

Clinical Features of Angiographically Occult Vascular Malformation

Sun Ho Lee*, Byung Kyu Cho and Dae Hee Han

*Department of Neurosurgery, Seoul National University
College of Medicine, Seoul 110-744, Korea*

Abstract—A retrospective review of 22 cases of angiographically occult vascular malformations (AOVMs), treated at our institution during the past 6 years, was conducted to define better the clinical and radiological manifestations as well as the effective treatment modality of these lesions. There were 12 (54.5%) arteriovenous malformations (AVMs), 8 (36.4%) cavernous angiomas, 1 (4.5%) venous angioma, and 1 (4.5%) unclassified vascular malformation.

The most frequent initial presentations included hemorrhages in 72.7% of the patients, seizures in 22.7%, and compression by mass lesion in 4.5%. Of the 16 hemorrhagic cases, 11 (58.7%) were AVMs and 3 (18.7%) were cavernous angiomas. The remaining 2 cases were a venous angioma and an unclassified vascular malformation. In almost all of the AVMs (11/12) the initial presentations were hemorrhages. Nine cases of AOVMs (40.9%) developed recurrent hemorrhages as confirmed by clinical, radiological, and operative features.

All of the MRI findings performed in the 17 cases showed thick circumscribed hypodense rims on T2-weighted images representing hemosiderin deposits. Various central intensities suggested the presence of hematomas in different stages.

Twenty patients underwent surgery with total excision and subsequent good results. Two patients, one with a pontine lesion and the other with a middle cerebellar peduncle lesion, underwent partial excision associated with consequent persistent neurological deficits.

This report suggests that the AOVMs are prone to cause hemorrhages or neurological deficit and surgical excision can provide an effective and safe treatment modality for this disease entity.

Key Words: *Angiographically occult vascular malformation (AOVM), Arteriovenous malformation, Venous angioma, Cavernous angioma, Hemorrhage, Seizure, Rebleeding*

INTRODUCTION

The term angiographically occult vascular malformation (AOVM) may be applied to a group of

intracranial vascular hamartomas (arteriovenous, cavernous, venous angioma, capillary telangiectasia or mixed) which are not visualized by cerebral angiography (Henderson and Gomez, 1969; Kramer and Wing, 1977; Bell *et al.*, 1978; Leblanc *et al.*, 1979; Lobato *et al.*, 1988).

AOVMs have occasionally been found in patients operated on for spontaneous brain hematoma (Crawford and Russel, 1956; Krayenbuhl and Sie-

* Author for correspondence

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benmann, 1965; Wakai *et al.*, 1985), uncontrolled seizure or intracranial hypertension syndrome (Patterson and McKissock, 1956; McCormick and Nofzinger, 1966; Tindall *et al.*, 1978; Wharen *et al.*, 1982; Steiger and Tew, 1984). Since the extensive use of computed tomography (CT) and the advent of magnetic resonance image (MRI) for the study of patients with cerebrovascular disease and epilepsy, detection of AOVMs has increased markedly (Gomori *et al.*, 1986). However the natural history of this disease entity is still obscure in terms of the rate of recurrent hemorrhage, surgical indication and its prognosis, the best form of treatment, and whether or not the different pathological types classically described have a consistently distinct profile (Giombini and Morello, 1978; Becker *et al.*, 1979; Michelsen, 1979; Mori *et al.*, 1980).

In order to ascertain these facts we analyzed 22 cases of AOVMs surgically treated in the Department of Neurosurgery, Seoul National University Hospital, between January 1984 and August 1990.

MATERIALS AND METHODS

The case records of 22 patients with angiographically occult vascular malformations (AOVMs), which were surgically confirmed in the Department of Neurosurgery of Seoul National University Hospital between 1984 and 1990, were reviewed. Full angiographic studies were performed in all cases and revealed no vascular abnormality except mass effect. CT scans were done in all 22 cases, and magnetic resonance images were acquired in 17 cases either with a 2.0T or 0.5T MRI scan.

Initial presentations were classified as hemorrhage, seizure or mass lesion according to the major cause of the presenting symptoms. The cases of seizure due to intracerebral hemorrhage were classified as hemorrhage. Pathological classification was done as either a definite case or a probable case. Probable cases were those with incomplete biopsy but which showed entangled vessels. These vessels were rather compatible with AVM in the operative field.

There were 13 definite cases and 9 probable cases. Among the 22 cases there were 12 AVMs,

which included probable cases, 8 cavernous angiomas, 1 venous angioma, and 1 unclassified vascular malformation. Surgery was classified into complete excision and partial excision. The surgical outcome was classified according to the change of the neurologic state after surgery as improved, stationary and aggravated.

RESULTS

1. Age and sex distribution

The age incidence varied from 3 years to 57 years with the mean age being 27.2 years and relatively prevalent in the young age group. The sex ratio of male to female was 9 to 13 (Fig. 1).

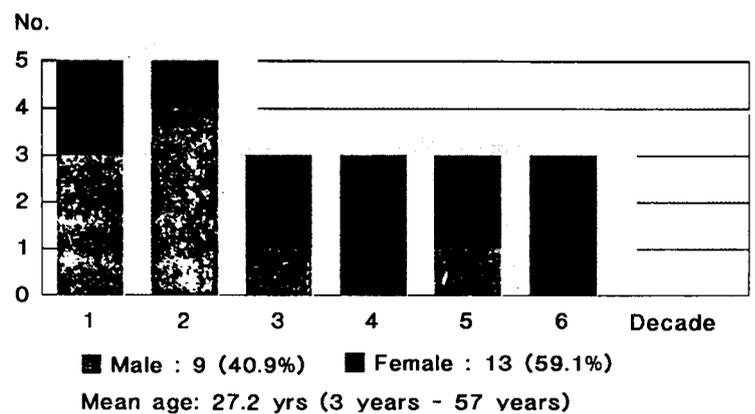


Fig. 1. Age and Sex Distribution

2. Location of AOVMs

Fourteen cases (63.6%) were located in the supratentorial area, and 8 cases (36.4) were in the infratentorial area. All 8 cavernous angiomas were located supratentorially. One of the cases of cavernous angioma was located as a multiple lesion, not only in the supratentorial but also in the infratentorial location, but it was classified as supratentorial according to the main lesion.

Among the 12 AVMs, 5 were located in the supratentorial area, and 7 were located in the infratentorial area showing a relatively high incidence in the infratentorial location. One venous angioma was located in the infratentorial location (Table 1).

Locations according to vascular territory showed that the MCA territory was the most prevalent location comprising 9 cases. Six cases were lo-

Table 1. Location of AOVMs (1)

	Supratentorial	Infratentorial	Total
AVM	5	7	12
CA	8		8
VA		1	1
UVM	1		1
Total	14 (63.6%)	8 (36.4%)	22

AOVM (angiographically occult vascular malformation)

AVM (arteriovenous malformation)

CA (cavernous angioma)

VA (venous angioma)

UVM (undclassified vascular malformation)

Table 2. Location of AOVMs (2)

Supratentorial (14) 63.6%	ACA Territory	3
	MCA Territory	9*
	PCA Territory	2
Infratentorial (8) 36.4%	Cerebellum	6
	Brain Stem	2

* includes 1 multiple case

ACA (anterior cerebral artery)

MCA (middle cerebral artery)

PCA (posterior cerebral artery)

cated in the cerebellum, and 2 were at the brain stem (Table 2).

3. Clinical manifestation

Initial presentation was caused by hemorrhage in 16 cases (70%), seizure in 5 cases, and mass lesion in 1 case. Among the 8 cavernous angiomas, 3 cases presented with hemorrhage, 4 with seizure, and 1 with mass lesion. However, among the other 14 vascular malformations, 13 (92.9%) presented with seizure. It is noteworthy that 11 out of 12 AVMs presented with hemorrhage (91.7%) (Table 3).

Recurrent hemorrhage, confirmed by clinical, radiological, and operative features, showed in 9 out of 22 cases (40.9%). However, subclinical microscopic bleeding, revealed by MRI or pathologic findings, was more than 90% of all AOVMs. Among the 16 cases which presented with hemorrhage, 7 (43.8%) showed recurrent hemorrhage, 5 presented with seizure, and 2 cases showed recur-

Table 3. Initial Presentation

	AVM	CA	VA	UVM	TOTAL
Hemorrhage	11	3	1	1	16
Seizure	1	4			5
Mass		1			1
Total	12	8	1	1	22

Table 4. Recurrent Hemorrhage*

	AVM	CA	VA	UVM	TOTAL
Hemorrhage	4	1	1	1	7
Seizure	1	1			2
Total	5	2	1	1	9

* Confirmed by clinical, radiological and operative features

rent hemorrhage. Five out of 12 AVMs and 2 out of 8 cavernous angiomas showed recurrent hemorrhages (Table 4).

4. Radiological findings

Computed tomographic findings showed hyperdensity or mixed density in 22 cases of precontrast CT scan. Among 17 post contrast scans, 5 showed mottled or rim enhancement, and 12 showed no enhancement. Calcification showed in 3 cases (Fig. 2).

Magnetic resonance images (MRI) produced a very peculiar finding showing a core of mixed signal intensity with a peripheral low signal intensity rim, which suggested hemosiderin deposits on the T2 WI in all 17 cases (Fig. 3).

5. Surgery and surgical outcome

Among the 22 cases, total excision was done in 20 cases and partial excision was performed in 2 cases which were located in the brain stem. There was no operative mortality, and the 9 cases which had no neurological deficit preoperatively showed no additional deficit. Among the 13 cases who had preoperative neurological deficit, such as cranial nerve palsy, motor weakness, or cerebellar dysfunction, improvement showed in 10 cases, stationary state in 2 cases and aggravation in 1 case (Table 5).

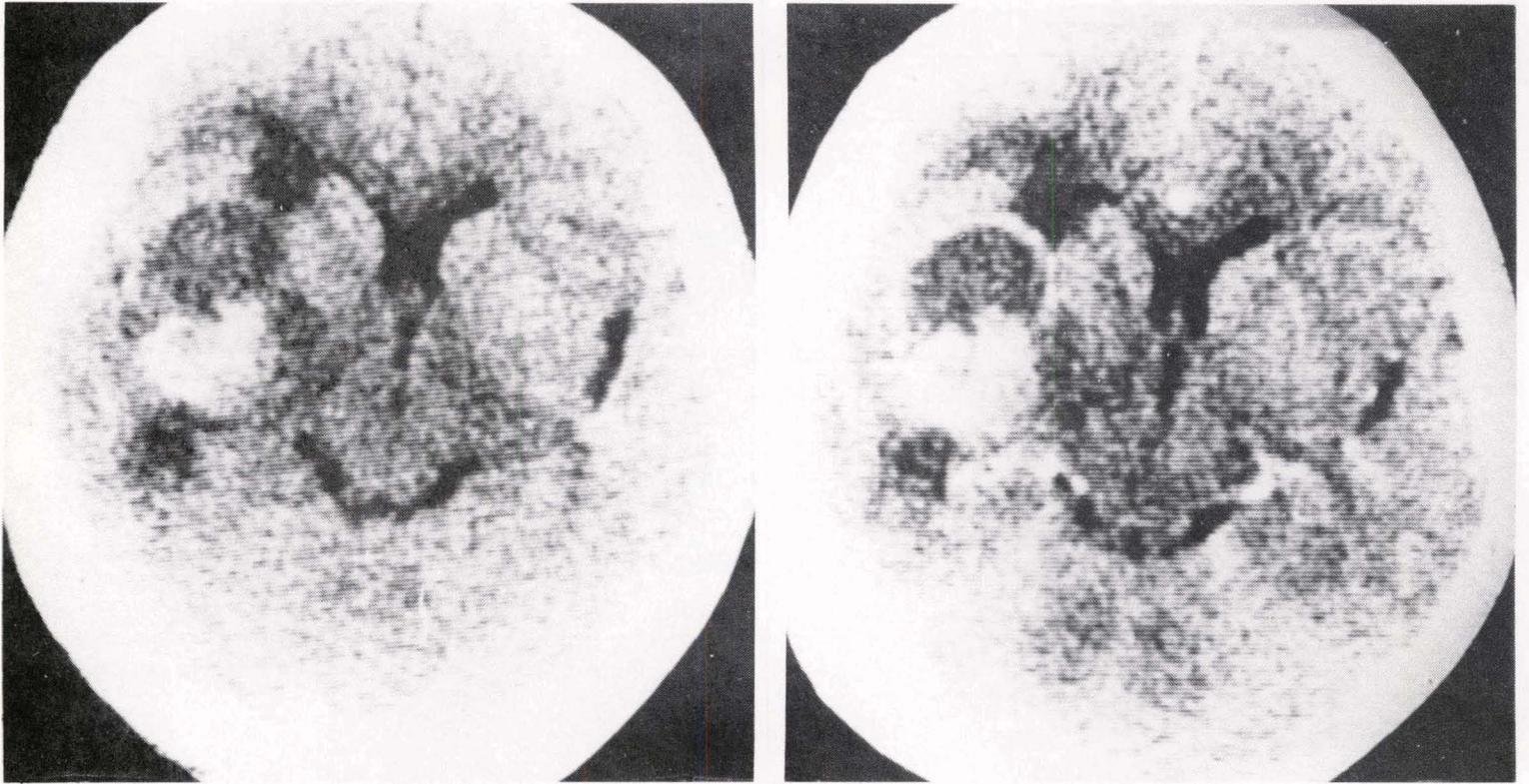


Fig. 2. Computed tomography (CT) of AOVM shows mixed density mass lesion on the right temporal lobe with inhomogeneous enhancement after contrast enhancement.

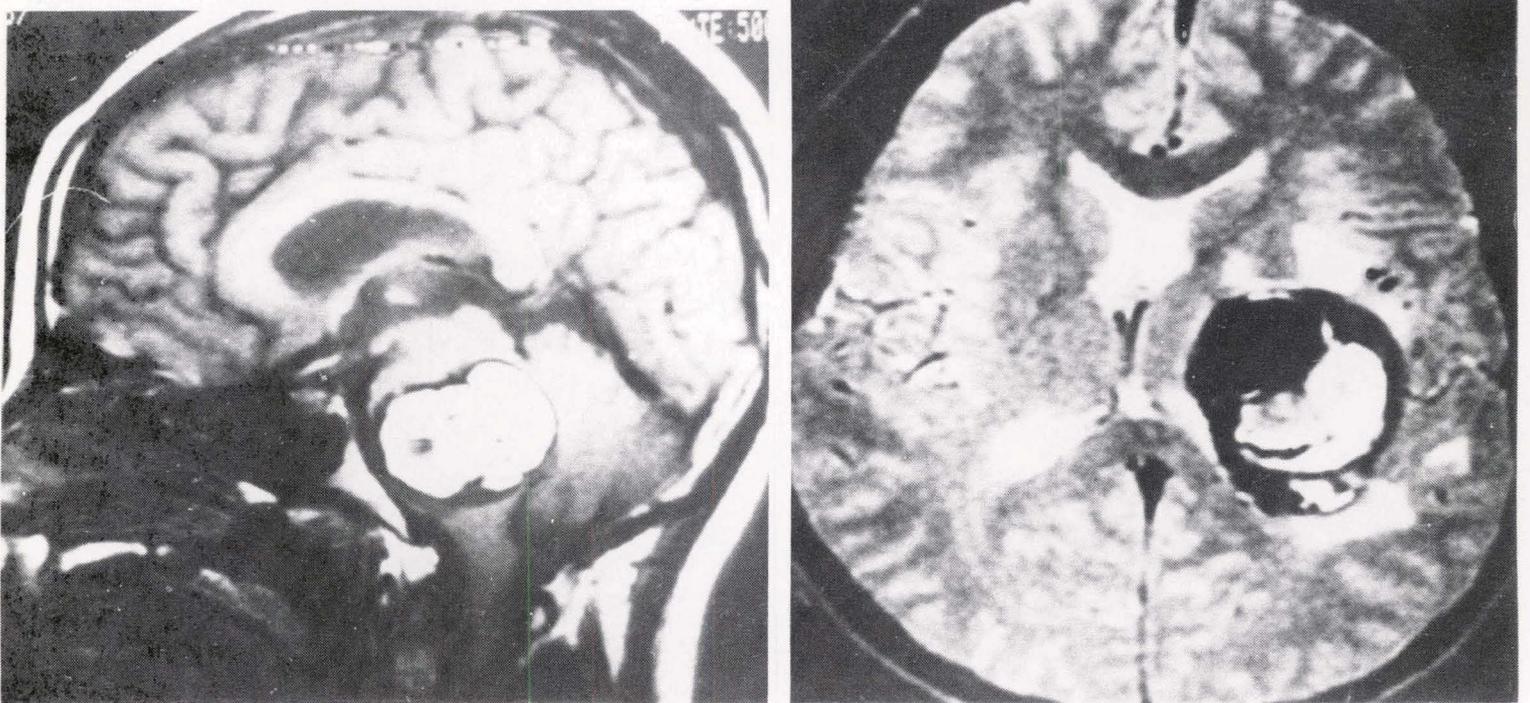


Fig. 3. Magnetic resonance image (MRI) of AOVM, T1WI, and T2WI shows a mixed signal intensity suggesting various stages of the hematoma with a surrounding low intensity rim.

The surgical outcome was generally quite good using microscopic surgery. However, the neurologic deficit of lesions at the functional area persisted

in 5 out of 7 cases with somewhat of an improvement (Table 6).

Table 5. Surgical Outcome of AOVMs

Preop	Postop
No ND* (9)	No ND (9)
ND (13)	Improved (10) Stationary (2) Aggravated (1)

* ND: neurological deficit (cranial nerve palsy, motor weakness, cerebellar dysfunction)

Table 6. Outcome of Functional Area

	Preop ND*	Postop ND*
Eloquent area (3)	3	2
Brain stem (2)	2	1
Basal ganglia (2)	2	2
Total	7	5

* ND: neurological deficit

DISCUSSION

Margolis *et al.* (1951) first described the small vascular malformation as the cause of massive intracranial spontaneous hemorrhage, but it was very difficult to diagnose before the advent of the CT scan. In 1956 Crawford and Russell subsequently used the term “cryptic vascular malformation” for the small vascular lesions clinically without angiography. Initially the term “cryptic” was applied to the group of vascular malformations based on their small size alone.

The more appropriate term, “angiographically occult vascular malformation” (AOVM), represents a myriad of vascular malformations with variable histology and includes cryptic AVMs, thrombosed AVMs, cavernous angiomas, and other histological vascular lesions escaping detection by cerebral angiography. This term is generally accepted now (Leblanc *et al.*, 1979; Lobato *et al.*, 1988; Tung *et al.*, 1990). The prevalent use of CT and the advent of MRI have greatly contributed to the detection of this disease entity, and MRI findings are quite characteristic in the diagnosis of this disease (Gomori *et al.*, 1986).

Lobato *et al.* (1988) reported 21 cases of AOVM

with a review of 241 previous cases. Pathologically, AVMs comprised 43.8%, cavernous angiomas 31.2%, venous angiomas 9.9%, capillary telangiectasis 3.8%, and mixed or unclassified angiomas 11%. In our series, 54.5% of AOVMs were AVMs, and 36.3% were cavernous angiomas.

Patients of all age groups were affected, but a peak symptomatic incidence occurred in the second and third decades of life. The average age of presentation was 31.9 years, and there was no prevalence for either sex.

Regarding the location and histopathology of the lesions, Lobato *et al.*, (1988) report that over 75% of AOVM were located in the supratentorial region, with the highest frequency being in the middle cerebral artery territory. In our series, 63.6% were located in the supratentorial region.

Clinical manifestation can be classified as hemorrhage, seizure or mass lesion. Wakai *et al.* (1985) report 58.7% of 172 cases of AOVM were accompanied by hemorrhage. In our series, 70% of the cases presented with hemorrhage.

Although the full spectrum of their natural history remains to be elucidated, angiographically occult vascular malformations are prone to cause recurrent hemorrhages (Wakai *et al.*, 1985; Lobato *et al.*, 1988).

Recurrent hemorrhage has been mentioned as the pathophysiological mechanism for neurological deficit, and with each successive recurrent hemorrhage the likelihood of a resultant persistent neurologic deficit increased.

It is difficult to determine the exact rate of rebleeding from these lesions. However, the rate of rebleeding appears considerable and may be similar to that of high-flow angiographically apparent AVMs. Lobato *et al.* (1988) report 24% of the patients they reviewed had a clinical course suggestive of recurrent hemorrhage, and Tung *et al.* (1990) report 33%, while our series showed 40.9%. However, it was reported that 80% to 90% of angiographically occult vascular malformations showed evidence of hemorrhage if subclinical hemorrhages were included. Even in asymptomatic cases of those presenting with seizures, MR imaging and pathological examination have documented the presence of hemosiderin, presumably the resi-

due of previously experienced subclinical hemorrhage (Wakai *et al.*, 1985). These findings are quite similar to our study.

The lack of angiographic identification in these vascular malformations may be due to the existence of one or more of the following features: (1) a lesion size below the resolution capacity of angiography, (2) compression of the vessel's lumen by a hematoma, (3) destruction of the abnormal vessels by macroscopic bleeding, (4) spontaneous thrombosis, (5) thrombosis secondary to gross hemorrhage, (6) partial thrombosis with sluggish circulation through the remaining patent vessels, (7) posthemorrhagic vascular spasm, (8) location of the lesion in the boundaries of the territories irrigated by different cerebral arteries, (9) dilution of the contrast medium in the enlarged cavernous vascular spaces of the malformation, and (10) defective angiographic techniques (Cohen *et al.*, 1982; Bruunelle *et al.*, 1983; Hashim *et al.*, 1985; El-Gohary *et al.*, 1987).

The CT scan may visualize the AOVM itself and/or the secondary changes induced in the surrounding tissue such as blood deposits, edema, atrophy, and calcification. The AOVMs appeared hyperdense on the nonenhanced CT scans in 95% of the cases and hypodense in 3%; they were not visualized in 2% of the cases.

Following intravenous injection of the contrast medium, the lesion was enhanced on CT scan in 74% of the cases; the enhancement was usually discrete. AOVMs of mottled appearance showing calcification may be indistinguishable from a low-grade glioma, meningioma or granuloma; magnetic resonance imaging may help to make a differential diagnosis by detecting the presence of hemosiderin (Kramer and Wing, 1977; Kucharczyk *et al.*, 1985; Lobato *et al.*, 1988).

MR images of these patients can often help suggest the diagnosis of AOVM with increased confidence and are most prominent on T2WI. These areas are interspersed with foci of mixed signal intensity patterns which correspond to the different stages of evolution within the hematoma. The low intensity or signal void areas most likely represent hemosiderin deposits (Kucharczyk, 1985; New *et al.*, 1986; Gomori *et al.*, 1986; Chang *et al.*, 1989).

There is a controversy about active surgery, especially in the eloquent area, because total removal in certain circumstances would be associated with unacceptable complications, especially in the case of pontine lesions, but surgical exploration is generally accepted with microsurgery. The surgical outcome is reported as generally good, with a 3% mortality, and the risk of repeated hemorrhage justifies the removal of these lesions whenever possible (Scott *et al.*, 1973; Chyatte, 1989; Ogilvy *et al.*, 1988).

Concerning radiation therapy, the use of radiosurgery has met with limited success in treating cavernous angiomas as opposed to high-flow AVMs (Steiner *et al.*, 1987), and recurrent hemorrhages may occur before complete obliteration of the lesion by radiosurgery or any other technique is achieved.

Because of the fear of hemorrhage, stereotactic biopsy is not generally recommended when this lesion is suspected.

In summary, this series of cases underscores the diagnostic effectiveness of MRI and the propensity of AOVMs to cause recurrent hemorrhages. Each successive hemorrhage tended to be associated with an increased incidence of permanent neurological deficit.

Surgery can be effective and relatively safe in removing these lesions even in hazardous areas of the brain.

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뇌혈관 조영술 상 잠재성인 뇌혈관 기형의 임상적 특성

서울대학교 의과대학 신경외과학교실

이선호 · 조병규 · 한대희

뇌혈관 조영술 상 잠재성인 뇌혈관 기형의 병태생리와 임상특성, 이에 따르는 적절한 치료법을 규명하기 위하여 과거 6년간 서울대학교병원 신경외과학교실에 입원하여 수술적 치료를 받은 22명의 뇌혈관 조영술 상 잠재성인 뇌혈관 기형을 가진 환자를 분석하였다. 이들 중 뇌동정맥 기형이 12예로 54.5%를 차지하였고 해면혈관종 36.4%, 정맥기형과 미분류 혈관기형이 각각 1예였다. 이들의 발현증상은 출혈이 가장 많아 16예로 72.7%를 차지하였고 간질 22.7% 종괴 효과에 의한 증상이 4.5%를 차지하였다.

16명의 출혈환자 중 58.7%가 동정맥기형이었고 18.7%가 해면혈관종이었다. 동정맥기형은 대부분 출혈로 발현되었다. 잠재성 뇌혈관 기형의 40.9%는 재출혈 소견을 보였다. 핵자기 공명영상은 이 질환에 특징적인 소견을 보이며 특히 T₂ 강조 영상에서는 다양한 시기의 혈종음영과 그 주위를 싸는 헤모시데린 침착을 의미하는 저음영이 특징적으로 나타났다.

20명 환자는 완전 절제를 하였으며 좋은 결과를 보였다. 뇌간이나 기능적으로 중요한 위치에 병소가 있던 2명 환자는 부분 적출만이 가능하였고 신경학적 손상은 잔존하였다. 이들 결과로서 뇌혈관 조영술 상 잠재성인 뇌혈관 기형은 출혈을 잘하고 출혈이 반복됨에 따라 신경학적 손상이 심해지므로 수술로 추가적인 손상을 심화시킬 위치가 아니면 적극적인 수술적 치료를 하는 것이 환자의 임상경과에 도움이 되리라 사료되었다.