Original article:

Supratentorial cerebral arteriovenous malformations : a clinical analysis

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Abstract:

Background: Arteriovenous malformations (AVM) of the brain are congenital vascular lesions accounting for approximately 2% of all hemorrhagic strokes in young and otherwise healthy individuals. Ninety percent of all AVMs are supratentorial. The most common presentation of an AVM is intracerebral haemorrhage (ICH) and Seizures.

Aims and objectives: To study the patient profile, clinical presentation, radiological features of patients with supratentorial AVMs and also to study surgical outcome and complications in these patients.

Material methods: This was a prospective study conducted at IPGMER, Kolkata from August 2011 to December 2013. All patients with supratentorial AVMs attending Bangur Institute of Neurosciences OPD and admitting in B.I.N. Hospital wards were included, studied and analysed according the standard proforma.

Results: Out of 24 enrolled patients 80% were below 40 years of age presenting with intra cranial haemorrhage(59%), chronic headache(31.82%), seizures (27.27%) and focal deficit (22.73%). Most common type was Parenchymal hematoma(69%) and location in frontal, parietal and temporal region (22.5% each). 50% of AVMs were located at eloquent cortex of brain and 77.3% had superficial venous drainage. 45.5% AVMs had dominant supply from MCA. Out of these 7 patients required microsurgical intervention and rest were kept under observation. Total 4 patients died during the course and others were followed up every quarterly for one year.

Conclusion: The presentation of supratentorial AVMs in young population is complex and treatment is highly individualized. Imaging for AVMs should be considered in patients presenting with chronic headache and seizures, so that early recognition of AVM and intervention can be done.

Key words: Arteriovenous malformations, Supratentorial, Spetzler and Martin Grading

Introduction:

Arteriovenous malformations (AVM) of the brain are congenital vascular lesions that account for approximately 2% of all hemorrhagic strokes.^{1,2} Despite the relative rarity of the disease (with an estimated current detection rate of approximately 1/100 000 person-years)³, AVMs pose a significant neurological problem because patients are mostly young and otherwise healthy. Moreover, the availability of non invasive imaging has rapidly increased the detection of incidental AVMs. The complex cerebrovascular anatomy of AVMs makes them a challenge to treat, and the treatment itself carries significant risks. To evaluate the possible benefit of a risky treatment, one needs to understand the natural history and prognosis of the disease. The complexity of AVMs makes them a rather heterogeneous group of lesions in terms of various factors possibly affecting the risk of rupture and subsequent hemorrhagic stroke.

Aims and objectives:

To study the patient profile, clinical presentation, natural history and radiological features of supratentorial cerebral arteriovenous malformations.

Also to review the surgical outcome and complications of supratentorial cerebral arteriovenous malformations.

Material and methods:

This was a prospective study done at Bangur Institute of Neurosciences at Institute of Post Graduate Medical Education and Research (IPGME & R) Kolkata from August 2011 to December 2013. All the consecutive patients of arterio-venous malformations attending Bangur Institute of Neurosciences OPD and admitting in B.I.N. Hospital wards were studied and having radiological diagnosis of supratentorial cerebral arteriovenous malformation were included in the study. Patients of other cerebrovascular malformations (venous angioma, cavernous malformations, capillary telengectasia and direct & infratentorial fistula), spinal cerebral arteriovenous malformation, ucontrolled comorbid condition, history of significant head trauma were excluded from the study.

Patient's demographic profile (age, sex, occupation etc) along with Clinical presentation and natural history were noted, radiological investigations (NCCT / Contrast CT Scan Brain, CT Angiography, MRI Brain (Plain And Contrast), MR Angiography, 4 Vessel Cerebral Digital Subtractions Angiography (DSA)) were studied and patients were classified according to Spetzler and Martin Grading³. Pre and Post operative course was observed and patients were followed up every quarterly for the next one year. Data was collected on standered proforma and analysed statistically.

Results:

Table 01: Age distribution of patients in the study.

Age in years	ALL PATIENTS		SURGICAL PATIENTS		OBSERVED PATIENTS	
	Number of	percentage	Number of	percentage	Number of	percentage
	patients		patients		patients	
< 20	6	27	3	43	3	20
21-40	12	55	3	43	9	60
> 40	4	18	1	14	3	20
TOTAL	22		7		15	
MEAN AGE	28.14		26.14		29.07	
AGE RANGE	11-55		13-48		11-55	

The total 22 patients analyzed in the study were of the age ranging from 11 years to 55 years and mean age of patients was 28.14 years.

Spetzler and Martin	SURGICAL PATIENTS		OBSERVED PATIENTS	
Grade				
	Number of	percentage	Number of patients	Percentage
	patients			
Ι	3	43	5	33
Π	0	0	4	27
III	4	57	4	27
IV	0	0	1	7
V	0	0	1	7
TOTAL	7		15	

Table 02: Spetzler and Martin Grading in surgical and observed patients.

3 (43%) of the total operated patients were Spetzler and Martin Grade I AVMs, while 4 (57%) were Grade III AVMs.

Presenting symptom	ALL PATIENTS		SURGICAL PATIENTS		OBSERVED PATIENTS	
	N = 22		N =7		N =15	
	Number of	percentage	Number of	percentage	Number of	percentage
	patients		patients		patients	
Hemorrhage	13	59.09	5	71.43	8	53.33
Seizures	6	27.27	2	28.57	4	26.67
Chronic headache	7	31.82	2	28.57	5	33.33
Focal deficit	5	22.73	1	14.29	4	26.67
Other /	1	4.55	0	0.00	1	6.67
asymptomatic						
History of previous	3	13.64	2	28.57	1	6.67
rupture						

Table 03: Frequency of Symptoms at presentation.

Most common presenting symptom in the patients was hemorrhage, seen in 13 (59.09%) patients. Seizure at presentation was seen in 6 (27.27%) patients. Chronic headache, focal neuro deficit and history of previous rupture was seen in 7(31.82%), 5(22.73%) and 3(13.64%) patients respectively. 1 out of 22 patients (4.55%) was diagnosed incidentally in a CT scan performed for unrelated reasons.

Table 04. Distribution of dominant arterial supply.

Dominant Artavial cumply	Number of	% of	
Dominant Arteriai suppry	Patients	Patients	
ACA	2	9.1	
ACA AND MCA	2	9.1	
LENTICULOSTRIATE	0	0.0	
MCA	10	45.5	
MCA AND ACA	2	9.1	
MCA AND PCA	3	13.6	
MCA, ACA AND PCA	2	9.1	
PCA	1	4.5	
Total	22	100.0	

10 (45.5%) AVMs had dominant supply from MCA.

 Table 05: Outcome in observed patients

OUTCOME	Number of	% of	
OU I COME	Patients	Patients	
REBLEED IN FOLOWUP	1	6.67	
NO REBLEED	14	93.33	
MINOR NEURODEFICIT	1	6.67	
MAJOR NEURODEFICIT	0	0	
DEATH	1	6.67	
Total	15	100.0	

Out of 15 observed patients 14 had no rebleed during followup period, 1 patient had minor neurodeficit and 1patient died in followup due to rebleed.

Table 06: Outcome in surgical patients in different Spetzler and Martin Grades.

OUTCOME	Spetzler and Martin Grades				
	Ι	II	III	IV	V
RECOVERED WITH NO NEURO DEFICIT.	2 (66.6%)	0	2 (50%)	0	0
MINOR NEURODEFICIT	0	0	0	0	0
MAJOR NEURODEFICIT	0	0	0	0	0
DEATH	1 (33.3%)	0	2 (50%)	0	0
TOTAL	3	0	4	0	0

Out of 3 operated Spetzler and Martin Grade I patients 2 (66.6%) recovered with no neurodeficit, while one patient (33.3%) died. Out of 4 operated Spetzler and Martin Grade III patients 2 (55.0%) recovered with no neurodeficit, while 2 patients (50.0%) died.

Discussion:

A total 24 cases of supratentorial AVMs, diagnosed angiographically and fulfilling the inclusion criteria were included in our study. Out of these, 2 patients were referred to the higher centre and who were subsequently lost to followup were excluded from the final analysis of the data.Average age of diagnosis in patients presented in this study was 28.14 years with majority of patients in the age group of 20 to 40 years. Ondra et al.⁴ outlined the natural history of AVMs among 160 patients who presented with symptomatic AVMs and were followed for a mean follow-up of 23.7 years. The mean age at presentation was 33 years.

Most studies report an equal or slight male preponderance in patients with AVMs. In our study there was significant male preponderance may be reflected due to small sample size.In present study 13 patients (59.1%) presented with hemorrhage with most common location of hemorrhage being intra parenchymal seen in 9 (69%) of the patient presenting with hemorrhage, this finding was consistent with other studies. 45 to 70% of patients presented with hemorrhage in large AVM series by Crawford et al.⁵ Of all AVM-related haemorrhages, 63% were intraparenchymal, 32% are subarachnoid, and 6% are intraventricular in the studies by Ondra et al and Crawford et al.^{4,5}

Frequency of seizures at presentation in the present study was 27.3% seen in 6 patients with generalized tonic clonic seizures in 4 patients and focal seizures in 2 patients. As per the study done by Crawford et al seizures are the second most common presentation of AVMs. 17% to 40% of patients with AVMs present with epileptic seizures.⁵ Other common presenting features were chronic headache and focal deficits noted in 7 (31.8%) and 5 (22.7%) patients respectively. As Laakso Aki has described approximately 6–14% of patients with cerebral AVMs present initially with headaches, and focal neurologic deficit due to mass effect or hemodynamic changes is seen in 3 to 10% of patients.⁶

In our study 12 (54.5%) patients had right hemispheric AVMs and 10 (45.5%) had left hemispheric AVMs. 50% of AVMs were located in eloquent areas of brain. Hofmeister et al in their study of 1289 consecutive AVM patients from 3 independent databases described overall proportion of patients with an eloquent AVM location in 71%. Lobar distribution of AVMs in our study was 22.7%, 13.6%, 4.5%, 22.7%, 22.7% and 13.6% in frontal, frontoparietal, occipital, parietal, temporal and temporoparietal locations respectively. Apsimon et al in their population based study described lobar distribution as frontal 16.3%, temporal 21.3%, frontoparietal 11.3%, parietal 8% and occipital 10.8 % which was similar to our study.8

In our study 12 (54.5%) patients had AVM nidus size less then 3cm, 9 (40.9%) had nidus size between 3 to 6 cm and only 1 (4.5%) patient had nidus size more then 6 cm. 17 (77.3%) patients had only superficial venous drainage of AVMs while rest 5 (22.7%) had both superficial and deep venous drainage. In study by Hofmeister et al the overall proportion of AVMs classified as small was 38% (95% CI 35% to 41%), 55% of AVMs were medium in size, and 7% were classified as large , results similar to present study.⁷ Dominant arterial supply of AVMs we studied in present study was from MCA in 45.5% of patients.

In present study we utilized standard Spetzler Martin grading system³ with this criteria

the incidence of AVMs in various grades was for grade I, 8 (36.4%); grade II, 4 (18.2%); grade III, 8 (36.4%); grade IV, 1 (4.5%); and for grade V, 1 (4.5%). In a large Finnish series of 623 AVMs accrued over 55 years (1951 to 2005), 13% were grade I, 29% were grade II, 32% were grade III, 19% were grade IV and 4% were grade V AVMs.⁹In present study 7 (29.2%) patients underwent microsurgical treatment. Of the operated patients 3(43%) were grade I and 4(57%) were grade III. In present series there was no significant morbidity in recovered patients and mortality of 42.8% is noted in operated patients.

Conclusion:

Early detection by high index of suspicion and imaging is important for diagnosing and management of AVMs. As the treatment is highly individualised , Intraoperative monitoring for AVMs located in eloquent regions with better postoperative intensive care is needed to reduce the postoperative morbidity and mortality in surgical management of AVMs.

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