

Multidisciplinary Approach for the Management of Brain Arteriovenous Malformations

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ABSTRACT

The contemporary management of complex brain arteriovenous malformations (AVMs) often requires a multidisciplinary approach, including embolization, microsurgical resection, and stereotactic radiosurgery (SRS).

Objective: *The main Objective of this study was to evaluate the significance of multimodality treatment of AVMs.*

Material and Methods: *The data for this study was collected from the four years audit of surgical management of Arterio-venous Malformation in King Faisal Specialist Hospital and research center Jeddah, from year 2014 to 2017 which include 24 patients with brain AVMs.*

Results: *In our study equal no. of male and female patients were recorded i.e. 12 patients in each group (50.00%). Seventeen (70.83%) patients were found in the first three decades of life indicating that the disease affects the younger age group. Main presenting complaint was seizures that was noted in 16 patient (66.66%) while headache was present in 15 patients (62.50%) followed by other signs and symptoms. Treatment of the patient was tailored according to the type of AVM its grade and the best possible way to take the complex disease. It included combination of pre-surgical or post-operative embolization, Complete or partial surgical resection of the lesion and stereotactic radiosurgery. The combination of these modalities were employed according to the situation of the patient to keep the plan of treatment safe, affective and ending finally in exclusion of the disease from the brain circulation. Pre-surgical embolization was successfully done in 7(29.16%) patients while surgical resection was done in all patients at different stages of treatment. With use of multidisplanary approach in this series; Improvement was seen in 16 (66.66%) of the patient, 6 (25.00%) revealed no improvement while one patient (04.16%) was deteriorated.*

Conclusion: *It was concluded that tailoring the treatment plan and use of multidisciplinary Approach for the management of Brain AVM depending upon their initial grades according to Spetzler Martin grading system results in maximum favorable results.*

Keywords: *Arteriovenous malformation, Embolization of AVM, Radiosurgery of AVM, Microsurgical resection of AVM.*

INTRODUCTION

Arteriovenous malformations (AVMs) of the brain is one the rare and complex vascular lesions that can causes significant morbidity and mortality, but with recent technical and technological advancement in clinical specialties of neurosurgery, endovascular surgery, and radiosurgery had improved patient

outcomes. Multidisciplinary approach for the management of brain AVMs had improved prognosis for patients. Unlike other pathologies of brain, for which therapeutic management from these different disciplines may be competitive, and management recommendations may be controversial, but with brain AVMs it provide an offers to works in collaboration

with these clinical specialists and propose integrated treatment strategies.

Cerebral AVMs are mainly considered as the disease of relatively younger age group affecting the initial four decades of life. The typical presentation of AVM include hemorrhage, seizures, neurological deficits, or headaches.¹ Intracranial AVMs have an annual hemorrhage rate of 1 to 4% and a re-hemorrhage rate of 6 to 18% in the year after first episode of hemorrhage. The morbidity and mortality rates associated with AVM rupture are as high as 53 to 81% and 10 to 18%, respectively.¹⁻⁴

Spletzler – Martin grading system is usually used to access the possible layout of treatment plan that may be either sole or combination of three modalities i.e. microsurgical resection, embolization and stereotactic radiosurgery. Preoperative embolization can be considered for high-grade AVMs, or as a targeted treatment for portions of the AVM difficult to deal with at surgery. Similarly SRS is alone a feasible option when the size of AVM is smaller than 2cm and located in deep and eloquent areas. Microsurgical resection is the best option mainly in Grade I and II AVMs where they can excised completely with producing any new neurological deficit but in complex AVM it is always helpful to reduce the circulation with pre-operative embolization then excise the nidus to the maximum extent and still left over portion can be subjected to SRS. The main goal behind all this exercise is to stay safe and treat the complex disease using the collective wisdom of Surgeon, Radiologist and Interventional Radiologist. This presents a artistic and strong combination of three disciplines focusing on the common target and eliminating the disease in more effective and safe manner. Multidisciplinary approach was individualized while considering the sum of procedural risks of each treatment.⁵

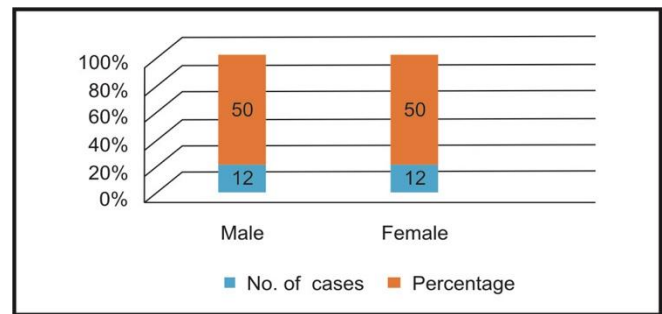
MATERIAL AND METHODS

Clinical data was obtained from the four years audit (2014-2017) of surgical management of Arteriovenous Malformation in King Faisal Specialist Hospital and research center Jeddah, All the patients suffering from this disease were analyzed. The patient treated conservatively were excluded from study and all those were included where microsurgical resection was done with or without combination of pre or post operative embolization and Steriotactic radiosurgery. A total of 24 patients with brain AVMs were recorded in 4 years that fulfilled our criteria. Demographics,

pre-operative Sign/symptoms, characteristics of AVMs and their grading was done using Spletzler-Martin Classification, investigation, treatment modalities, post-op outcomes were studied and statistically analyzed. At the end of study we concluded the strategy that was the best in our opinion and which we are following in our institution for the management of Complex Brain AVMs.

RESULTS

Between 2014 and 2017, 24 patients were included in the study that fulfilled the inclusion criteria. The Gender distribution was equal in our series we recorded 12 (50.00%) patients each in both male and female group and with ratio of 1:1 (Graph 1).



Graph 1: Gender Distribution of Series.

In age distribution the maximum cases were found in the first three decade of life and the no. remained 17 (70.83%) with mean age 23 years. Eight cases (33.33%) were in their 2nd decay of life which indicates the affiliation of disease to the younger age groups. The age diversity in our series can be seen in (Table 1).

Table 1: The age diversity in our series can be seen in.

Age in Years	No. of Cases	Percentage
0 – 10	3	12.5%
11 – 20	8	33.3%
21 – 30	6	25%
31 – 40	3	12.5%
41 – 50	3	12.5%
51 – 60	0	0%
61 – 70	1	4.16%

Initial evaluation of the patients were done with history and complete physical examination and the most common presentation found was seizures, which were noted in 16 patients (66.67%) of cases. These Seizures were either focal or generalized or starting initially as focal and then secondarily generalizing hence giving a possible prediction of the site of origin.⁹ Other most notable presentations was headaches seen in 15 cases (62.00%). In 8 (33.33%) cases focal neurological deficit was present while vomiting was present in 3 cases (12.00%). other sign/symptoms include memory disturbance, visual deterioration, motor aphasia, facial palsy, loss of consciousness, neck stiffness, and sphincter loss. The detailed account of clinical presentation is summarized in Table 2.

Table 2: Clinical Presentation of the patients.

Symptom/Sign	No	Percentage
Headache	15	62.5%
Vomiting	3	12.5%
Neurological deficit	8	33.33%
Seizures	16	66.67%
Miscellaneous	10	
Memory disturbance	2	
Visual deterioration	1	
Motor Aphasia	2	
Facial palsy	1	
Loss of Consciousness	2	
Neck stiffness	1	
Sphincter Loss	1	

In our series 7 (29.16%) cases were present with rupture and 17 (70.83%) cases were diagnosed as unruptured. In ruptured cases maximum cases i-e 6 (85.70%) out of 7 presented with Intracerebral Haematoma while one (14.20%) presented with SDH. Non of the ruptured case presented with Brain stem bleed of Sub arachnoid Haemorrhage (Table 3).

Table 3: Radiological Presentation.

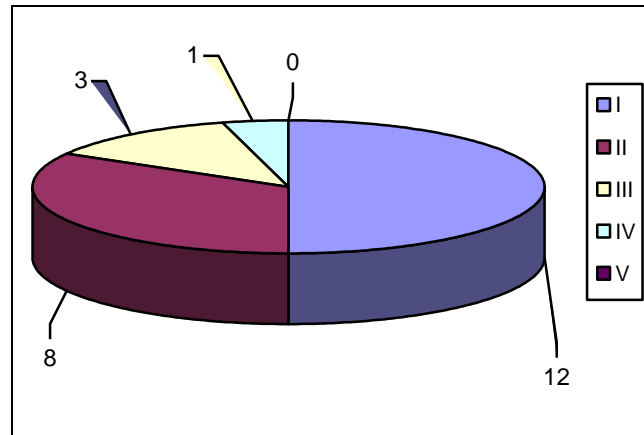
Ruptured	7 (29.16%)	Un-rupture	17 (70.83%)
ICH	6 (85.7%)		
SDH	1 (14.2%)		
SAH	00		
Brain Stem bleed	00		

The diagnostic tools used for final intervention were Cerebral 4 vessels angiogram, MRI with MRA and MRV and CT angiogram. These were employed either in isolated form or where required in combination of more than one investigation to get maximum information before any intervention. Breakdown of definitive investigation in our series can be seen in Table 4.

Table 4: Definitive Investigations Used in Series.

CT Angiogram	MRI/MRA/MRV	Angiography Cerebral
11 (45.83%)	18 (75.00%)	14 (58.33%)

Grading of the patient was done using the famous Spetzler-Martin grading system. It was found in our series that 20 cases (83.33%) were in good grades on arrival while there were 3 cases (12.50%) in Grade III while only one case (04.16%) was categorized in Grade IV. No patient was classified as Grade V. This can be seen in Table 5 and Graph 2.



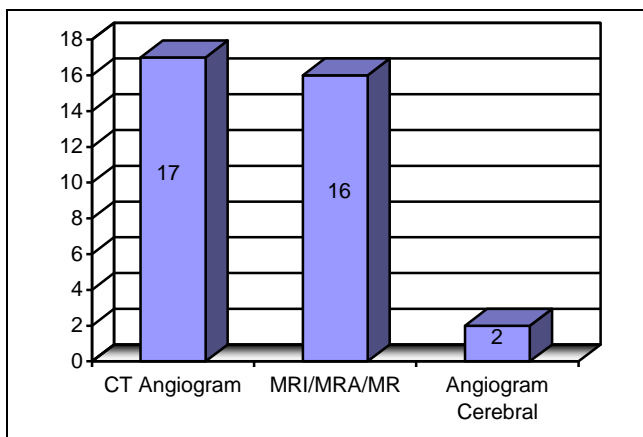
Graph 2: Grading of cases according to Spetzler-Martin Scale.

Table 5: Grading of cases according to Spetzler-Martin Scale.

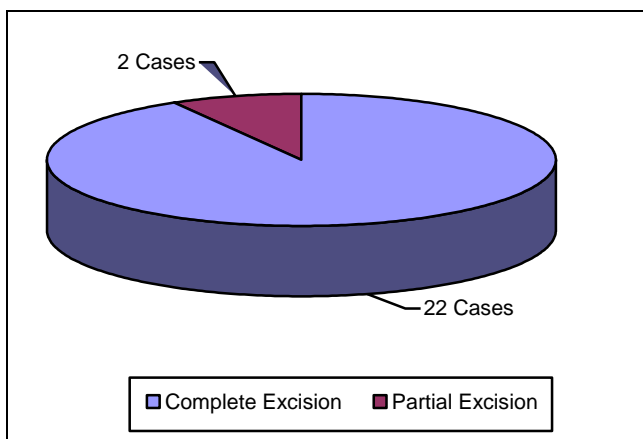
Spetzler- Martin Scale	No. of Cases	Percentage
I	12	50.00%
II	8	33.33%
III	3	12.50%

IV	1	4.16%
V	0	0%

In our series 10 (41.65%) cases underwent pre-operative embolization. In 1 (4.16%) case the procedure was end in failure and in another 2 (08.33%) cases it was complicated with haemorrhage. All the patients underwent microsurgical excision and during surgery the surgical haemorrhage was carefully assessed and it was found that in 21 cases (87.50%) it was < 500 ml and in 3 cases (12.5%) haemorrhage was more > 500 ml. Post operative excision was confirmed with help CT angiogram, MRI, MRA, MRV and Cerebral Angiogram. Graph no. 3. Complete excision was confirmed in 22 cases (91.66%) while partial excision was noted in 2 cases (8.33%) Graph 4. One of these cases with partial excision was re-explored and



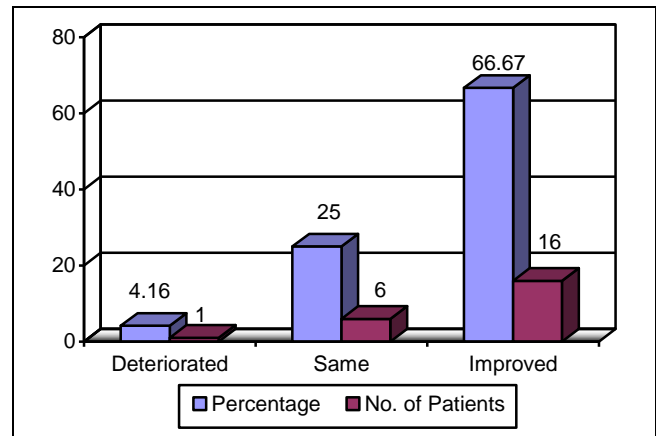
Graph 3: Post-operative Investigations.



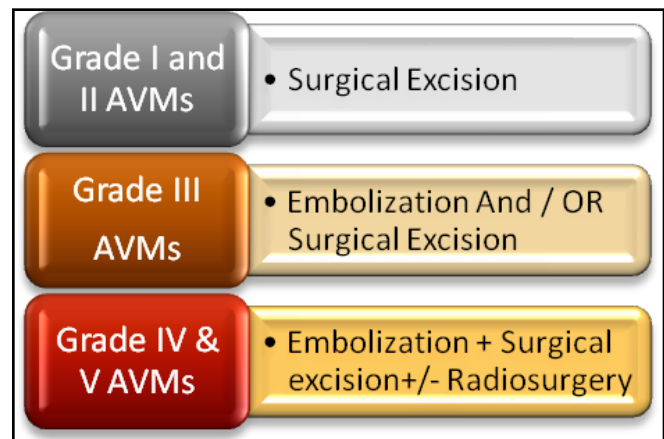
Graph 4: Results of Microsurgical excision of AVMs.

remaining part is excised while the other one was sent for SRS.

Glasgow outcome scale was used to see the outcome of patients at the end of two weeks of surgery. And we found that clinical improvement in 16 cases (66.67%), No obvious change from pre-operative status in 6 cases (25.00%) and one patient further deteriorated (4.16%) after surgery. This is summarized in Graph 5.



Graph 5: Surgical outcome evaluation after 2 weeks of final stage treatment using Glasgow Outcome Scale.



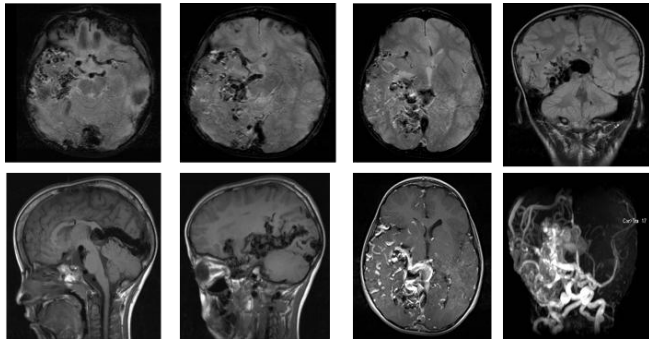
Graph 5: A Case from Our Series.

DISCUSSION

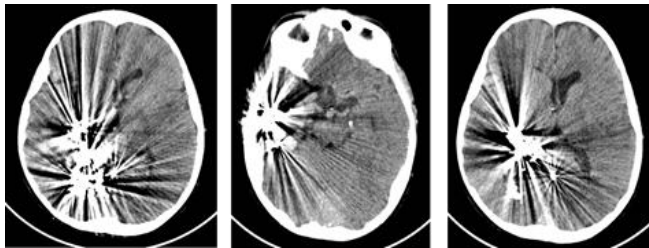
Intracranial AVMs patients are mostly diagnosed before they reached the age of 40, thus it is seldom occur in elderly patients.¹¹ 29.16% of AVMs ruptured resulted in intracranial hemorrhage and subdural hematoma while 70% AVMs were diagnosed earlier

before they were ruptured by clinical and radiological assessment. Most commonly intracerebral hemorrhage occurs, but subarachnoid hemorrhage and intraventricular hemorrhage are also seen as well. Severe vasospasm from AVM-related hemorrhage usually don't occur, but it can occasionally reported.¹² Vascular malformation-related steal phenomena by altering the perfusion of the tissue at the site of the

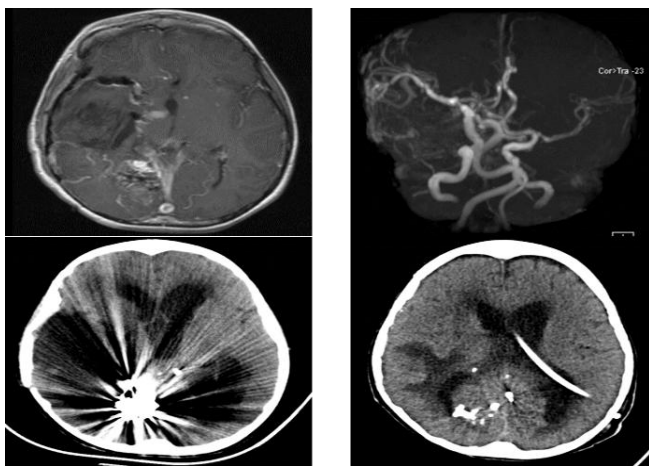
AVM, causing the focal neurological deficits are rarely reported.¹⁰ Intracranial AVMs are mostly diagnosed with of diagnostic radiological studies.



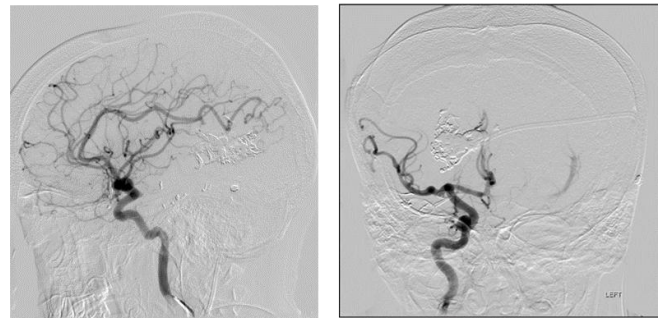
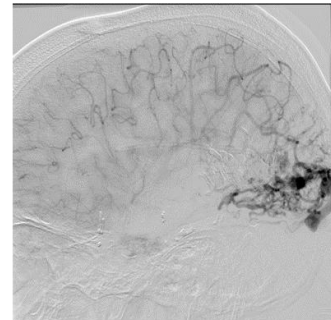
(a) A 7 year old male diagnosed with a complex and huge AVM involving Temporoparietooccipital region, 2010.



(b) Pre-operative Embolization, 2011.



(c) Excision of the temporoparietal portion of AVM. Patient developed Hydrocephalus and Insertion of VP Shunt, 2011.



(d) Residual AVM in Occipital area for which SRS was done and complete resolution of AVM was seen after, 2012 and Complete obliteration, 2013.

Image 1 (a, b, c, d): A perfect example of Multi-disciplinary Approach to AVM.

Computed tomography (CT) without contrast although had low sensitivity, but calcification and hyper densities are noted but enhancement of the lesion can be achieved by administration of contrast.¹³ Magnetic resonance imaging (MRI), shows an inhomogeneous signal void on T1- and T2-weighted sequences, commonly with hemosiderin depicting prior hemorrhage.¹⁴⁻¹⁵ Magnetic resonance angiography is noninvasive imaging technique which provide some information but without detailing factors such as presence of intra-nidal or feeding artery aneurysms, comprehensive data on venous drainage patterns, or subtle AVM nidus characterization.⁹ Cerebral angiography is considered as “gold standard” imaging study for defining feeders, nidus and draining veins of the AVM. Spetzler-Martin Grade (SMG) scale is commonly used for grading brain AVMs.⁶ Points are assigned depending upon the size of nidus, its venous drainage and presence within or away from the eloquent areas. Grade I and II lesions

are generally considered benign surgical lesions with very low incidence neurological deficits, while grade IV and V lesions are frequently reported to have significant surgically induced neurological deficits and Grade VI lesions are inoperable.⁷The risk of subsequent hemorrhage is increased when the brain AVM accompanied with hemorrhage or deep venous drainage or when it is associated with aneurysms, or when the lesion is deeply located.^{7,8}

The gold-standard in brain AVM management is surgery whenever viable. The specific surgical approach along with the use of electrophysiological neuromonitoring were dependent on AVM location and the preferences of treating neurosurgeons.

Although surgical resection remains the standard for the definitive eradication of intracranial AVMs, endovascular embolization before microsurgery has enhance the safety and efficacy of AVM management and prognosis of the patient.¹⁶⁻¹⁷ The result of preoperative embolization of AVMs has shown reduction in operation time and intraoperative bleeding, surgical complications or neurological deficits.¹⁸

The multimodalities of treatment like embolization, microsurgery, and radiosurgery combination are treated in certain types of AVMs. The first decision is between microsurgery, radiosurgery, or observation. The Spetzler-Martin grading system is an essential part of this first decision, with low-grade AVMs (Grades I, II, and III) usually considered for microsurgical resection, and high-grade AVMs (Grades IV and V) usually observed or exposed to multidisciplinary approach. Individual AVM dictate the combination of two or more modalities for the best possible treatment required for its eradication from the circulation. This may require a multistage treatment including multiple sessions of embolization, attempt to remove surgically accessible lesion and followed by Stereotactic Radiosurgery.

Amongst the low-grade AVMs are those in inaccessible locations, highly eloquent locations, and/or nonhemorrhagic presentations that have not produced any neurological deficits, and some of these lesions might be better suited to stereotactic radiosurgery. Candidates with good surgical prognosis are evaluated for embolization, if there is low associated risk and clear benefit to the surgical resection. AVMs patients that cannot be embolized due to significant risk or other clinical issues (e.g., hematoma and elevated ICP) are managed without this adjunct. AVMs deemed good radiosurgical lesions are

either small-volume niduses with a high probability of obliteration, in which case traditional single-stage radiosurgery is used, or are large niduses that require volume-staged radiosurgery. Patients with intracranial AVMs should be evaluated for associated aneurysms on feeding arteries, in the circle of Willis, and within the nidus. Aneurysms that are at least 7 mm in diameter are often treated. In addition, aneurysms that are associated with high-flow AVMs, are exposed to increased hemodynamic stress, or have a dysplastic morphology are also considered for treatment. Untreated aneurysms associated with AVMs need serial imaging monitored and if they enlarge, it should be treated immediately. After an AVM has been treated with multidisciplinary approach, angiography is used to determine whether the AVM has been obliterated. Nothing short of complete angiographic obliteration can be considered curative or protective against future hemorrhage. Patients with a residual AVM are re-entered into the algorithm to determine how best to manage the remaining nidus, and, often, AVMs that were incompletely obliterated with one modality are considered for another modality. Thus, incompletely obliterated AVMs are considered for microsurgery after previous radiosurgery, and vice versa. Sometimes, residual arteriovenous shunting after microsurgical resection represents a technical error that is addressed immediately with reoperation, particularly when venous outflow has been altered by the resection or the patient presented with rupture. Similarly, after radiosurgery, some residual AVMs remain poorly accessible surgically and are treated with an additional radiation dose. However, many of the three-modality strategies were developed when individual modalities incompletely obliterated the AVM and the management “crossed over” to other modalities.

Stereotactic radiosurgery refers to treatment method in which the entire AVM nidus is irradiated. Hypofractionated stereotactic irradiation and volume-staged radiosurgery are used for AVMs with diameter more than 3cm and volume greater than 10cm³ and generally requires more than one treatment session.

CONCLUSION

AVMs challenge the neurosurgeon, and one must synthesize different option in decision to treat. A coordinated strategy must be designed to optimize the different therapeutic modalities of embolization, microsurgery, and radiosurgery. Intraoperatively, the

technical challenges and dangers associated with AVMs are extreme. Postoperatively, there are difficult recovery and rehabilitation challenges to be met. A multidisciplinary team approach, involving neurosurgeons, neurointerventional radiologists, neurologists, neuroanesthesiologists, radiation oncologists, research scientists, and clinical nurses, is essential. Cooperative synergy at critical clinical crossroads optimizes patient outcomes, answers important clinical research questions, and leads to the development of innovative therapies for brain AVMs. Image 1 gave a summary what we are following in K.F.S.H and R.C Jeddah.

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