the surgical field. Progressive brain fullness, narrowing of the surgical corridor, or outward herniation of brain may be the only signs of active bleeding. The hematoma may suddenly erupt into the surgical field. When there is an unexplained change in operative conditions and a concealed hemorrhage is suspected, areas with previous bleeding, persistent oozing, or friable arteries should be reexplored. A trail of edematous or hemorrhagic brain can often be followed to a clot, which is then evacuated and explored for the bleeder. Concealed ventricular hemorrhage quickly fills the ventricles and herniates brain outward, demanding aggressive entry into the ventricle to control the bleeder.

Intraoperative Rupture Due to Nidal Penetration

Dissecting too close to the AVM and violating its margin can rupture the nidus,[22] although this was our least common cause. Judging the right dissection distance is an ongoing process that must balance the eloquence of the adjacent parenchyma with the dangers of dissecting close to the nidal margin. This process can be difficult with diffuse AVMs and those treated radiosurgically with altered margins.[5,12,17] Small breaches are sometimes controlled with cautery, but it can also widen the hole and aggravate bleeding. A small piece of Nu-knit and gentle pressure with the sucker will control minor bleeding. For more significant bleeding, a fixed retractor blade on the Nu-knit will free the hands to continue operating. This hemostatic retraction applies pressure, buys time, and even advances circumdissection by lifting the nidus. The retractor is released after the bleeding site has clotted. Retracting an AVM can, by itself, result in AVM bleeding if it is excessive or if the blade sticks to the AVM margin. Therefore, Nu-knit is used to pad the blade and retraction pressure is minimized.

Intraoperative Rupture Due to Venous Occlusion

Frank AVM rupture usually occurs after inadvertently occluding an AVM's venous outflow.[7,10,18] A large red vessel is eagerly cauterized and cut, expecting drastic reduction in the AVM's arterial supply. Instead, the AVM becomes tense with increased intranidal pressure, feeding arteries become difficult to coagulate, and the nidal margins pop with scattered points of hemorrhage remote from operative site. That misinterpreted "artery" was actually a major draining vein and the AVM is now on the verge of rupture. It is no longer possible to meticulously dissect the remaining planes or to control these random bleeding sites around the nidus. Instead, a commando resection must deliver the AVM before it erupts. There is an interval between the venous occlusion and rupture that is variable and unpredictable, and therefore the resection must accelerate. The occluded vein is no longer functional and can be divided to mobilize the AVM. The AVM is aggressively retracted to hunt down the remaining feeding arteries. Active bleeding requires a large sucker to maintain visualization, and excess bleeding is allowed to run out of the bed, rather than down into the bed. Attention is directed to larger, coagulable feeding vessels. Delicate feeders that would otherwise be carefully closed with microclips may have to wait until after the AVM is removed. Typically, there are two or three last feeding vessels on the polar opposite side of the nidus, and they are found by mobilizing the nidus out of the resection bed, even as the blood pools in it. After occluding these feeding vessels, the AVM slackens. Removal of the AVM opens the bed and makes it easier to scour for active bleeders, typically the smaller ones that were ignored.

The commando resection is a last resort when faced with impending or frank AVM rupture. It is not meant for simple arterial or venous bleeding, or for minor nidal bleeding. Rupture of an AVM calls for decisive action, and the commando resection represents a point of no return. It is a test of one's mettle, requiring the recognition of a mistake and the courage to battle on. There are few situations in neurosurgery more challenging than this one. One must summon skill, insight, and grit at the end of a long case. One must quiet the rush of fear, confusion, regret, stress, frustration, and excitement. One must find calm, clarity, and confidence to continue operating. One must instruct nurses passing instruments, neurosurgeons providing assistance, and anesthesiologists replenishing the blood loss. Although the orderly rhythm of a meticulous AVM resection is suddenly replaced with frenzied chaos, it need not shake one's confidence. All of the AVMs in this experience were resected and no patient died on the operating table. Getting through an intraoperative AVM rupture and a commando resection is an important milestone. It is inevitable for the busy AVM surgeon; it exposes technical errors that can be rectified, like misinterpreting red veins; it establishes one's ability to manage a disaster; and it builds confidence for all subsequent AVM resections, knowing that even the worst conditions are manageable. In the end, it's just bleeding and one's ability to keep working despite bleeding. While the amount of bleeding in these explosive situations can be shocking, it can also be ignored, fixating instead on the last arterial targets and completion of the surgical plan.

Conclusions

Intraoperative AVM rupture is an uncommon complication caused by pathological arterial anatomy and by technical mistakes in judging the dissection distance from the AVM margin and in mishandling or misinterpreting the draining veins. The decrease in intraoperative rupture rate over time suggests the existence of a learning curve. In contrast, intraoperative rupture due to arterial bleeding reflects the difficulty with dysplastic feeding vessels and deep perforator anatomy rather than neurosurgeon experience. Our results demonstrate that intraoperative AVM rupture negatively impacts patient outcome, and that skills in managing this catastrophe are critical.

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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: Lawton. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Rodríguez-Hernández. Statistical analysis: Torné. Study supervision: Lawton.

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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, pontocerebellar 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.

Table 1
Summary of pfAVMs clinical presentation for ruptured and unruptured cases.

	Clinical presentation			
	Ruptured AVMs ($n=49$)		Unruptured AVMs ($n=11$)	
	N	%	*N*	%
Symptoms				
Loss of consciousness	21	42	–	–
Headache	19	38.8	–	–
Cerebellar syndrome	6	12.2	2	18.2
Other neurological deficits	2	4.1	1	9.1
Seizures	1	2	1	9.1
Incidental	–	–	7	63.6

Regarding anatomical classification, cerebellar hemisphere AVM was the most frequent subtype, with 32 cases out of 60 (53.3%). The remaining anatomical subtypes in decreasing order of frequency were vermian AVM (11/60; 18.3%), brain stem AVM (10/60; 16.6%), pontocerebellar angle AVM (6/60; 10%), and tonsillar AVM (1/60; 1.7%).

3.3. Management

The management strategies adopted in our patients are summarized in Table 2. Most patients (34/60; 56.7%) had their AVM surgically removed. Out of these surgically managed patients, 7 (7/60; 11.7%) received previous treatment that consisted of preoperative embolization in two cases (2/60; 3.3%) and a combination of embolization and radiosurgery in the remaining five cases (5/60; 8.3%).

Five patients (5/60; 8.3%) had their AVM-associated aneurysms treated by endovascular means and did not receive any further treatment. One patient was a young female who presented with bleeding, had her associated aneurysm embolized, and died from a rebleed while awaiting surgical removal of her AVM. The remaining four patients were all over 60 years of age. Comparing this group of patients who only had their aneurysm treated with those who had both the aneurysm and the AVM treated, we found a statistically significant difference in the mean age of both groups (60 vs. 42.1 years; $p=0.048$).

Three patients (3/60; 5%) had their AVM embolized as the sole treatment and three (3/60; 5%) received radiosurgery alone. Two of the endovascularly treated patients preferred to receive embolization alone after being informed of the risks and benefits of all management options. The remaining patient was transferred to our institution in a poor neurological state after embolization and bleeding of an SM grade V pfAVM. His family declined further treatment of the AVM. Of the three patients who received radiosurgery, two chose this treatment option based on personal preferences even though they were both offered surgery as the first recommended treatment option. One patient had an SM grade 2 hemispheric AVM that presented with hemorrhage. He had been evaluated at another institution, where he was also offered surgery, and transferred to our department for a second opinion. The other patient had an unruptured SM grade 2 vermian AVM. The third patient harbored an SM grade 3 brainstem AVM that presented with bleeding but was deemed surgically unremovable, and therefore radiosurgery was offered as the only available treatment option at that time.

Overall, 13 patients (21.7%) were conservatively managed and only received symptomatic treatment. Most (7/13; 53.8%) arrived at our institution in deep coma and due to lack of any improvement active treatment was ruled out. Four of these "deep coma" patients had brainstem AVMs. In the remaining six patients (46.2%), conservative management was selected due to family or patient preferences (one patient), patient age (three patients were over 65 years old), AVM location (one brainstem AVM), or comorbidity (one patient was in the early postoperative period after liver transplant).

Table 2
Summary of pfAVM characteristics, treatments and clinical outcomes.

Characteristics	Cerebellar AVMs ($n=50$)		Brainstem AVMs ($n=10$)	
	N	%	*N*	%
Hemorrhagic presentation				
Yes	41	82	8	80
No	9	18	2	20
Spetzler–Martin grade				
I	15	30	0	0
II	16	32	0	0
III	13	26	9	90
IV	0	0	1	10
V	4	4	0	0
Management strategy				
Microsurgery	25	50	2	20
Microsurgery + endovascular	2	4	0	0
Microsurgery + endovascular + radiosurgery	3	6	2	20
Endovascular	8	16	0	0
Radiosurgery	2	4	1	10
Endovascular + radiosurgery	2	4	0	0
Conservative	8	16	5	50
Hydrocephalus	23	46	7	70
Clinical outcome (GOS)				
Dead (1)	7	14	5	50
Persistent vegetative state (2)	1	2	0	0
Severe disability (3)	3	6	2	20
Moderate disability (4)	7	14	1	10
Good recovery (5)	32	64	2	20

3.4. Hydrocephalus

Thirty pfAVM patients (30/60; 50%) presented or developed hydrocephalus as defined by neuroradiological criteria (Evan's index > 0.30) and 24 (24/60; 40%) received treatment for it. In 12 patients (12/60; 20%) temporary external ventricular drainage was sufficient to manage hydrocephalus, but the remaining 12 patients (12/60; 20%) required a permanent cerebrospinal fluid (CSF) shunt placement (Table 3). Two patients with previously diagnosed hydrocephalus presented postoperative CSF leak related to EVD malfunction, but none of the pfAVM patients in our series had an EVD or shunt placed only because of a CSF leak.

In six patients (6/60; 10%) hydrocephalus was left untreated. Four of these six patients presented in deep coma with signs and/or symptoms of non-reversible brainstem injury and therefore any active treatment was ruled out. The remaining two patients had mild hydrocephalus. One arrived at hospital with a Glasgow coma scale (GCS) score of 12. She had an associated aneurysm embolized and died from an AVM rebleed while awaiting surgical treatment. The other patient had a GCS score of 14 and hydrocephalus treatment was withheld until microsurgical removal of the AVM was performed. However, this patient's surgery was complicated with an intraoperative AVM rupture. The patient was kept deeply sedated during the following days and died from a respiratory complication.

Most hydrocephalus (27/30; 90%) developed in patients presenting with hemorrhage. In three patients with unruptured AVM, hydrocephalus developed in the postoperative period after microsurgical AVM removal (3/30; 30%).

Table 3
Summary of hydrocephalus incidence and treatment in AVM patients.

	Supratentorial AVMs ($n = 314$)		Posterior fossa AVMs			
			Cerebellar AVMs ($n = 50$)		Brainstem AVMs ($n = 10$)	
	N	%	N	%	N	%
Hemorrhagic presentation	122	38.9	41	82	8	80
Temporary EVD	12	9.8	12	29.3	0	0
Permanent shunt	8	6.6	7	17	2	25
Non-hemorrhagic presentation	192	61.1	9	18	2	20
Temporary EVD	9	4.7	0	0	0	0
Permanent shunt	3	1.6	1	11.1	2	100

Table 4
Bleeding pattern relationship to hydrocephalus and mortality in pfAVM patients presenting with hemorrhage.

	Bleeding pattern								Total ($n = 49$)	
	Group 1: IPH ($n = 13$)		Group 2: IVH ($n = 14$)		Group 3: IPH + IVH ($n = 16$)		Group 4: SAH ($n = 6$)			
	N	%	N	%	N	%	N	%	N	%
Hydrocephalus	6	46.1	10	71.4	11	68.7	0	0	27	55.1
EVD	5	38.4	9	64.3	7	43.7	0	0	21	42.8
Permanent shunt	2	15.4	5	35.7	2	12.5	0	0	9	18.3
Mortality	0	0	3	21.4	9	56.2	0	0	12	24.4

IPH, intraparenchymal hematoma; IVH, intraventricular hemorrhage; SAH, subarachnoid hemorrhage; EVD, external ventricular drainage.

In terms of bleeding pattern, IVH patients with or without associated IPH were significantly more likely to present hydrocephalus than those with IPH alone ($p = 0.017$; Table 4). Twelve patients (12/60; 20%) presented with hemorrhage due to AVM-related aneurysm, but only six had SAH alone (6/60; 10%). None of those six SAH patients developed hydrocephalus.

Thirteen pfAVM patients with hydrocephalus initially presented in coma (GCS ≤ 8). Three showed signs or symptoms of severe brainstem injury and did not receive any treatment. Of the 10 patients who had an external ventricular drain (EVD) placed to manage hydrocephalus, four (40%) had a good outcome (Fig. 1).

With regard to anatomical location, cerebellar AVM patients required a permanent CSF shunt in 16% of cases (8/50), while brainstem AVM patients received a permanent shunt in 40% (4/10) of cases ($p = 0.101$) (Table 3).

In the supratentorial AVM patient group registered in our database, 20 out of 122 (16.4%) with hemorrhagic presentation and 12 out of 192 (6.3%) with unruptured AVMs developed hydrocephalus. Overall, 11 supratentorial AVM patients out of 314 (3.5%) required a permanent CSF shunt as part of the management strategy. In contrast with these data, pfAVM patients showed a significantly higher need for both temporary EVD (6.7 vs. 20%, $p < 0.05$) and permanent CSF shunts (3.5 vs. 20%, $p < 0.05$) (Table 3).

3.5. Clinical outcome

All patients underwent follow-up and were clinically evaluated at 6 months after clinical onset or first treatment. Overall, 12 patients died (20.3%). Six of these patients (50%), who had arrived to our center already in a deep coma, died before any kind of treatment could be offered. One patient with hemorrhagic presentation due to an associated aneurysm had the aneurysm embolized and died from an AVM bleed while awaiting surgery. The remaining five patients underwent AVM surgery. The cause of death in three patients was pneumonia in the postoperative period, while two died during the follow-up period due to systemic causes related with their initially poor neurological state.

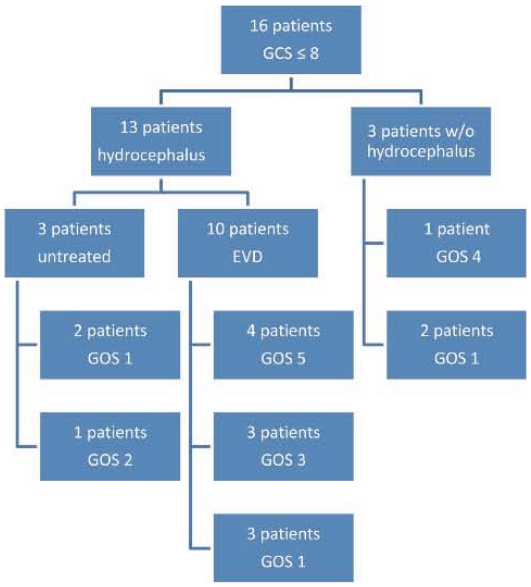

Fig. 1. Scheme showing outcomes of patients presenting in coma (GCS ≤ 8). Abbreviations used in this graphic: GCS, Glasgow coma scale; w/o, without; EVD, external ventricular drainage; GOS, Glasgow outcome score.

Of the initial 60 patients, 42 (70%) presented good outcomes (GOS score of 4–5) at the 6-month follow-up while six patients (10%) had poor outcomes (GOS score of 3–2) (Table 2).

We observed a morbi-mortality rate among pfAVM patients harboring aneurysms of 41.6% (5/12) compared to 27% (13/48) among pfAVM patients without aneurysms, however no statistically significant difference was found ($p = 0.324$).

We observed the poorest outcome in brainstem AVMs (7/10; 70%) when compared with other cerebellar cases (11/50; 22%) ($p = 0.002$). Half of the brainstem AVM patients died at 6 months and two were in poor neurological state, resulting in a 70% morbi-mortality rate (7/10). Cerebellar AVM patients had better outcomes, with a 22% morbi-mortality rate (11/50) that was significantly lower than that observed in brainstem patients ($p = 0.002$).

4. Discussion

4.1. Peculiarities in clinical presentation

The present study reports our experience in the management of a large series of pfAVMs, which accounted for 16% of our AVM cases. As previously described by other authors [7–9], most patients (82%) presented with hemorrhage. Out of the unruptured cases, more than half were incidentally diagnosed and only a few cases presented neurological symptoms that prompted the diagnostic workout.

The causes behind increased hemorrhagic presentation in pfAVMs, which is a commonly accepted phenomenon, remain unclear. Some of the proposed possible causes for increased bleeding are a higher incidence of deep venous drainage [8], hemodynamic peculiarities [10], smaller nidus size [11,12], and frequent stenosis in the venous drainage [13,14]. An intriguing new idea is that many pfAVMs are simply asymptomatic (no neurological symptoms other than bleeding) and are only diagnosed when they rupture [4,15]. Recent studies by Khaw et al. [8] and Rodríguez-Hernández et al.[9] found that the percentage of spAVM patients presenting with seizures approximated the difference in hemorrhagic presentation when compared to pfAVM patients. Therefore, apart from patients experiencing seizures they found no statistically significant differences in hemorrhagic presentation between pfAVMs and supratentorial AVMs. This "silent course" hypothesis is also supported by the findings in our series and those reported by Stapf et al., which suggest that patients with pfAVMs are diagnosed at an older age than those with spAVMs [16]. The absence of non-hemorrhagic neurological symptoms (such as seizures) in younger patients could explain why pfAVMs might remain undiagnosed, and therefore untreated, until bleeding occurs.

A second controversial topic on hemorrhagic presentation involves brainstem AVMs. Brainstem AVMs account for 2 to 6% of all brain AVMs [17]. Of the 374 AVMs evaluated in our center, 10 presented brainstem AVM (3.7% of all brain AVMs). It is difficult to determine the risk of bleeding in these lesions due to their low prevalence, but it appears to be similar to that of other pfAVMs [18,19]. Some authors report a lower bleeding risk attributed to early diagnosis because of the cranial nerve and brainstem symptoms associated with AVM pulsatility [15,20]. In our series, brainstem AVM patients presented with hemorrhage in 80% of cases, a figure that did not differ from that of our cerebellar AVM patients.

Most of our unruptured pfAVM patients were diagnosed incidentally, a fact that further supports the "silent course" hypothesis. Other clinical forms of presentation included headache, cranial nerve palsies, and typical cerebellar symptoms such as dysmetria, dysarthria, or gait imbalance. Oddly enough, two patients in our series, one of whom harbored an unruptured AVM, presented with seizures. This form of presentation is infrequent in pfAVMs and has been described only rarely [21].

4.2. Hydrocephalus

One important difference found between pfAVMs and spAVMs was the incidence of hydrocephalus. To the best of our knowledge, this difference has not been previously investigated or examined in detail [22]. Twenty percent of our pfAVM patients required a temporary EVD and another 20% required a permanent shunt system, while in supratentorial AVM patients 6.7% required EVD placement and 3.5% required permanent CSF shunt. The aforementioned higher hemorrhagic risk with pfAVMs could be a plausible explanation for the higher hydrocephalus incidence. Furthermore, pfAVM hemorrhage often invades the ventricular system and we found that the presence of intraventricular hemorrhage is associated with higher rates of hydrocephalus. Therefore, we suspect that the increased tendency to bleed, together with a frequent intraventricular hemorrhage invasion, could be behind the higher incidence of hydrocephalus in pfAVM patients.

More than half of patients with ruptured pfAVM in our study presented with hydrocephalus, but in only 18.3% of cases a permanent CSF shunt was necessary. The vast majority of patients with bleeding requiring temporary ventricular drainage were able to subsequently avoid the placement of a permanent shunt. If we single out brainstem AVM patients, 40% in our series required a permanent shunt, although this finding was not statistically different from the cerebellar AVM patient group ($p = 0.083$).

With regard to the specific bleeding patterns, intraventricular bleeding was linked to hydrocephalus in 70% of the cases, but only 23.3% of patients presenting IVH (with or without IPH) eventually required a permanent shunt. Surprisingly, there were differences between the two IVH groups: we found that the percentage of cases needing a permanent shunt was higher in Group 2 (only IVH; 35.7% shunt placement) than in Group 3 (IVH with IPH; 12.5% shunt placement). The higher mortality observed in IVH patients with IPH could explain this difference because patients did not survive long enough to be considered for permanent CSF shunt placement. In any case, hydrocephalus incidence was significantly higher in patients with any kind of IVH (Group 2 and Group 3) than in those with IPH alone (Group 1) ($p = 0.017$).

To summarize, hydrocephalus is a common complication in pfAVM patients, unlike those with spAVMs. The placement of an EVD should be a priority in patients that present with a decreased level of consciousness due to hydrocephalus. However, many of them avoid permanent shunt placement and can have a good outcome, even when first presenting in coma. A higher incidence of hydrocephalus appears to be associated with a higher incidence of hemorrhagic presentation and the frequent intraventricular invasion observed in these AVM patients.

4.3. Treatment

Generally speaking, microsurgical removal remains the first treatment choice for cerebral AVMs. Other management options, such as embolization and radiosurgery, have been used as a single treatment option or in combination with other treatments. More than half of our patients (56%) were treated with surgery as a definitive treatment, but in seven of these patients other therapeutic options were previously combined as adjuvant to surgery. In most cases, surgical treatment was delayed and performed electively [4,23]. Emergency surgery is indicated when there is an associated aneurysm that may be responsible for the hemorrhage. In the absence of associated aneurysms, hematomas larger than 3 cm with compression of the fourth ventricle and obliteration of the basal cisterns also warrant emergency surgery [24]. Staged surgery has also been described when there is significant edema that could hinder AVM resection. In these cases, the hematoma could be evacuated at the first stage, thus delaying the definitive AVM removal until parenchymal edema resolves 2–3 weeks later [23,25].

Preoperative embolization serves two purposes in pfAVM management: (1) the treatment of associated aneurysms and (2) partial occlusion of the AVM nidus prior to surgery. Radiosurgery is useful

in deep and small AVMs that have not been previously embolized because embolization might interfere with the radiosurgical treatment. Bowden et al. obtained occlusion rates of 76% at 5 years in pfAVM patients treated with radiosurgery alone [26]. In contrast, Kelly et al. [27] advocated for multidisciplinary treatments because isolated radiosurgery did not prove useful in their experience. In our series, only six patients received one of these two treatments alone.

Brainstem AVMs are a special subset of pfAVMs. There are several reports on surgical treatment for brainstem AVMs [28,29], but they describe complex approaches and demanding microsurgical techniques that carry with them a high morbidity and mortality in patients who, in many cases, already have severe neurological impairment. Accessibility through the subarachnoid space and nidus size are key aspects in determining the feasibility of a microsurgical treatment for this type of pfAVM [29,30]. Nonetheless, tailored rebleeding risk and treatment alternatives offered by radiosurgery and embolization should be carefully assessed before deciding on surgical removal. Factors such as patient age, comorbidities, intrinsic brainstem location, poor neurological state, or patient preferences influence the final therapeutic decision [31,32]. Of our 10 brainstem AVMs patients, two were treated with microsurgery alone, one with radiosurgery, and two with a combined treatment including embolization, radiosurgery, and microsurgery. In the remaining five brainstem AVM patients, the final decision was to refrain from performing any active treatment.

4.4. Clinical outcomes

The high mortality rate (20%) observed in pfAVM patients is largely explained by the higher hemorrhagic presentation and usually poor initial neurological status of the patients upon arrival to the hospital. Overall, 32% of our pfAVM patients presented with a GCS that was less than or equal to 8. A lower tolerance of the posterior fossa to hematoma volume, a frequently observed injury to brainstem structures, and consequent hydrocephalus may explain the large number of pfAVM patients presenting in coma.

Nevertheless, patients who manage to survive the acute phase of their severe condition usually show a good functional status at 6 months. In our series, 70% of pfAVM patients had a GOS greater than or equal to 4 at the 6-month time mark, a finding that does not differ from previous studies reporting good final outcomes in 70 to 87% of cases [4,19,33]. Several theories try to explain these unexpectedly good final outcomes. First, the aforementioned initial coma is often related to other complications such as hydrocephalus, which is usually easily treated. Furthermore, cerebellar lesions that do not affect the dentate nuclei should not prevent the patients' future functional independence [19]. Finally, some of the initially observed motor deficits are temporary and recoverable. Therefore, poor initial neurological status in pfAVM patient per se should not be sufficient criteria to withdraw care.

Despite a similar incidence of hemorrhagic presentation, patients harboring brainstem AVMs have a worse prognosis than those with other pfAVMs. In our brainstem AVM patient group, five (50%) died within the first 6 months and only three (30%) had a good functional outcome (GOS $\geq$ 4). Although surgical treatment can be successful in these patients, it is usually highly complex and deficits derived from either the surgical treatment itself or the initial AVM bleed can obscure the clinical outcome. The lower incidence of brainstem AVMs, the higher risks associated with treatment, and the poorer natural outcomes call for a tightly tailored, case-to-case evaluation in order to give an accurate prognosis and recommend the best management strategy to the patients and their families.

5. Conclusion

Most pfAVMs patients present with hemorrhage, but the causes behind this characteristic clinical behavior remain unclear. Suggested differences in age at presentation, seizure incidence, and hemodynamic factors associated with the particular anatomy of the posterior fossa are intriguing factors that could elucidate this matter, but further research is needed.

Unlike in supratentorial AVMs, hydrocephalus is a common complication in pfAVMs. The usual hemorrhagic presentation with frequent intraventricular invasion might be the cause of the higher hydrocephalus incidence we observed. As a preventable cause of neurological impairment, diligent treatment of acute hydrocephalus is mandatory. However, out of the 40% of patients requiring initial treatment, half can end up avoiding a permanent CSF shunt.

Increased likelihood of hemorrhagic presentation and acute hydrocephalus result in poorer neurological status initially, but good functional outcome at early follow-up in a large number of patients should be expected, therefore justifying a more aggressive management strategy. Microsurgical resection, with or without preoperative embolization, remains the first treatment option for ruptured pfAVMs and most unruptured cases. Best timing could be argued, but leaves no room for doubt in cases of associated aneurysms and/or mass effect from the hemorrhage. Radiosurgery is usually the preferred alternative for most brainstem AVMs, which constitute a special subtype of pfAVMs.

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3. Resultados y discusión

El neurocirujano forma parte activa en el diagnóstico, tratamiento y seguimiento del paciente afecto de patología vascular cerebral. En los últimos años se han producido importantes avances, sobretodo en lo referente al tratamiento mediante las nuevas técnicas endovasculares que parece ser una de las principales causas de los mejores resultados clínicos(8).

A pesar de todo, el pronóstico de estos pacientes continua siendo en muchas ocasiones malo e incierto(86). La patología cerebrovascular permanece entre las causas con mayor mortalidad y morbilidad en nuestro medio. Son habituales los déficits motores, sensitivos y cognitivos con los que sobreviven muchos de estos pacientes. Estas secuelas representaran además un elevado gasto de recursos humanos y económicos que debe tenerse en cuenta también en la toma de decisiones. La percepción que tiene el propio individuo al año del ictus de su propia enfermedad o la que tienen sus familiares puede llegar a ser más satisfactoria de lo esperado inicialmente, incluso en el caso de patología con secuelas neurológicas muy invalidantes e irreversibles(87). Sin duda, esta percepción de lo que la enfermedad representa para el paciente y familiares se basa en un importante componente social y cultural. Sobre el neurocirujano recae un importante papel; no sólo deberá ser un experto en la realización de su técnica quirúrgica si no que deberá realizar una certera predicción de la evolución clínica y posibles déficits que presentará el paciente durante su proceso.

3.1. Resultados

3.1.1. Hemorragia subaracnoidea y lupus eritematoso sistémico

Realizamos una revisión de nuestra base de datos prospectiva de 641 pacientes con HSA entre 1996 y 2013, identificando tres pacientes previamente diagnosticados de LES. Las características del sangrado y la evolución clínica que tuvieron son analizadas en el trabajo. Efectuamos una búsqueda sistemática en la literatura utilizando como palabras clave en PubMed: "*subarachnoid hemorrhage*" y "*systemic lupus erythematosus*" en el periodo comprendido entre 1980 y 2013 **(tabla 2)**. Al realizar la revisión de la literatura obtenemos 39 pacientes con HSA y LES. Cinco de los 11 pacientes con HSA no aneurismática presentaron un mal pronóstico funcional. De los 27 restantes, seis presentaban aneurismas múltiples y seis aneurismas en disposiciones atípicas o aneurismas fusiformes. Al examinar la cúpula de un paciente con pseudoaneurisma en una localización atípica intervenido en nuestro centro, no se observó un infiltrado inflamatorio que sugiriera un proceso vasculítico intercurrente.

Tabla 2: Pronóstico funcional de los pacientes con HSA y LES descritos en la literatura.

	No. de pacientes	Tipo de aneurisma	Localización del aneurisma	Enfermedad sistémica	Pronóstico
Asai et al., 1989	1	Múltiple, sacular	ACoP,AB,ACoA	Vasculitis activa por patología	Muerte
Atanes et al., 1989	1	Múltiple, sacular	ACoA, ACoP	LES activo	No deficits
Baizabal Carvallo et al., 2007	10	5 aneurismas, 5 angio-negativas	4 ACM, 1 ACoP	4 con LES activo	1 HSA angio-negativa y 3 pacientes con aneurisma mueren
Brah et al., 2012	1	Múltiple, fusiforme	ASC, Arteria lenticulostriada	LES activo	Muerte
Gillard et al., 2002	1	Múltiple, fusiforme	ASC, ACoA, ACM	LES activo	NR
Hashimoto et al, 1986	2	sacular	AB, ACoA	LES activo	1 déficits neurológicos
Kelley et al., 1980	1	fusiforme	ACoP	Vasculitis activa por patología	Muerte
Mimori et al.,2000	10	4 saculares, 4 angio-negativas y 2 sin arteriografía	ACM, 2 ACoP, 1 ACoA	5 con LES activo	7 muertes
Nagayama et al, 1989	1	sacular	BA	LES sin actividad	Déficits neurológicos
Nakai et al.,2000	1	sacular	ACS	LES sin actividad	Buena recuperación
Lim et al., 2013	1	Tipo blister	ACI	NR	Muerte
Owada et al, 2009	2	1 múltiple, 1 sacular	ACM bilateral y ACI	LES activo en ambos	Buena recuperación
Refai et al., 2008	1	HSA angio-negativa	No	LES sin actividad	No déficits neurológicos
Rozet et al, 2004	1	sacular	PICA	LES sin actividad	Muerte
Sakaki et al., 1990	2	sacular	ACM, ACoA	Vasculitis activida por patología	1 buena recuperación
Sánchez-Ojanguren et al., 1999	1	sacular	ACoA	LES activo	Muerte
Segura et al., 1998	1	HSA angio-negativa	No	LES activo	No déficits
Tang et al, 2011	1	Multiple, fusiforme	(ACI, ACM…)	LES activo	Déficits neurológicos

Abreviaciones: ACoP: arteria comunicante posterior, AB: arteria basilar; ACoA: arteria comunicante anterior, ACM: arteria cerebral media; HSA: hemorragia subaracnoidea; LES: lupus eritamoso sistémico; ASC: arteria cerebelosa superior; ACI: arteria carótida interna; PICA: arteria cerebelosa postero-inferior; NR: No reportado.

3.1.2. Paciente en coma con hematoma subdural aneurismático

Un total de 440 pacientes con HSA aneurismática fueron valorados en nuestro centro a partir del 2005. Diecinueve (4,3%) presentaban un hematoma subdural y 13 (2,9%) se diagnosticaron con un bajo nivel de conciencia (GCS≤8). La localización más frecuente fue la ACoP (5; 38.4%) seguida de la ACM (4; 30,7%) y la ACoA (4; 30,7%). Cuatro pacientes presentaban un hematoma intraparenquimatoso con laceración cortical (4; 30,7%). La mediana del grosor del hematoma fue de 10 mm con un rango entre 3 y 15 mm. El tratamiento del aneurisma fue mediante clipaje quirúrgico en nueve pacientes, el resto se trató mediante un procedimiento endovascular. Un total de ocho pacientes requirieron una craniectomía descompresiva primaria.

Cinco pacientes fallecieron durante el postoperatorio. De los supervivientes, la mayoría (5/8; 62.5%) tuvieron un buen pronóstico funcional (GOS 4-5) al evaluarse entre los 6 y 12 meses. Los pacientes en coma con subdural que evolucionaron a un buen pronóstico funcional fueron parecidos a aquellos pacientes en coma sólo con hemorragia subaracnoidea (HSDa; 38,5% vs HSA; 26,4%).

3.1.3. Rotura intraoperatoria de las MAVs

La rotura intraoperatoria de una MAV es una de las complicaciones más temidas en neurocirugía y convierte un técnica minuciosa y reflexionada en un cirugía improvisada donde el objetivo pasa a ser el de salvar la vida del paciente. Definimos la rotura intraoperatoria de una MAV como un sangrado torrencial de difícil control muy superior al sangrado típico durante las resecciones habituales. La rotura intraoperatoria de una MAV se da en menor frecuencia que la rotura intraoperatoria de los aneurismas cerebrales(88,89) y requiere de una técnica quirúrgica distinta para solucionarlo.

Los datos del estudio pertenecen a una cohorte de 591 MAVs tratadas quirúrgicamente en la servicio de neurocirugía de la UCSF (*University of California,*

San Francisco) durante un periodo de 15 años (1997-2013) por un mismo cirujano. Identificamos 32(5%) pacientes con rotura intraoperatoria tras revisar retrospectivamente las hojas quirúrgicas y los apuntes realizados por el cirujano en una base de datos prospectiva. Las características clínicas, radiológicas y causas de la rotura han sido analizadas retrospectivamente (**tabla 3**) al igual que el pronóstico funcional (**tabla 4**).

En nuestro estudio obtenemos una predominancia de sujetos varones 21/32 (66%) con un media de edad de 43 años. Del total, en 11 pacientes se había realizado una embolización prequirúrgica y en 2 una oclusión incompleta de la MAV mediante radiocirugía. En 5(16%) de los pacientes existía un aneurisma asociado, sin ser la causa de la rotura intraoperatoria de la MAV. Observamos una distribución heterogénea de las MAVs en distintas localizaciones, siendo predominantes las lesiones supratentoriales (25/32; 78%) a las infratentoriales (7/32; 22%). Se obtuvo un leve mayor riesgo de rotura intraoperatoria en las MAVfp (7/95; 7%) al compararlas con las MAVsp (25/496; 5%) sin alcanzar diferencias significativas (p=0,36).

La mayoría de roturas se producen en MAVs grado III (44%) y grado IV-V (31%) de la clasificación de Spetzler-Martin(79). En 20(63%) pacientes la MAV fue catalogada como difusa, siendo el doble de frecuente que en el resto de MAVs.

Las causas de sangrado fueron categorizadas en tres tipos:

- Sangrado o rotura por arterias nutricias perforantes en el margen profundo de la MAV.
- Penetración del nidus malformativo durante la resección.
- Rotura del nidus malformativo por oclusión prematura de la vena de drenaje.

El sangrado procedente de las arterias perforantes fue el más frecuente en 18(56%) casos, la rotura accidental o cierre precoz de la vena de drenaje ocurrió en 10(31%) y la invasión del nidus fue la causa menos frecuente, sólo observándose en 4(13%) pacientes. Del total, en 18(56%) casos el cirujano pudo finalizar la cirugía con una

resección perinidal; en el resto, el sangrado no permitió una resección reglada requiriendo una cirugía con márgenes y a la carrera. Más de la mitad de la roturas intraoperatorias 17(53%) se produjeron durante los primeros 5 años de experiencia quirúrgica del cirujano. Se observa una disminución significativa de roturas intraoperatorias entre el primer periodo 17/180(9%) al compararlo con el segundo (5/177,3%) y el tercero (10/234,4%) (*p*=0,037). La causa de rotura más importante durante los primeros cinco años fue la oclusión de la vena de drenaje (7/17; 41,1%), seguida por la invasión del nidus (4/17; 23,5%) y finalmente por el sangrado de las arterias perforantes (6/17; 35,2%). Describimos una clara curva de aprendizaje entre el primer periodo y los siguientes con un mayor número de rotura debido a un error en la técnica quirúrgica (oclusión prematura de la vena de drenaje y penetración del nidus). Durante estos primeros 5 años, la resección con márgenes por sangrado incontrolable también fue mayor (9/17; 53%), secundaria a una menor capacidad técnica para controlar la hemorragia al compararlo con los periodos siguientes.

Centrándonos en el pronóstico, observamos que ocho pacientes fallecieron; la mitad en el postquirúrgico inmediato y el resto de forma más diferida en relación con complicaciones derivadas de la rotura de la MAV. A pesar de todo, en la mayoría de pacientes no se evidenció un empeoramiento clínico en la evaluación postoperatoria 18/32 (56%). Comparamos mediante el mRS (*modified rankin score*) los resultados obtenidos en la cohorte de pacientes con rotura intraoperatoria (media: 2,8) con el resto de MAVs (media: 1,5), observando un peor pronóstico funcional en aquellos pacientes que han presentado una rotura intraoperatoria (*p* = 0.001).

Tabla 3: Sumario de los pacientes con rotura intraoperatoria

Edad	**Localización MAV**	**Grado MAV**	**Tratamientos previos**	**Causa rotura intraoperatoria**				
		Escala S-M	Escala suplementaria	Radiocirugía	Embolización	Arterial	Nidal	Vena drenaje
41	Parietal	4	4	Yes	No	No	Yes	No
25	Frontal	3	3	No	No	No	No	Yes
67	Cerebelar	3	4	No	No	No	No	Yes
40	Frontal	1	2	No	No	No	No	Yes
15	Occipital	4	2	No	Yes	No	No	Yes
33	Temporal	4	2	No	Yes	Yes	No	No
63	Cerebelar	3	4	No	No	Yes	No	No
46	Occipital	3	4	No	Yes	Yes	No	No
46	Occipital	2	4	No	No	Yes	No	No
34	Cerebelar	4	4	No	Yes	Yes	No	No
68	Ganglios	2	4	No	No	Yes	No	No
31	Frontal	2	2	No	Yes	Yes	No	No
75	Tronco	3	3	No	No	No	No	Yes
11	Cerebelar	3	3	No	No	Yes	No	No
46	cerebelar	3	4	No	Yes	Yes	No	No
37	Parietal	5	3	No	No	Yes	No	No
69	Parietal	3	4	No	No	No	No	Yes
41	Parietal	3	4	No	No	Yes	No	No
40	Frontal	3	4	Yes	No	No	Yes	No
23	Frontal	4	3	No	No	Yes	No	No
31	Temporal	3	4	No	No	No	Yes	No
26	Occipital	5	4	No	Yes	No	No	Yes
46	Temporal	2	4	No	No	Yes	No	No
37	Temporal	1	4	No	No	No	No	Yes
45	Temporal	3	3	No	Yes	Yes	No	No
41	Parietal	4	4	No	No	No	Yes	No
48	Frontal	4	4	No	Yes	No	No	Yes
61	Temporal	2	4	No	No	Yes	No	No
65	Cerebelar	3	4	No	No	Yes	No	No
20	Temporal	4	2	No	Yes	Yes	No	No
90	Temporal	1	4	No	No	Yes	No	No
36	Parietal	3	4	No	Yes	No	No	Yes

Tabla 4: Comparativa del pronóstico funcional según la causa de rotura intraoperatoria

	Total		**Sangrado perforantes**		**Invasión nidus**		**Oclusión vena drenaje**	
	N	**%**	**N**	**%**	**N**	**%**	**N**	**%**
mRS preoperatorio								
0	4	12,5	2	6,2	1	3,1	1	3,1
1	8	25	5	15,6	1	3,1	2	6,2
2	5	15,6	3	9,4	0	0	2	6,2
3	4	12,5	2	6,2	0	0	2	6,2
4	8	25	4	12,5	2	6,2	2	6,2
5	3	9,4	2	6,2	0	0	1	3,1
6	0	0	0	0	0	0	0	0
mRS final								
0	5	15,6	2	6,2	1	3,1	2	6,2
1	6	18,8	5	15,6	1	3,1	0	0
2	4	12,5	2	6,2	0	0	2	6,2
3	8	25	3	9,4	0	0	5	15,6
4	1	3,1	1	3,1	0	0	0	0
5	0	0	0	0	0	0	0	0
6	8	25	5	15,6	2	6,2	1	3,1
Pronóstico relativo								
Mejoría	11	34,4	8	25	0	0	3	9,4
No cambio	7	21,9	2	6,2	2	6,2	3	9,4
Empeoramiento	6	18,8	3	9,3	0	0	3	9,4
Muerte	8	25	5	15,6	2	6,2	1	3,1

3.1.4. MAVs de fosa posterior

A partir de un registro de 374 pacientes con MAVs cerebrales diagnosticados de forma prospectiva en el "Hospital Universitario de la Vall d'Hebron" durante un periodo de 20 años se identifican 60(16%) MAVs de fosa posterior (MAVfp). Un total de 10(16,7%) pacientes tenían la lesión localizada en el tronco encefálico. Describimos un mayor riesgo de hemorragia (49/60; 82%) al comparar estas lesiones con el resto de MAVsp (MAVs supratentoriales) (122/314; 38,8%; p<0.05). La forma principal de presentación fue la pérdida de consciencia (21/49; 42%) y la cefalea brusca (19/49; 38,8%). Entre los pacientes con MAVs no rotas, la mayoría fueron diagnosticados de forma incidental al realizar la prueba de imagen por otras causas (7/11; 63,6%).

La hidrocefalia fue una complicación frecuente en estos pacientes (30/60; 50%). La mayoría de pacientes con hidrocefalia (24/30; 80%) se trataron; sólo en seis se decidió no realizar ningún tratamiento por el mal estado neurológico inicial y las irreversibles lesiones que se observaban en las pruebas de imagen. Las MAVfp presentan un mayor requerimiento de drenaje externo (DVE) (20% vs. 6,7%, p <0,05) y de derivación ventriculo-peritoneal (DVP) (20% vs. 3,5%, p <0,05) al comparárlo con el resto de MAVsp. Al centrarnos en el patrón de sangrado, observamos un mayor riesgo de hidrocefalia en aquellos pacientes que presentaron hemorragia intraventricular (HIV) asociada o no a un hematoma intraparenquimatoso (HIP) si se compara con los que presentaron un HIP aislado (p =0.017).

Al centrarnos en el pronóstico funcional de estos pacientes, observamos una elevada mortalidad inicial (12/60; 20.3%); sin embargo el pronóstico funcional de los supervivientes no fue malo. Los resultados funcionales fueron evaluados a los seis meses de seguimiento mediante la escala de GOS(*Glasgow Outcome Scale*) (90). Un total de 42(70%) pacientes presentaban un GOS de 4 o 5 en esta evaluación. No observamos diferencias en el pronóstico funcional entre aquellas MAVfp con aneurismas asociados. Al comparar los distintos tipos de MAVfp según su

localización, observamos un peor pronóstico en las MAV de tronco encefálico (7/10; 70%) al compararlas con el resto de MAVfp (11/50; 22%; *p*=0,002).

3.2. Discusión

3.2.1. Hemorragia subaracnoidea

3.2.1.1. Descripción de tres patrones en relación con el pronóstico funcional en los pacientes con HSA y LES

Existe poca literatura en referencia a como una enfermedad previa puede afectar a la evolución, clínica y pronóstico de un paciente que ha presentado una HSA. Aunque diversos trabajos se centran en factores de riesgo como la hipertensión arterial (HTA), diabetes mellitus (DM) o dislipemia (DLP) (8,17), pocos estudian las complicaciones del sangrado en pacientes afectos de otras patologías.

El LES constituye una enfermedad autoinmune que afecta frecuentemente a gente joven caracterizada por la presencia de inmunocomplejos anti-DNA y Anti-Ro. La mitad de los pacientes con LES tienen afectación cerebral, el ictus representa un 15% de las complicaciones neurológicas siendo el más frecuente el ictus isquémico(91). La incidencia de la HSA en el LES parece ser superior a la población normal(82). Las causas de esta mayor incidencia no están claras en la literatura y pueden ser debidas a fenómenos vasculíticos, efectos adversos del tratamiento crónico con inmunosupresores o a el mayor riesgo de presentar aterioesclerosis que sufren estos pacientes (82,92-98).

Aunque es frecuente la presentación de pacientes con LES y aneurismas saculares simples como ocurre en la población general(99), de forma común se describen en la literatura casos con aneurismas poco habituales y evoluciones tórpidas de la HSA(49,95,97,100-106).

Estas peculiaridades, que parecen seguir unos patrones no descritos previamente, se observan persistentemente en las publicaciones de pacientes con HSA y LES.

Nuestro trabajo describe tres nuevos patrones clínicos a considerar y su relación con el pronóstico funcional:

- HSA no aneurismática con mala evolución clínica(49,106,107).
- Aneurismas múltiples(95,100-102).
- Aneurismas en disposiciones atípicas o fusiformes(95,97,100,103-105,108-110).

La revisión de nuestros datos aporta un caso ilustrativo para cada uno de estos patrones.

Diversos casos de HSA no aneurismática en LES son descritos en la literatura(111,112), raramente estos pacientes presentan un sangrado perimesencefálico típico de la HSA angiograficamente negativa o idiopática(10,11,13), en algunos de ellos se solicitó incluso confirmación postmortem de la ausencia de aneurismas(49,113). Generalmente, la HSA idiopática se caracteriza por un patrón perimesencefálico(16) secundario a un sangrado venoso, habitual en gente añosa y con un muy buen pronóstico funcional. Nada que ver con las HSA negativas descritas en nuestra revisión, dónde el mal pronóstico funcional es cercano al 50% de los pacientes. De los 11 pacientes descritos en la literatura, cuatro fallecieron y uno presentó un mal pronóstico funcional. En nuestro articulo describimos una paciente diagnosticada de HSA angio-negativa con un grado de IV en la escala de Fisher(53) que presentó un resangrado y que finalmente falleció. Este mal resultado clínico puede explicarse por las complicaciones médicas de estos enfermos, como la nefropatía y los procesos infecciosos. Las discrasias sanguíneas son frecuentes y pueden explicar en parte los sangrados masivos sin presencia de aneurismas(114-116). Nuestra paciente presentó en el momento agudo una plaquetopenia severa con valores de 10.000 plaquetas.

La prevalencia de aneurismas múltiples varia según la población de estudio siendo superior en las series japonesas. La prevalencia real en muchos medios como el nuestro es desconocida. Owada et al. encontró una proporción de aneurismas múltiples en pacientes con LES de 31.6% en una revisión de la literatura japonesa

al respecto (102). En nuestro revisión, encontramos una proporción ligeramente superior a la esperada de 22.2%. La causa de este aumento de la proporción de aneurismas múltiples es desconocida. Únicamente Asai et al. diagnostica una vasculitis transmural al estudiar uno de los aneurismas de un paciente con LES y aneurismas multiples(117).

Los aneurismas en localizaciones atípicas y fusiformes son frecuentes en el LES. Un 22.2% de los pacientes presentan en nuestra revisión aneurismas poco habituales en la población general. En muchas ocasiones, estos aneurismas son un reto tanto para el tratamiento endovascular como para la cirugía, requiriendo la realización de procedimientos poco habituales en el día a día. El paciente que describimos presentaba un aneurisma muy poco frecuente localizado en una rama M4 de la ACM y que requirió un "*trapping*" de la arteria para poder ser tratado.

3.2.1.2. Actitud del clínico ante el paciente con HSA y LES

Los hallazgos descritos previamente tienen una importante implicación clínica. El médico debe ser consciente del mal pronóstico que presentan los pacientes con HSA negativa y LES, aún tratándose de pacientes jóvenes. A diferencia de lo que ocurre en la población general, la HSA idiopática en el LES es una enfermedad con una elevada morbimortalidad. En lo referente a la mayor presencia de aneurismas múltiples en pacientes con LES, el clínico debe concentrarse en la búsqueda de otros aneurismas que puedan pasar desapercibidos inicialmente. La búsqueda minuciosa de otras lesiones evitará tener que repetir procedimientos para tratar aneurismas que no se habían detectado de forma inicial.

La realización de una angiografía de los cuatro vasos intracraneales, si bien de forma habitual es mandatoria, no tiene ninguna excusa de no realizarse en estos pacientes. La inspección minuciosa de la arteriografía no sólo debe servir para la búsqueda de aneurismas múltiples, sino también de aneurismas en localizaciones atípicas que puedan por tanto pasar desapercibidos. Estos aneurismas, además, presentan morfologías no saculares en una mayor proporción que en la población general(118-120), lo que representa un reto quirúrgico para el neurocirujano como

para el radiólogo intervencionista. Los aneurismas fusiformes y aneurismas tipo "*blister*" son considerados aneurismas complejos, aumentan el riesgo de complicaciones quirúrgicas y de posibilidad de muerte o secuelas neurológicas severas(119). En estos pacientes, el beneficio que nos aporta la arteriogarfía es mucho mayor que en otros casos de HSA. Deberemos realizar esta prueba de imagen siempre que sea posible por su importancia como indicador de valor pronóstico.

La inflamación vascular parece no ser la única causa de las peculiaridades de los aneurismas en el LES; podrían estar implicados los tratamientos inmunosupresores y la prematura aterosclerosis que sufren estos pacientes(92). La vasculitis focal como causa relacionada con la enfermedad sólo ha sido descrita en tres ocasiones en la literatura(94,96,117). Al examinar una de las cúpulas de los aneurismas intervenidos en nuestro centro no observamos un proceso vasculítico en la pared. De todas maneras, y en una enfermedad que cursa a brotes, puede ser que estemos evaluando el resultado de un proceso inflamatorio focal ya finalizado. Nuestro hallazgo, orienta a posibles causas multifactoriales en la etiología de los aneurismas en esta enfermedad, en la que la vasculitis tendría un papel aún no esclarecido.

Futuros estudios deben permitir reforzar estos hallazgos. Centrarse en la fisiopatología que permita determinar cuál de las causas tiene una mayor implicación en un determinado individuo. Deberíamos poder estudiar más muestras anatomo-patológicas de cúpulas aneurismáticas para poder saber si presentan un proceso vascultítico o lo han presentado en un pasado; sobretodo en pacientes con aneurismas múltiples y lesiones atípicas. El neurocirujano debe prestar atención delante de una pacientes con HSA y LES. Debe conocer sus características y su mal pronóstico funcional.

3.2.1.3. Valor pronóstico del hematoma subdural en la HSA aneurismática

Los Hematomas subdurales relacionados con un rotura aneurismática representan entre un 2-10% de todos los pacientes con HSA(121,122). La mayoría de casos son

debidos a la rotura de un aneurisma de la ACoP; es conocido que estos aneurismas pueden proyectar la cúpula hacia el espacio subdural con más facilidad que otros(123-125). En nuestra serie, la mayoría de los HSDa fueron a consecuencia de un aneurisma en esta localización. Otras causas menos frecuentes de hematomas subdurales son secundarios a aneurismas gigantes que afloran por las cisuras(126,127) o a aneurismas que han producido un hematoma intraparenquimatoso que termina drenando al espacio subdural(124,128); en este último caso hablaremos de "laceración cortical aneurismática".

En los pacientes con HSDa se recomienda el tratamiento quirúrgico del aneurisma al permitir la misma cirugía de clipaje aneurismático la evacuación del hematoma y la realización de una craniectomía descompresiva primaria si es necesario(129). Sorprende el elevado número de pacientes de nuestra serie que presentaban un grosor del HSDa igual o inferior a 10 mm, justo el límite de lo considerado quirúrgico en otras patologías como el traumatismo craneoencefálico(130,131). Futuros trabajos deberían esclarecer si en el caso de los HSDa se debería considerar quirúrgico un grosor de hematoma subdural menor que el establecido para los pacientes traumáticos.

Es controvertido el pronóstico funcional de los pacientes con HSDa; la mayoría de literatura se basa en pequeñas series(124,127,132-138) donde se mezclan pacientes con buen y mal estado de conciencia inicial(139) con conclusiones contradictorias. La revisión más extensa de la literatura incluye 10 articulos con 111 pacientes donde sólo un 23% con mal estado neurológico inicial consiguen un buen pronóstico funcional(140). Westermaier et al. obtiene al realizar un tratamiento quirúrgico agresivo de entrada un sorprendente buen resultado funcional en cinco de siete casos en pacientes comatosos(137). Nuestros resultados, si excluímos una elevada mortalidad postoperatoria del 38,5%, también son esperanzadores obteniendo un 62,5% de pacientes con buen pronóstico funcional entre los supervivientes. No observamos un peor pronóstico funcional de los pacientes con HSDa si los comparamos con los que presentaron HSA de forma aislada. Nuestros resultados reflejan que la presencia de un hematoma subdural en un paciente en

coma con HSA no debería considerarse como un predictor de mal pronóstico funcional por si mismo.

De todas formas, existe un subgrupo de pacientes que si que parecen presentar una peor evolución clínica. Es el caso de los pacientes en los que el HSDa se asocia a un hematoma intraparenquimatoso (laceraciones corticales aneurismáticas). Describimos 4 de estos pacientes en los que sólo uno de ellos presentó un buen resultado funcional. Estos pacientes podrían tener una evolución similar a los "estallidos de lóbulos" que observamos en los pacientes traumáticos(131,141-143).

3.2.2. MAV cerebrales

El tratamiento de las MAVs continúa siendo una importante causa de controversia en el mundo neuroquirúrgico. El estudio ARUBA no ha cubierto las expectativas depositadas en él. Los resultados obtenidos en contra del tratamiento de las MAVs no rotas y a favor la historia natural se fundamentan sobre severos sesgos metodológicos y de selección(69,74-77). Es poco probable que estos resultados cambien la actitud de los cirujanos, por otra parte crean indecisión con respecto al tratamiento óptimo a seguir entre los médicos de otras especialidades. La incertidumbre en el tratamiento de las MAVs no rotas sigue y se extiende a las rotas; la pobre casuística al tratarse de una enfermedad minoritaria, la falta de datos sobre su historia natural(68) y los devastadores resultados del tratamiento en algunos casos avivan esta controversia. Los argumentos en contra del tratamiento de las MAVs se basan en justificar el tratamiento como una elevada causa de morbimortalidad en pacientes con un riesgo real no conocido de rotura. Estos autores parecen sugerirnos que debemos permanecer con los ojos cerrados delante el goteo de MAVs que se rompen espontáneamente, argumentando que el tratamiento de las mismas de forma preventiva es una peor opción(75,144).

La mayoría de autores coinciden en la necesidad de tratamientos multidisciplinares en los que la cirugía sigue siendo primordial. Aunque el tipo de tratamiento puede

presentar dudas, el lugar que ocupa cada especialista habitualmente queda bien definido. La clave en el tratamiento de las MAVs, sigue a día de hoy, en poder discernir entre aquellos pacientes que se benefician del tratamiento de los que no. Para ayudar al clínico en esta toma de decisiones es imprescindible conocer más a fondo la historia natural de la enfermedad y focalizarnos en indicadores que nos permitan predecir la evolución clínica de los pacientes que se van a tratar.

3.2.2.1. Indicadores de mal pronóstico en relación con el sangrado

La rotura intraoperatoria de las MAVs es una complicación poco común (5%) con una frecuencia inferior a la rotura intraoperatoria de los aneurismas (7-15%)(88,89). Los aneurismas rotos muestran una fragilidad en la pared arterial que hace que puedan romperse con mayor facilidad durante la disección a diferencia de las MAVs(145) que parecen no tener un mayor riesgo de rotura al relacionarlas con el sangrado previo. A diferencia de la cirugía aneurismática, en donde los pasos quirúrgicos a seguir en frente a una rotura intraoperatoria están bien establecidos, la rotura intraoperatoria de una MAV requiere capacidad de improvisación por parte del neurocirujano. En nuestra serie se observa un peor pronóstico funcional en aquellas MAVs que se rompieron intraoperatoriamente; en un 44% de los pacientes se obtuvo un empeoramiento neurológico posterior a la cirugía. La mortalidad fue muy elevada, siendo un 12,5% en el postoperatorio inmediato y un 12,5% secundaria a la cirugía. Estos datos refuerzan el hecho que la rotura intraoperatoria de las MAVs es una complicación grave y representa un indicador de mal pronóstico en la evolución clínica. Al revisar las causas de rotura intraoperatoria de las MAVs nos encontramos con tres causas principales: El sangrado arterial por arterias perforantes, la invasión del nidus y la oclusión prematura de la vena de drenaje principal.

El sangrado arterial de las arterias perforantes habitualmente proviene del parénquima que rodea la MAV. Estas abigarradas arterias presentan unas paredes dismórficas que dificultan su coagulación. La utilización de microclips es una buena opción en la mayoría de casos. De forma habitual, estas arterias se encuentran

profundas dentro del parénquima y requieren de la resección de tejido sano para una correcta visualización y clipaje. Estos procedimientos deben hacerse en medio de un sangrado profuso, el cirujano debe resecar tejido cerebral con el consiguiente riesgo de nuevos déficits en áreas elocuentes. El sangrado de las arterias perforantes debe evitarse con tratamientos endovasculares previos a la cirugía(146). Aunque el acceso endovascular a estas arterias en ocasiones es difícil, la embolización si es posible puede ayudar a la resección del cirujano evitando que se produzca un sangrado masivo con el correspondiente empeoramiento del pronóstico funcional. El cirujano debe revisar conjuntamente con el neuroradiólogo intervencionista las arterias perforantes que le pueden presentar problemas durante su procedimiento quirúrgico y planificar una embolización previa si el caso lo requiere; también debe tener especial cuidado en aquellos casos de MAVs que limitan con el sistema ventricular y en las que se produce un edema cerebral masivo sin observarse un sangrado en el campo quirúrgico lo explique. Estas arterias pueden romperse y sangrar de forma descontrolada dentro del ventrículo. Se deberá entonces acceder rápidamente al ventrículo para intentar identificar el punto de sangrado y solucionarlo.

Muchas veces la diferencia en el campo quirúrgico entre lo que podría ser un arteria o una vena arterializada no es fácil, la apariencia macroscópica puede ser parecida y puede prestar a errores. Un sangrado repentino en relación a la oclusión de un vaso debe hacernos pensar en una oclusión prematura de la vena de drenaje(147-149). En estos casos, la lesión cambiará a un aspecto tenso por un aumento de la presión intranidal y se iniciarán sangrados espontáneos en sitios lejanos al lugar de trabajo. Existe un intervalo indeterminado entre la oclusión de la vena de drenaje principal y la rotura definitiva de la MAV. El cirujano debe percatarse de esta situación; la vena de drenaje ya no es funcionante y el objetivo principal pasará a ser el de finalizar la cirugía lo más rápidamente posible cogulando las aferencias principales antes de que se produzca la rotura.

La violación del nidus es la complicación menos frecuente en las roturas intraoperatorias, sólo fue registrada en 4(13%) pacientes. Trabajar muy cerca de la

MAV para evitar tejido elocuente puede llevarnos a esta complicación(150). En MAVs difusas o en las que han recibido radiocirugía previamente los márgenes con el parenquima cerebral son menos evidentes(80,151,152). En ocasiones, el agujero realizado en el nidus malformativo puede controlarse mediante la coagulación bipolar; aunque esta técnica puede ser contraproducente e incluso agrandar el propio defecto. Si se produce un sangrado descontrolado, la presión de una de las espátulas del separador con ayuda de celulosa oxidada puede darnos una tregua y permitirnos tener libres las manos para poder finalizar la cirugía.

Estas dos últimas causas estudiadas, tiene una relación directa con la experiencia del cirujano. En nuestra revisión describimos un aumento de roturas intraoperatorias durante los primeros cinco años de experiencia, posteriormente la tasa de roturas pasa de un 9% a un 3%. Todos los pacientes en los que la causa fue debida a una violación del nidus y el 70% de los pacientes en los que se produjo una oclusión prematura de la vena de drenaje se produjeron durante este primer periodo, indicando que estos errores técnicos pueden ser corregidos con la experiencia. La rotura de la MAV por el sangrado de arterias perforantes se encuentra más distribuida en el tiempo. A diferencia de las otras causas descritas previamente, la rotura de las arterias perforantes no tiene una relación tan clara con la falta de experiencia del cirujano. En muchas ocasiones, estas arterias profundas no pueden embolizarse previamente y el cirujano tiene que hacer la cirugía a sabiendas del elevado riesgo de sangrado que representará la manipulación de estos vasos.

En nuestro trabajo, demostramos que la rotura intraoperatoria de las MAVs empeora el pronóstico funcional de estos pacientes. Analizamos las tres causas principales, dos de las cuales son evitables si la cirugía la realiza un neurocirujano vascular con experiencia. A pesar de todo, la causa más frecuente de rotura intraoperatoria es por un sangrado masivo debido a las arterias perforantes, normalmente en la parte final de la resección de la MAV. Aconsejamos el estudio previo de estas arterias y siempre que se pueda con un riesgo aceptable, la embolización de las mismas. Las arterias perforantes no han formado parte de las diversas clasificaciones que existen para determinar el riesgo quirúrgico de las

MAVs(79,80), si que han sido utilizados otros indicadores como la presencia de un nidus difuso, que en nuestra revisión es el doble de frecuente que en el resto de MAVs. Creemos, que el cirujano debe tener presente estas arterias; el estudio de las mismas debe formar parte de sus decisiones prequirúrgicas e intraoperatorias. La presencia de arterias perforantes de gran calibre que no pueden ser tratadas endovascularmente puede ser un factor que nos haga decidirnos para el no tratamiento de una MAV.

3.2.2.2. Localización de las MAVs como indicador pronóstico

Al analizar el riesgo de sangrado intraoperatorio según la localización de la MAV observamos que un 25% de la roturas intraoperatorias tienen lugar en MAVfp mientras que un 78% son supratentoriales. Encontramos una mayor tasa de roturas intraoperatorias en MAVfp (7/95; 7%) si lo comparamos con las supratentoriales (25/496; 5%), aunque estos resultados no son significativos (p =0,36).

Al analizar los datos de nuestro hospital, nos centramos en el estudio de las MAVfp. Obtenemos de forma parecida al resto de la literatura un mayor riesgo de presentación con hemorragia. Un 82% de las MAVfp se presentaron clínicamente con sangrado a diferencia que las supratentoriales que fue sólo en un 38,8%. No se conoce la causa de la mayor tendencia al sangrado de la MAVfp, incluso hay autores que sugieren que la incidencia de hemorragia es parecida a las supratentoriales y que la mayor presentación con sangrado sería una falsa percepción clínica, al ser la hemorragia la principal causa de debut; permanecerían pues un gran número de MAVfp asintomáticas no diagnosticadas en la población general(62). Se han expuesto distintas hipótesis que podrían explicar este mayor sangrado como: la mayor predisposición a presentar un drenaje profundo, causas hemodinámicas e incluso el menor tamaño del nidus(153). A parte de un posible mayor riesgo de resangrado, las MAVfp con hemorragia presentan un peor estado de conciencia inicial si se compara con las MAVsp. La menor tolerancia volumétrica al sangrado de fosa posterior, la posible afectación del tronco cerebral y la mayor propensión a la hidrocefalia podrían estar entre las causas de la presencia de un

mayor número de pacientes en coma. En nuestro estudio, 16(32%) pacientes se presentaron en coma (GCS≤8).

La alta mortalidad (20,3%) de los pacientes con MAVfp se explica en gran parte por la mayor presentación con sangrado y el mal estado neurológico (GCS) inicial en el que llegan a nuestros centros. A pesar de todo, aquellos pacientes que sobrevivieron presentaron a los 6 meses un buen estado funcional. Un 70% de la MAVfp tenían un GOS de 4 y 5 a los 6 meses del diagnóstico.

3.2.2.3. La hidrocefalia como indicador pronóstico

La hidrocefalia es un complicación frecuente en los pacientes afectos de una MAVfp (30/60; 50%). Esta complicación no sólo es más frecuente en las MAVfp que en las MAVsp, además es mayor la necesidad de tratamiento con colocación de un drenaje o una derivación permanente. El bajo nivel de conciencia es por si mismo un indicador de mal pronóstico en la gran mayoría de patologías neuroquirúrgicas. Aunque la mayor presencia de hidrocefalia en estos pacientes podría explicar en parte la mayor presentación en coma de los pacientes con MAVfp que sangran(23). Diez pacientes se presentaron en coma con hidrocefalia sin afectación clínica ni radiológica de tronco encefálico, a todos ellos se les trató la hidrocefalia. El resultado funcional fue de un GOS de 5 en 4 pacientes, GOS de 3 en 3 pacientes y GOS de 1 en 3 pacientes. Observamos que un 40% de estos pacientes sólo presentaban una mínima afectación neurológica en el seguimiento a los 6 meses. La hidrocefalia podría ser una causa reversible de coma en algunos pacientes con MAVfp. El tratamiento precoz de esta complicación podría aumentar el número de pacientes con buen resultado funcional en el seguimiento tardío.

Al estudiar distintos patrones de sangrado, observamos que los pacientes con hemorragia intraventicular en el TC inicial tienen una probabilidad aumentada de presentar hidrocefalia. Presentar un hematoma intraparenquimatoso no aumenta esta probabilidad si no va acompañado de hemorragia intraventricular.

Conclusión

Conclusión

- En los pacientes con HSA y LES, describimos tres patrones no previamente descritos en la literatura: Mayor presencia de aneurismas múltiples, aneurismas fusiformes/blister o en localización atípicas y HSA angiográficamente negativa.
- Describimos una mala evolución clínica en aquellos pacientes afectos de LES que han presentado una HSA angiograficamente negativa, comparando la benignidad del proceso en pacientes no afectos por esta enfermedad.
- La presencia de un hematoma subdural en un paciente en coma con HSA no debería ser considerado como un predictor de mal pronóstico funcional por si mismo.
- Describimos la “laceración cortica aneurismática” como una nueva entidad que presenta un peor pronóstico funcional con semejanzas al “estallido de lóbulo” observado en los pacientes con traumatismos craneoencefálicos.
- Demostramos que la rotura intraoperatoria de las MAVs es un factor de mal pronóstico empeorando el estado funcional a largo término.
- A partir de una serie extensa de MAV tratadas quirúrgicamente, determinamos tres causas relacionadas con esta complicación; dos de ellas evitables con una correcta planificación prequirúrgica.
- Corroboramos que existe una clara curva de aprendizaje en el tratamiento quirúrgico de las MAVs cerebrales, con una significativa disminución de esta complicación en relación a la experiencia del cirujano.
- Confirmamos que las MAVfp presentan un mayor riesgo de rotura espontánea y un mismo riesgo de rotura intraoperatoria que las MAVsp.
- Demostramos que la incidencia de hidrocefalia es superior en las MAVfp y podría explicar el peor estado neurológico inicial en estos pacientes.
- Demostramos que la incidencia de hidrocefalia es superior en aquellos pacientes que prestan un patrón con sangre intraventricular.

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