

A Clinical Study of One Hundred and Twenty Six Cases of Cerebral Arteriovenous Malformations¹

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= Abstract = One hundred twenty six cases of cerebral arteriovenous malformation (AVM), admitted to the neurosurgical department of Seoul National University Hospital between January 1973 and June 1987, were evaluated retrospectively with reference to clinical features, epidemiology and the outcome of surgical or conservative treatment. The peak incidence was at the third decade and the presenting symptoms were hemorrhage, seizure and headache in order of frequency. Hemorrhage was more frequent in the second and third decades while seizure was more frequent in the third and fourth decades. Small AVM's, less than 2 cm in diameter, were presented with hemorrhage more frequently than seizure, while large AVM's, more than 6 cm in diameter, were presented with seizure more frequently. The clinical outcome was better in adults than in children and patients presented with seizure showed better outcomes than patients with hemorrhage. The patients presented by hemorrhage had poor outcomes with conservative treatment and therefore surgical treatment was needed in these patients. Surgical treatment was also helpful for the patients presented by seizure.

Key words: *Arteriovenous malformation, Hemorrhage, Seizure, Headache, Surgical treatment*

INTRODUCTION

Arteriovenous malformations AVM's of the brain are congenital lesions that arise at an early fetal stage, forming the direct arterial to venous communications without an intervening capillary bed. As a child's brain develops it is presumed that the additional arterial contributions and small repeated hemorrhage happens to be a more common occurrence of symptoms in the second and third decades than in childhood (Stein *et al.* 1980).

With the development of sophisticated neuroradiologic techniques and improved treatment, these patients have become significant when considering the possibility of a cure or substantial

modification of the disastrous consequences of leaving these lesions to their natural course.

The natural history of cerebral AVM is not known concerning the rebleeding rate, mortality, mechanism of seizure, the surgical effect on the seizure, and the outcome according to the treatment modalities. We reviewed our experiences with AVM's and tried to compare the surgical management with conservative treatment on the retrospective basis.

The case records of 126 consecutive patients with cerebral AVM's who were admitted to the neurosurgical department of Seoul National University Hospital between January 1973 and June 1987, were reviewed.

MATERIALS AND METHODS

The case records of 126 consecutive patients with cerebral AVM's who were admitted to the neurosurgical department of Seoul National University Hospital between January 1973 and June

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1987, were reviewed. This review did not include patients with extracerebral arteriovenous fistula or caroticocavernous fistula. All cases were diagnosed on the basis of angiography and computed tomography of the brain.

Presenting symptoms were classified according to the first major symptoms and location was classified as hemispheric type, deep seated type, posterior fossa type and dural type (which is located mainly at the dura), on the basis of angiography. Deep seated type included the AVM's at the basal ganglia, thalamus, brain stem and ventricles.

The size of AVM was measured three dimensionally on the angiography in diameter, and large AVM which is over 6 cm in diameter. The treatment modality was classified as conservative and surgical treatment. The conservative group included the medical treatment or removal of hematoma only or the ventriculoperitoneal shunt for hydrocephalus.

The surgical treatment group included the total or partial removal of AVM or the obliteration of feeding vessels. Patient outcome was classified as good, fair or poor. The good category is the patient who has normal or minor neurologic deficits which do not prevent gainful employment, but the patient may have minor headaches, infrequent seizures, a trace of weakness or a visual field loss from an old hemorrhage. The fair category is the patient who is plagued by recurring hemorrhage, troublesome epilepsy, headaches or a progression of a neurological deficits such that gainful employment is limited, but not excluded, because of these symptoms. The patient is still capable of self-support with proper medical management. The poor category is the patient who is totally dependent on others for supportive care either at home or within an institution. Statistical analysis was done using chi-square test.

RESULTS

1) Age and sex distribution

The peak incidence of intracranial AVM's was in the third decade followed by the second decade and the fourth decade. The sex ratio of male to female was 2 to 1.

The peak incidence of males fell in the third, fourth and second decades while with females it fell in the third and second decades (Fig. 1).

2) Clinical manifestations

Presenting symptoms:

Presenting symptoms were hemorrhage, seizure and headache in order of frequency. Hemorrhage comprised 58.7% and seizure 27.8%. The incidence of rebleeding in these hemorrhagic patients was 21.6% (16 out of 74) and mortality due to rebleeding was 25% (4 out of 16) (Table 1).

Presenting symptom and age incidence:

Hemorrhage was more frequent in the second and third decades while seizure was more frequent in the third and fourth decades (Fig. 2).

Type of hemorrhage and seizure:

Among the locations of hemorrhage intracerebral hematoma was most frequent comprising 75.7%. Among the seizure type generalized seizure was most frequent (Table 2,3).

Location of AVM:

The location of intracranial AVM's was hemispheric, deep seated, posterior fossa and dural location in order of frequency (Table 4). Among the 126 AVM's aneurysms were combined in 7 cases

Table 1. Presenting symptoms

Symptom	No. of Cases	%
Hemorrhage	74	58.7
Seizure	35	27.8
Headache	6	4.8
Progressive hemiparesis	2	1.6
Acute hemiparesis	2	1.6
Proptosis	3	2.4
Paresthesia	1	0.8
Forehead mass	1	0.8
Blindness	1	0.8
Tinnitus	1	0.8
Total	126	

Table 2. Patients presenting with hemorrhage

Type of hemorrhage	No. of cases	%
Intracerebral hematoma	56	75.7
Intraventricular hemorrhage	10	13.5
Subarachnoid hemorrhage	8	10.8
Total	74	

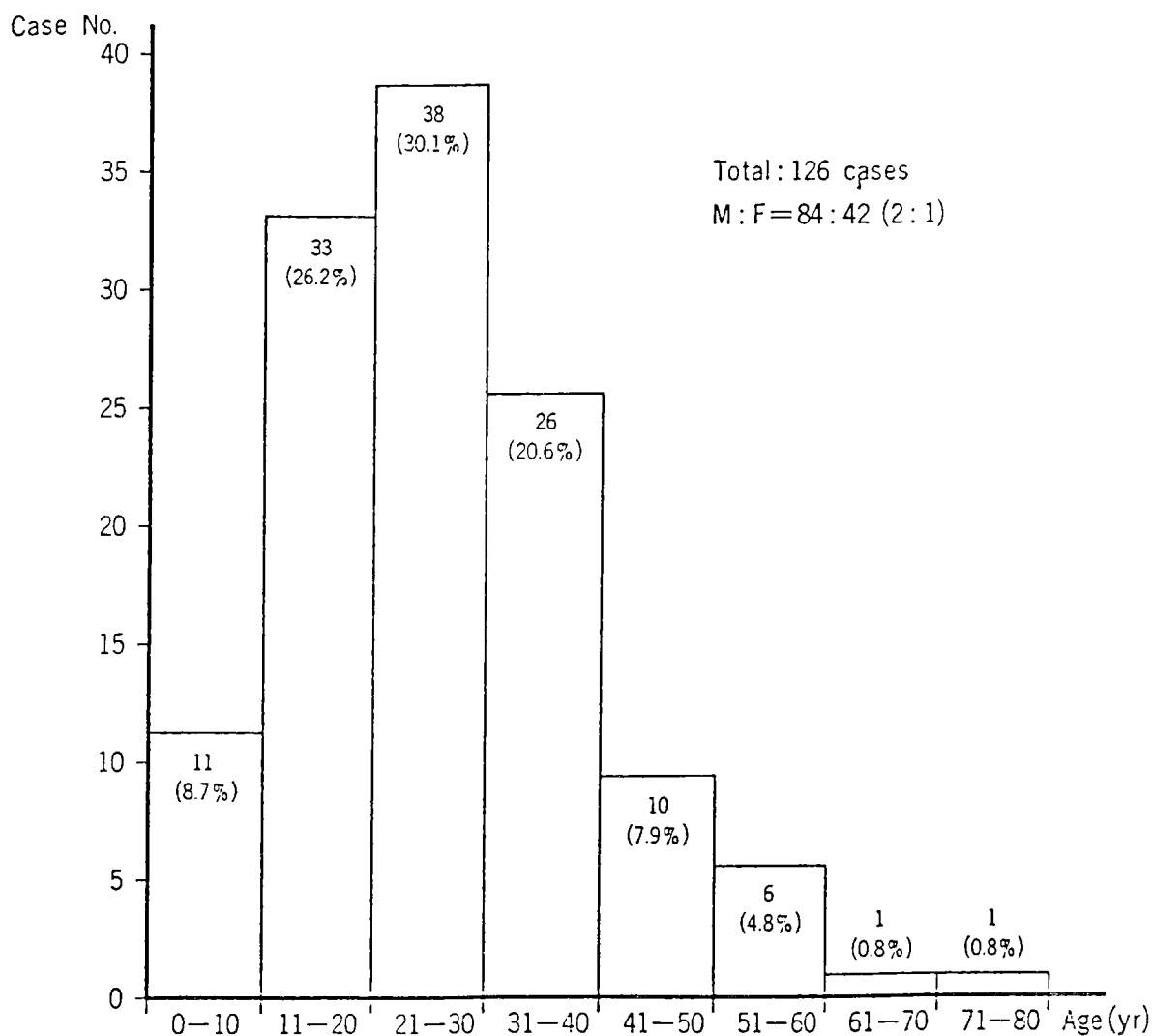


Fig. 1. Age distribution.

Table 3. Patients presenting with seizure

Type of seizure	No. of cases	%
Generalized seizure	24	68.6
Focal motor seizure	10	28.6
Psychomotor seizure	1	2.9
Total	35	

Table 4. Location of intracranial AVM

Location	No. of cases	%
Hemispheric	92	73.0
Deep	22	17.5
Post. fossa	9	7.1
Dural	3	2.4
Total	126	

(4.8%).

The size and presentation:

The size of AVM's is related to the presenting symptoms. Small AVM's, less than 2 cm in diameter, were presenting more frequently with hemorrhage than with seizure, while large AVM's, more than 6 cm in diameter, were presented with seizure more frequently (Table 5). There was statistical significance ($p < 0.005$).

3) Treatment and analysis of outcome

Surgical therapy was done in 50 cases and conservative treatment was done in 76 cases. Among the surgical treatment total removal was possible in 27.8% (Table 6). Among 126 cases 80 cases were able to be followed up. The average duration of follow up was 9.4 months (Table 7).

Relationship between age and outcome:

We analyzed the relationship between age and outcome. Among 27 children who could be fol-

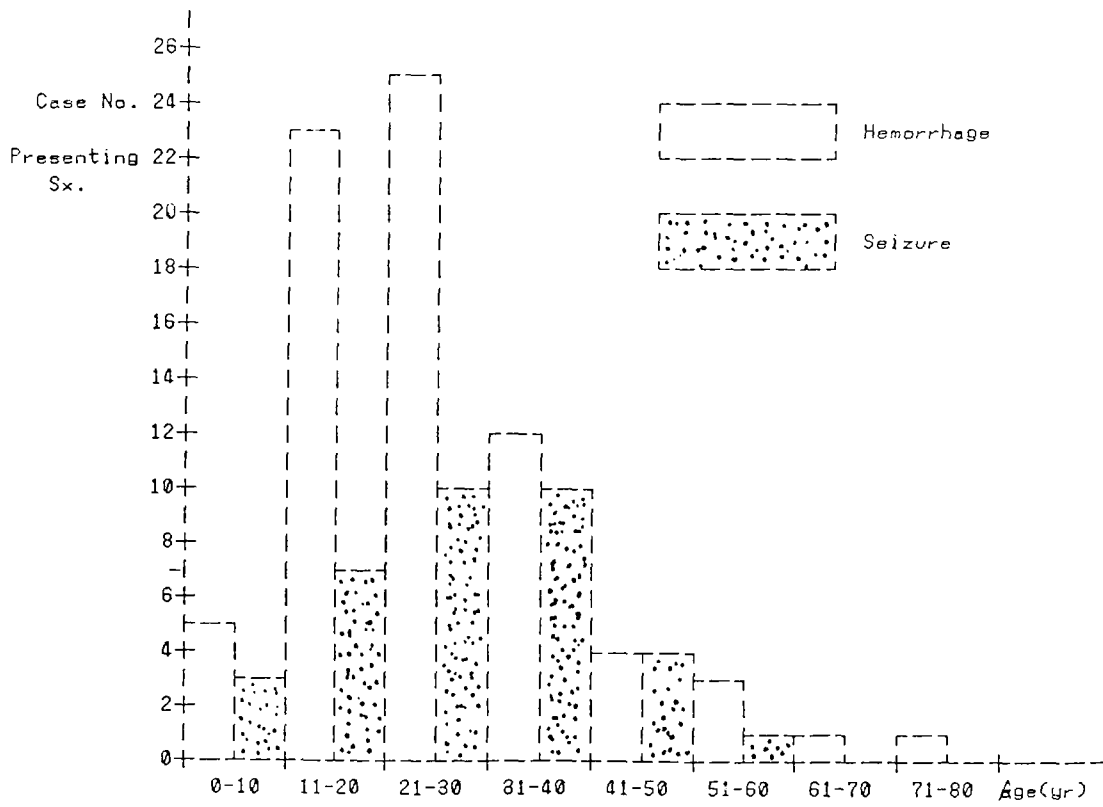


Fig. 2. Age distribution of patients who had hemorrhage or seizure as a presenting symptom.

Table 5. Relationship between size of AVM and symptom

Size(diameter) symptom	Small (<2cm)	Medium (2-6cm)	Large (>6cm)
Hemorrhage	30(85.7%)	17(41.5%)	5(25.0%)
Seizure	2(5.7%)	17(41.5%)	10(50.0%)
Total	35	41	20

Table 7. Follow-up (1 month-65 months)

Treatment group	F/U cases/ total	Average duration (month)
Conservative	43/ 76	8.8
Surgical	37/ 50	10.3
Total	80/126	9.4

Table 6. Modalities of treatment

	Modalities	No. of cases	%
Surgical therapy	—Total removal of AVM	35	27.8
	—Partial removal of AVM	5	4.0
	—Clipping of feeder	10	7.9
Conservative therapy	—Medical Tx only	67	53.2
	—Hematoma evacuation	6	4.8
	—V-P shunt	2	1.6
	—Clipping of associated aneurysm	1	0.8
Total		126	

Table 8. Relationship between age and outcome

Outcome	Children(0-16 yr)		Adult(above 16 yr)	
	No. of cases (%)		No. of cases (%)	
Good	14	(51.9)	49	(62.8)
Fair	9	(33.3)	15	(19.2)
Poor	1	(3.7)	7	(9.0)
Dead	3	(11.1)	7	(9.0)
Total	27		78	

Good: normal or minor neurologic deficits, gainful employment

Fair: self-support but limited gainful employment

Poor: totally dependent for supportive care

Dead

Table 9. Relationship between presenting symptom and outcome

Outcome	Hemorrhage		Seizure	
	No. of cases (%)		No. of cases (%)	
Good	34	(56.7)	24	(75.0)
Fair	13	(21.7)	7	(21.9)
Poor	5	(8.3)	0	(0.0)
Dead	8	(13.3)	1	(3.1)
Total	60		32	

Table 10. Overall result

Outcome	Conservative therapy		Surgical therapy	
	No. of cases (%)		No. of cases (%)	
Good	27	(48.2)	36	(73.5)
Fair	16	(28.6)	8	(16.3)
Poor	5	(8.9)	3	(6.1)
Dead	8	(14.3)	2	(4.1)
Total	56		49	

lowed up, 51.9% were good and among 78 adult patients, 62.8% were good. There was no statistical difference ($p > 0.05$) (Table 8).

Relationship between presenting symptom and outcome:

Sixty patients presented with hemorrhage and 32 patients with seizure were analyzed. The patients presented with seizure had better outcome (75%) than patients presented with hemorrhage (56%).

Table 11. Presenting symptom vs. outcome in surgical therapy

Outcome	Hemorrhage		Seizure	
	No. of cases (%)		No. of cases (%)	
Good	24	(70.6)	12	(85.7)
Fair	7	(20.6)	1	(7.1)
Poor	2	(5.9)	0	(0.0)
Dead	1	(2.9)	1	(7.1)
Total	34		14	

Table 12. Presenting Symptom vs. outcome in conservative therapy

Outcome	Hemorrhage		Seizure	
	No. of cases (%)		No. of cases (%)	
Good	10	(38.50)	12	(66.7)
Fair	6	(23.1)	6	(33.3)
Poor	3	(11.5)	0	(0.0)
Dead	7	(26.9)	0	(0.0)
Total	26		18	

This was statistically significant ($p < 0.1$). Mortality was also high in the hemorrhage group (Table 9).

Overall results of conservative therapy and surgical treatment:

Among 56 patients with the conservative treatment, 48.2% showed good outcome while 14.3% died. Of the 49 patients with the surgical treatment, 73.5% showed good outcome and 4.1% died. Statistical analysis was done between the two groups according to the good group and the others. There was statistical significance between the two groups ($p < 0.05$) (Table 10).

Surgical outcome according to the presenting symptom:

With surgery the patients presented with hemorrhage had good results in 70.6% of the cases while the seizure patients also had quite good results. There was no statistical significance ($p > 0.05$) (Table 11).

Outcome of conservative treatment according to the presenting symptom:

The patients with seizures had good outcomes in 66.7% of the cases while patients with hemorrhage had good outcome in 38.5% of the cases and 26.9% died. This was statistically significant ($p < 0.05$) (Table 12). So we concluded that surgical

treatment was needed in these patients presented with hemorrhage.

DISCUSSION

Arteriovenous malformation (AVM) of the brain are uncommon congenital lesion causing symptoms between the second and fifth decade and our cases developed most frequently in the second and third decade with a male preponderance as indicated in other reports. (Foster *et al.* 1972; Perret and Nishioka 1966; Parkinson and Bachers 1980).

The prevalence of AVM's has been estimated as about one-seventh of that of intracranial aneurysm. Therefore we note that approximately 0.14% of the populations harbors an AVM (Michelsen 1984). The natural history of cerebral AVM is not known about the rebleeding rate, mortality, the mechanism of seizure and the surgical effect on the seizure.

In our cases presenting symptoms were intracranial hemorrhage, seizure, and headache in order of frequency as reported by others. Perret and Nishioka (1966) reported that among the intracranial hemorrhage, intracerebral hemorrhage was the most frequent in 63% of cases, followed by subarachnoid hemorrhage and intraventricular hemorrhage, in order of frequency. While in our cases intracerebral hemorrhage was the most frequent, intraventricular hemorrhage was the next one in frequency. Among the seizure types, the generalized type was the most frequent differing from the Perret's report. For the occurrence of seizure there are several mechanisms such as occult hemorrhage or steal phenomenon (Nornes and Grip 1980).

Aneurysm is reported to be associated with AVM in 2.7 to 8.7% of cases (Hayashi *et al.* 1981; Luesenhop 1984) and our cases fell into this range. There are many reports about the relationship between the size of the AVM and the presenting symptom (Foster *et al.* 1972; Graf *et al.* 1983; Guidetti and Delitala 1980; Waltimo 1973). As in other reports our cases of small AVM tended to present with hemorrhage while large AVM was presented with seizure more frequently. In regard to the rebleeding of AVM, the incidence of rebleeding rate was reported to be 23% to 67% and the death rate due to rebleeding was about 40% while the interval of rebleeding was quite variable (Perret and Nishioka 1966; Svien and McRae 1965; Fults and Kelly 1984). The rebleeding rate was 21.6% and the death rate was 25% in our results.

The relationship between the patient's age and outcome was of no statistical difference between the adult and child group as in the report of Fults and Kelly (1984) but there needs to be more accumulated data to clarify this point. The relationship between the initial symptom and outcome revealed that patients with hemorrhages had poorer outcomes than that of patients with seizures. There were 9 cases of mortality among which 8 of the cases were attributed to hemorrhage. For the surgical treatment of these lesions there is much controversy according to the location of the lesion, the size, the patient's age and clinical status, and the technical aspect, but with the development of recent radiological and surgical facilities the surgical indication of this disease is broadened (Luesenhop and Rosa 1984).

Dandy, Cushing and Bailey were pioneers in describing the surgical problems created by AVM's of the brain and Olivecrona in 1932 was the first to successfully excise an AVM. As the surgery has become more sophisticated, there has been a reduction in morbidity and mortality of AVM cases. Successful removal has also been recorded with AVM's of the speech and motor area, as well as subcortical areas, without additional neurologic deficits.

Pool and Potts (1965) listed the advantages of excising AVM's, including the relief of seizures not alleviated by medication, the relief of progressive neurological behaviour of intellectual deterioration, and the prevention and relief of serious hemorrhage, especially if it is recurrent.

We also decided the surgical indications on the basis of many aspects but among the patients who were recommended to have surgical removal about 30% of patients refused the surgery for paramedical reasons. In our cases total removal of AVM was possible in 35 cases, partial removal in 5 cases, and clipping of feeder in 10 cases, without mortality. On comparing the outcome between the conservative treatment and surgical group, the surgical outcome was superior to the conservative treatment with statistical significance, although it is important to consider which patients were selected for surgery. Moreover the patients presented with hemorrhage had poor outcome with conservative treatment therefore surgical treatment was more helpful for these patients.

For the better understanding of this disease there should be a prospective clinical study in line

with the experimental study for its pathophysiological mechanism.

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= 국문초록 =

126예의 뇌동정맥 기형에 대한 임상적 연구

서울대학교 의과대학 신경외과학교실

한대회 · 이선호 · 최길수

1973년 1월부터 1987년 6월까지 서울대학교병원 신경외과학교실에서 뇌혈관촬영 및 전산화단층촬영으로 진단된 126예의 뇌동정맥 기형을 대상으로 임상적 특성 및 보존적 치료 및 수술적 치료에 대한 효과를 분석하였다.

1. 호발연령은 10대와 20대 였으며 남녀 비율을 2:1 이었다.
2. 발현증상은 출혈, 전간, 두통의 순이었고 출혈은 10대와 20대에서 호발하였으며 전간은 20대와 30대에서 호발하였다. 출혈의 부위는 뇌실질내출혈이 가장 많았고 전간의 형태는 전신성 전간이 가장 많았다.
3. 동정맥기형의 발생위치는 대뇌반구형이 가장 많았고 기저핵, 시상, 뇌간 및 뇌실을 포함한 심부형, 후두외형, 경막형의 순서였다.
4. 직경 2 cm 이하의 작은 동정맥 기형은 직경이 6 cm이상의 큰 동정맥 기형에 대해 전간보다는 출혈에 의해 발현되는 경우가 많았다.
5. 예후는 소아에서보다 성인에서 양호하였으며 예후는 전간에 의해 발현된 경우가 출혈에 의한 경우보다 양호하였다.
6. 출혈에 의해 발현된 예들은 보존적 치료로는 예후가 불량하였으며, 이런 경우에는 수술적 치료가 필요하다고 사료되었으며 전간으로 발현된 환자에서도 수술적 치료의 결과는 양호하였다.