The Spetzler-Martin Grading System and Management of Patients with Intracranial Arteriovenous Malformation in a Tertiary Referral Hospital

[1]**SA Al-Edrus,** [1]**SN Suhaimi,** [2]**AR Noor Azman,** [3]**AZ Latif &** [4]**M Sobri**
[1]*Department of Imaging, Faculty of Medicine and Health Sciences*
Universiti Putra Malaysia,Serdang, Selangor, Malaysia
[2]*Department of Neurosurgery, Hospital Sultanah Aminah, Malaysia*
[3]*Department of Neurosurgery, Hospital Kuala Lumpur, Malaysia*
[4]*Department of Radiology, Faculty of Medicine, Universiti Kebangsaan Malaysia*

ABSTRACT

Introduction: An arteriovenous malformation (AVM) is an abnormal collection of blood vessels in which arterial blood flows directly into the draining vein without the normal interposed capillaries. The Spetzler-Martin grading system has been widely accepted worldwide to estimate the pretreatment risks and predict the outcome of patients with intracranial AVM. In Malaysia, we still do not have the baseline data of this grading system. **Methods:** A total of 33 patients from a tertiary referral hospital diagnosed with intracranial AVM based on neuroimaging findings over a 4-year period were studied. Medical records were traced and neuroimaging findings were analysed. The AVMs were graded according to the Spetzler-Martin grading system and Fisher's exact test was used to assess statistical difference between the grades of the AVM and management plan for the patients. **Results:** Four patients were graded as Grade 1, 9 patients as Grade II, 10 patients as Grade III, 6 patients as Grade IV and 4 patients as Grade V. Ten patients were treated conservatively; six patients underwent surgery and embolisation respectively. Four patients underwent radiosurgery and a combination of embolisation while surgery and radiosurgery were given to 5 patients and 2 patients respectively. Statistically significant difference ($p=0.016$) was found between the Spetzler-Martin grading system and the management of intracranial AVMs. **Conclusion:** The management decision was not made based on the grading of the AVMs. It is recommended that all AVM patients be routinely graded according to this system prior to treatment.

Keywords: Intracranial arteriovenous malformation, Spetzler-Martin grading

INTRODUCTION

An AVM is an abnormal collection of blood vessels in which arterial blood flows directly into the draining vein without the normal interposed capillaries. A contrast enhanced computed tomography (CT) of the brain is often the initial imaging modality used to diagnose intracranial AVM. However, magnetic resonance imaging (MRI) gives more precise information to assess the size and anatomical localisation of the AVM nidus, although it is still difficult to delineate the true nidus. Therefore, 4-vessel catheter angiography with digital subtraction

*Corresponding author: Dr Sharifah Aishah AL-Edrus; Email: *aishahaledrus@gmail.com*

is the most important pre-operative study and remains the gold standard in delineating the vascular anatomy. The precise angio-architecture of an AVM with fast frame angiogram, once correlated with imaging data, leads to appropriate grading and pre-operative planning.

The Spetzler-Martin grading system has been widely accepted worldwide to estimate the pretreatment risks and predict the outcome of patients with intracranial AVM. The proper management of AVM is complex and requires an understanding of their natural history and effect on the cerebral circulation and metabolism. It requires proper and meticulous pre-treatment planning, multimodality treatment options and modern post-operative and post-procedural care.

Currently, the majority of neuro-surgeons and interventional neuro-radiologists use the Spetzler-Martin grading system to estimate the pretreatment risk and predict the outcome of patients with intracranial AVMs.[1] In this grading system, AVMs receive a grade of I through V, which corresponds to a point scale depending on the size of the lesion, the eloquence of adjacent brain tissue and the pattern of venous drainage. Higher-grade lesions, being more difficult to treat, have higher degrees of patient morbidity and mortality associated with their treatment.

In Malaysia, we still do not have the baseline data of this grading system. AVM patients are not routinely graded using the Spetzler-Martin grading system even though it is already widely accepted. In this study, we focused on the management decision of intracranial AVMs in correlation with this grading system.

METHODS

This was a retrospective study carried out at the Diagnostic Imaging Unit and the record office of Neurology and Neurosurgery Department. Universal sampling method, where all patients above 12 years old, who were diagnosed with intracranial AVMs over a 4-year period based on neuroimaging findings, were included. The medical records and neuroimaging of these patients were traced and relevant information was recorded in a proforma. Two radiologists reviewed the neuroimaging images for the Spetzler-Martin grading. The proforma included:

1. Patient's demographic data: name, identification number, sex, age and race.
2. Clinical presentation and final diagnosis.
3. Neuroimaging findings from Magnetic Resonance Imaging (MRI) and Computed Tomography (CT): site of the lesion (eloquent or non-eloquent part of the brain), types of haemorrhage, types of hydrocephalus, cerebral oedema and calcification.
4. Cerebral angiogram findings: size and site of the lesion, venous drainage and arterial supply of the nidus of the AVM.
5. Management of the patient: surgery, radiosurgery, embolisation, combined therapy or conservative treatment.

Spetzler-Martin Grading of Cerebral AVMs

i. *Size of the lesion*: The size of the AVM was measured by taking the maximum transverse diameter of the nidus. It was determined from the angiogram or MRI and was classified into 3 categories: small (< 3cm), medium (3–6cm) and large (>6cm) with 1, 2 and 3 points assigned for each of the category, respectively.

ii. *Site of the lesion*: The site of the lesion was determined from the CT Scan or MRI. It was classified as an eloquent area if the involved area of the brain had an identifiable neurological function, and if injured, will result in a disabling neurological function. It was considered as a non-eloquent area if it involved areas with much more subtle neurological function. In the Spetzler-Martin grading scheme, the areas which are considered eloquent are the sensorimotor, language and visual cortex, the hypothalamus and thalamus, the internal capsule, the brain stem, the cerebellar peduncles and the deep cerebellar nuclei. Areas with less important neurological function, or areas in which injury does not cause permanent disabling deficit, for example, the anterior portion of the frontal or temporal lobes and the cerebellar cortex, are considered as non-eloquent areas. One point was given to involvement of an eloquent area and 0 point for a non-eloquent area.

iii. *Pattern of venous drainage*: The course of the draining vein was determined from the angiogram. It was considered superficial if all the drainage from the AVM was through the cortical venous system. It was considered deep if any or all of the drainage was through the deep veins (e.g. internal cerebral vein). Zero and 1 point was assigned for each category respectively.

The grades of the lesions were derived by summing the points assigned for each category (Table 1).

Table 1. The Spetzler- Martin grading system for intracranial AVM

Graded feature	Score
Size of AVM	
Small (< 3cm)	1
Medium (3–6 cm)	2
Large (>3 cm)	3
Eloquence of adjacent brain	
Eloquent	0
Non-eloquent	1
Pattern of venous drainage	
Superficial only	0
Deep	1

Data were analysed using Statistical Package for Social Sciences (SPSS) version 10.0. The Fisher's exact test was used to determine the difference between the Spetzler-Martin grading system and the management of the patients. The null hypothesis stated that there is no difference between the Spetzler-Martin grading system and the management of patients with intracranial AVMs. Level of significance was set at 0.05 with 95% confidence interval.

RESULTS

Demographic Data

A total of 33 patients was included in this study. The patients' age ranged from 12 to 66 years with a mean age of 33.39 years. Twenty were males (60.6%) and 13 were females (39.4%). The majority of the patients were Malays (n = 20), followed by the Chinese (n = 8), Indians (n = 2) and others (n =3).

Clinical Presentation

Haemorrhage (intraparenchymal, subaracnoid, intraventricular or a combination of any 2 or all) occurred in 39.4% of patients (n=13). Eight out of the 13 patients presented with neurological deficits. Other clinical presentations, such as seizures, were present in 15.2% of the patients, headache in 18.2% and combination of both in 9.1% of the patients.

Size of AVMs

The size of the AVMs ranged between 0.6cm to 8.4cm. Nine AVMs (27.3%) were categorised as small lesions, 20 AVMs (60.6%) were medium sized and 4 (12.1%) were giant AVMs.

Pattern of Venous Drainage

Fifteen patients (45.5%) had superficial venous drainage which involved either the superficial cerebral veins (SCV) or superior (great) anastomotic veins of Trolard or both. Eighteen other patients (54.5%) presented with deep venous drainage that involved either the internal cerebral veins (ICV) or inferior anastomotic veins of Labbe or both.

Location of AVMs

Nineteen patients (57.6%) had AVMs involving the left cerebral hemisphere, 12 (36.4%) had in the right cerebral hemisphere and 2 patients (6.1%) had AVMs in the posterior fossa. Seventeen patients (51.5%) had AVMs located in eloquent areas while 16 patients (48.5%) had in non-eloquent areas.

Determination of Grades

Four patients were categorised as Grade I (12.1%), 9 as Grade II (27.3%), 10 as Grade III (30.3%), 6 as Grade IV (18.2%) and 4 as Grade V (12.1%).

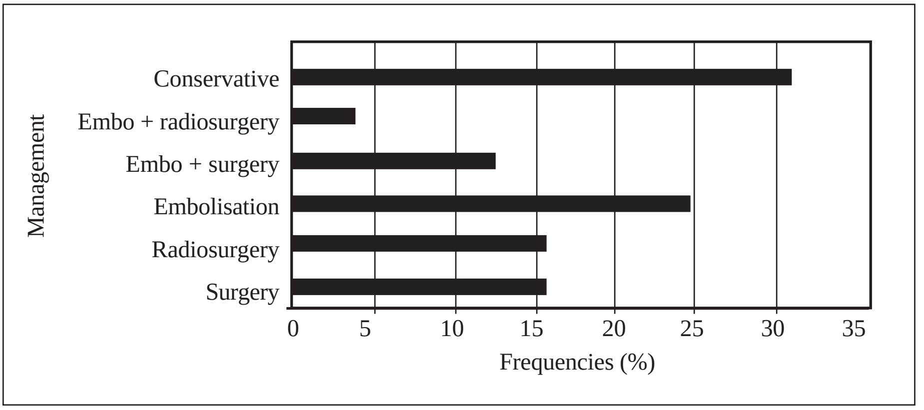

Figure 1. Different management plans for patients with AVMs

Table 2. Different management plans according to the Spetzler-Martin grading system

	Grading of AVM					
	GD I	GD II	GD III	GD IV	GD V	TOTAL
Surgery	3		3			6
Radiosurgery		1	2	1		4
Embolization		2	3		1	6
Embo+surgery		1	1	3		5
Embo+radiosurgery	1			1		2
Conservative		5	1	1	3	10
TOTAL	4	9	10	6	4	33

Management of AVMs

Figure 1 and Table 2 summarises the different management plans for patients in this study.

Association between Grading and Management of AVMs

Fisher's probability exact test was done to determine the difference between the Spetzler – Martin grading system and management of the AVMs and it showed a statistically significant difference (p= 0.016). The management decision was therefore not based on the grading system.

DISCUSSION

AVM affects both sexes equally although our study showed a higher incidence in males. Patients typically present with symptoms during early childhood and may seek medical attention for one or a combination of the following problems: intracranial haemorrhage, seizures, headache, focal neurological deficits, impairment of higher cortical function and bruit. In our study, the majority of patients presented with intracranial haemorhage (39.4%). The decision to proceed with the treatment option is based on the natural history of the disorder combined with the patient's general condition. Ideally, any treatment decision should be taken by a multidisciplinary team composed of the neurologist, neuro-surgeon, radiotherapist and neuro-radiologist.[2] Microsurgery, radiosurgery, embolisation and a combination of these techniques are currently used to treat and reduce neurological morbidity and mortality due to AVMs. Some of the AVM patients are treated conservatively. Whichever treatment is chosen, the only acceptable result for an AVM is an angiographically demonstrated complete obliteration. Treatment-related permanent neurological deficit and death from any method of AVM treatment has a rate of between 0% to 20% with a mean rate of 8%.[3-7]

In 2001, a writing group was formed by the Stroke Council of the American Stroke Association to review published data for intracranial AVMs and to develop practice recommendations regarding epidemiology, natural history, potential treatment strategies and outcomes.[8] These management guidelines have recommended an approach according to the Spetzler-Martin grading. The main recommendations of this group are:

1. For Grades I and II AVMs, surgical excision should be considered as the primary single treatment.
2. For Grades IV and V AVMs, surgery alone is unsuitable.
3. Stereotactic radiosurgery is the preferred single treatment for small (<3cm) Grades I and II AVMs if the vascular anatomy is unsuitable for surgery.
4. Treatment of choice for other suitable Grades II-V lesions is a combined approach to completely eradicate the AVM nidus using embolisation (perhaps repeatedly) prior to surgery or stereotactic radiosurgery.
5. Palliative embolisation (without complete AVM obliteration) may be beneficial for intractable epilepsy refractory to antiepilectic treatment, or when a progressive neurological deficit is thought to be due to high flow or venous hypertension.

In our centre, lesions are treated on a case-by-case basis. When comparing the above guidelines to the management decisions of our patients, some discrepencies were noted. In our patients, surgery was performed not only in Grade I AVMs but also in 3 patients with Grade III AVMs. Radiosurgery was also performed on 2 patients with Grade III and one patient with Grade IV AVM. Embolisation, either alone or combined with surgery or radiosurgery, were performed in all AVM grades. Therefore, the above guidelines, although reflecting the current practice in North America, are not strictly followed and may be less representative of practices in other parts of the world.

Surgery was performed in a number of patients in our centre possibly because of the availability of experienced neurosurgeons as well as established neurosurgical facilities. With newer improved microsurgical techniques, disabling post-surgical complications have seen a downward trend over recent years and the reliability of the Spetzler- Martin grading system for the pre-operative surgical risk and the development of permanent neurological disability following surgery have been proven.[9-11] Embolisation, being the newer treatment method for intracranial AVM is gaining popularity but has only been available in our centre in recent years. The number of procedures done has been limited by the high cost of the procedure which includes endovascular catheters and thromboembolic material used, as well as a designated neuro-interventional radiologist. At the moment, this mode of treatment is only available in a few centres in Malaysia with trained neuro-interventional radiologists and post-procedural intensive care and neurosurgical backup. Outcome of embolisation therapy has improved over the last decade with treatment related permanent neurological disability and death rate ranging between 4 to 18%,[3] depending on the embolic material used. Although the Spetzler-Martin grading system was initially designed and validated to predict surgical treatment outcome, mixed results were found in predicting the treatment outcome following embolisation with some authors finding it helpful [12,13] and others who did not.[14] The aim of embolisation is to obliterate or reduce the size of the AVM and is frequently followed by surgical removal or radiosurgery.

Although helpful and useful in determining the surgical risk, it must be noted that other important morphologic characteristics of the AVM such as the type and number of arterial feeders and draining veins and the presence of intranidal or feeder aneurysms are not included in the grading system. These findings may contribute significantly to the treatment risk although they have not been studied in a population large enough to be identified as poor outcome predictors. Therefore, clinicians facing therapeutic decisions in AVM patients, including treatment risks may be left with the notion that larger AVMs with deep drainage in difficult regions of the brain may bear a high treatment risk.

Based on our study results, there was a significant difference between the management decision for our AVM patients and the Spetzler-Martin grading system. Although the grading system was designed and validated to predict surgical outcome, it has also been evaluated in the combined management of AVMs and clinical deterioration due to treatment has been shown to increase according to the grade of the AVM. The limitation of our study was there was no follow-up on the clinical outcome of each patient following each management decision. Therefore, comparisons could not be made regarding the clinical outcome in patients who were treated in accordance to the recommendation for each AVM grading and those who were not.

A further follow-up study needs to be done to determine treatment related morbidity and mortality in our local population and its association with the Spetzler-Martin grading system. Clinicians should be made aware of the grading system in the pretreatment evaluation of patients with intracranial AVMs so that their patients are able to better understand their treatment risks and the prognostification of their disease.

CONCLUSION

There is a significant difference between the Spetzler-Martin grading system and the management of intracranial AVMs in our centre. It can be seen that the management decision was therefore not based on the grading of the AVMs. It is recommended that all AVM patients be routinely graded according to this system prior to treatment.

REFERENCES

[1] Spetzler RF, Martin NA. A proposed grading system for arteriovenous malformation. J. Neurosurg 1986; 65:476–483.

[2] R Al-Shahi, Warlow C. Arteriovenous malformations of the brain: ready to randomize? J. Neurol. Neurosurg. Psychiatry 2005;76: 1327–29.

[3] The Arteriovenous Malformation Group. Arteriovenous malformation of the brain in adults. N Engl J Med 1999; 340: 1812–1818.

[4] Paulsen RD, Steinberg GK, Norbash AM, Marcellus ML, Lopez JR, Marks MP. Embolisation of rolandic cortex areteriovenous malformation. Neurosurg 1999; 44: 479–484.

[5] Fritzel RT, Fischer WS. Cure, morbidity and mortality associated with embolisation of brain arteriovenous malformations; a review of 1246 patients in 32 series over a 35-year period. Neurosurg 1995; 37:1031–1039.

[6] Wilkholm G, Lundqvist C, Svendsen P. Transarterial embolisation of cerebral arteriovenous malformations: improvement of results with experience. AJNR Am J Neuroradiol 1995; 16: 1811–1817.

[7] Mast H, Koennecke HC, Meisel J, Osipor A, Hartmann A, Lasjaunias P, Pile-Spellman J, Hacein-Bey L, Young YL, Mohr JP. Therapy of cerebral arteriovenous malformation. Nervenarzt 1998; 69: 287–295.

[8] Ogilvy CS, Stieg PE, Ewad I, Brown RD Jr, Kondziolka D, Rossenwasser R, Young WL, Hademenos G. Recommendations for the management of intracranial arteriovenous malformations:a statement for healthcare professionals from a special writing group of the Stroke Council, American Stroke Association. Stroke 2001; 32: 1458–1471.

[9] Hamilton MG, Spetzler RF. The prospective application of a grading system for arteriovenous malformations. Neurosurgery 1994; 34: 2–6.

[10] C. Schaller, J. Schramm, D. Haun. Significance of factors contributing to surgical complications and to late outcome after elective surgery of cerebral arteriovenous malformations. J Neurol Neurosurg Psychiatry 1998; 65: 547–554.

[11] Hartmann A, Stapf C. Hofmeister C, Mohr JP, Sciacca RR, Stein BM, Faulstich A, Mast H. Determinants of neurological outcomes after surgery from brain arteriovenous malformation. Stroke 2000; 31: 2361–2364.

[12] Steinberg GK, Lane B, Marks MP. Endovascular therapy for intracranial vascular lesions. West J Med 1990; 153: 542–543.

[13] Henkes H, Nahser HC, Berg-Dammer E, Weber W, Lange S, Kuhne D. Endovascular therapy of brain AVMs prior to radiosurgery. Neurol Res 1998; 20: 479–492.

[14] Hartmann A, Pile-Spellman J, Stapf C, Sciacca RR, Faulstich A, Mohr JP, Shumacher HC, Mast H. Risk of endovascular treatment of brain arteriovenous malformations. Stroke 2002; 33: 1816–1820.