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A Case of Venous Angioma with Arteriovenous Shunts —Case Report—

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ABSTRACT

A 35-year-old man presented with a sudden headache and disturbance of consciousness. On admission, his consciousness level was Japan Coma Scale 100. Computed tomography disclosed a subarachnoid hemorrhage (SAH) and right cerebellar hematoma. Angiography was performed and, at first, arteriovenous malformation of the posterior fossa was diagnosed. Then external decompression of the posterior fossa and ventricular drainage were performed, followed by barbiturate therapy. Repeat angiography revealed that the lesion was a venous angioma with arteriovenous shunts. On day 37, subtotal removal of the lesion was performed. Intraoperatively, acute brain swelling emerged and partial internal decompression of the right cerebellar hemisphere was performed. The postoperative course was comparatively good and the patient was discharged with very mild ataxia. The patient is now being followed up in our outpatient clinic.

Key words: *Venous angioma, AV shunt, Posterior fossa*

Vascular malformations in the brain are classified into five groups: telangiectasia, varix, cavernous angioma, arteriovenous malformation (AVM) and venous angioma[9]. Venous angioma is characterized by an absence of feeding arteries and demonstration of dilated medullary veins and collecting central veins. In addition, angioma is observed exclusively in the venous phase in the angiogram. Venous angioma with arteriovenous shunts (VA with AVS) are described by Huang[5] as medullary venous malformations with an arterial component, and this type of venous angioma has a distinctive capillary blush in the arterial phase in the angiogram. Thirty-four such cases have been reported in the literature, including our case. This entity is not well known and its management is discussed here.

CASE REPORT

A 35-year-old man presented with a sudden headache and disturbance of consciousness, and was hospitalized by ambulance. On admission, his consciousness was 100 (Japan Coma Scale) without apparent hemiparesis. Computed Tomography showed a subarachnoid hemorrhage and right cerebellar hematoma with acute hydrocephalus (Fig. 1). Angiography was immediately performed and vascular malformation was seen at the right posterior fossa. Initially, we thought this lesion was an arteriovenous malformation coexisting with a venous angioma (Fig. 2). In order to control the intracranial pressure, ventricular drainage via the right posterior horn and external decompression of the posterior fossa were performed, followed by barbiturate therapy for five days. His consciousness level improved gradually and on the fourteenth day after admission, his consciousness became clear and hydrocephalus did not occur even after the removal of ventricular drainage. MRI on the twenty third day showed a cerebellar hematoma and flow void, indicating venous angioma (Fig. 3). For the purpose of surgical resec-

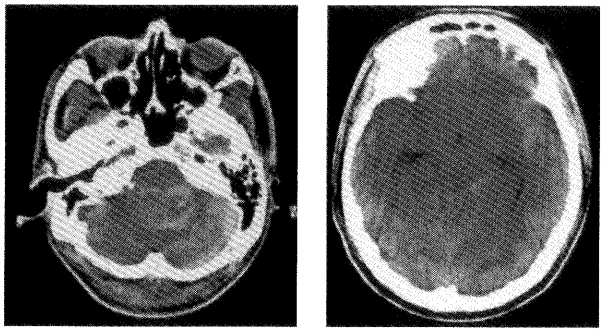

Fig. 1. CT on admission showed SAH and left cerebellar hemorrhage. Acute hydrocephalus was also recognized.

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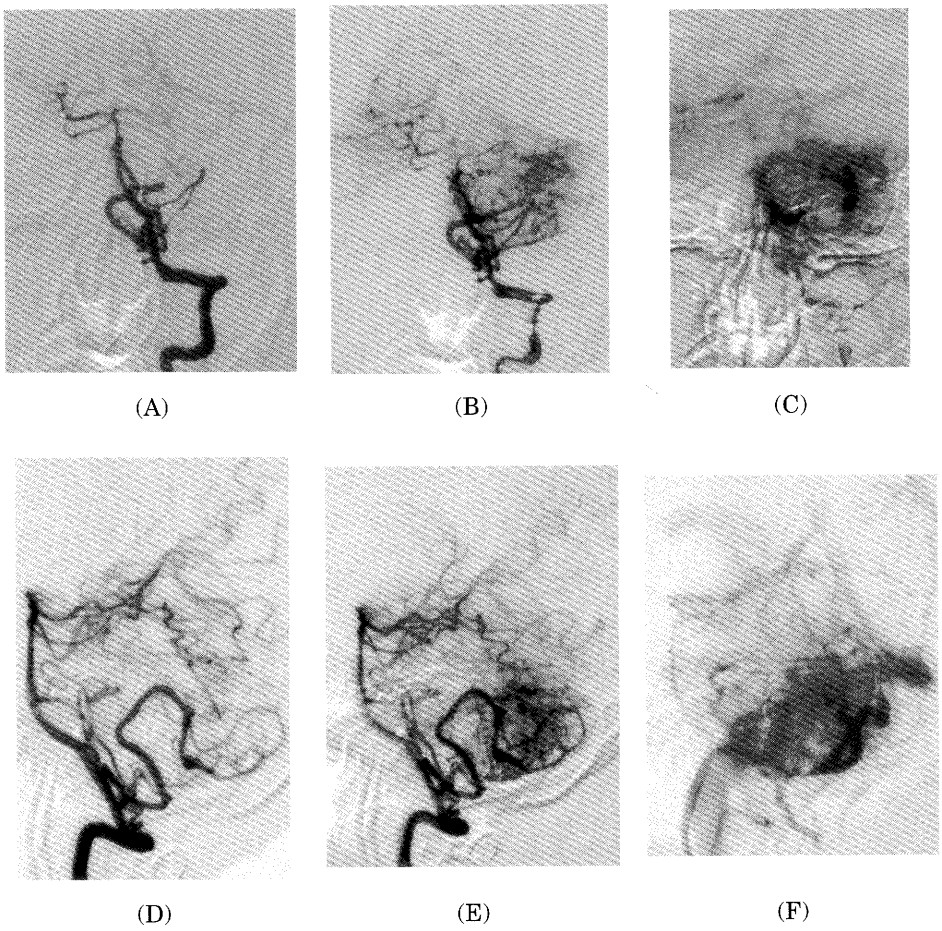

Fig. 2. An angiogram disclosed a capillary blush in the left cerebellar hemisphere in both the arterial phase (A, D) and capillary phase (B, E), and these drained into the venous angioma.
This lesion looked like AVM, however, no nidus was seen. Venous angioma was seen only in the venous phase (C, F).

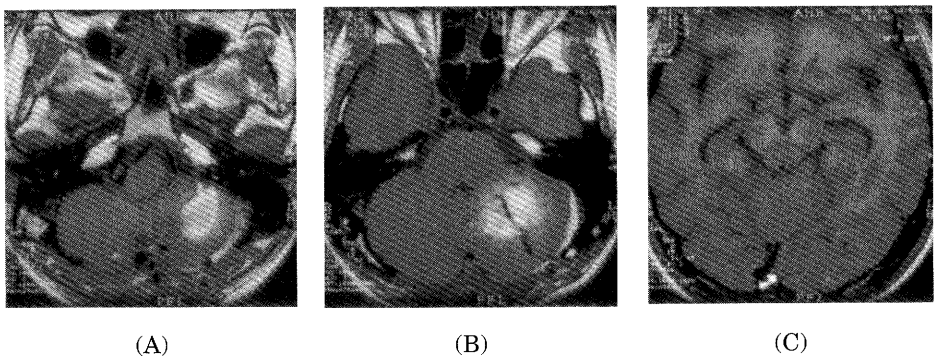

Fig. 3. MRI on the twenty third day showed a left cerebellar hematoma and flow void that indicated venous angioma. Tightness in the posterior fossa was improved compared to the CT on admission.

tion of the lesion, angiography was performed again (Fig. 4). In the arterial phase, a dilated posteroinferior cerebellar artery (PICA) with distal aneurysm and a capillary blush in the right cerebellar hemisphere were observed. A dilated medullary vein and early venous filling were also seen. In the initial angiogram, venous angioma was seen only in the venous phase. However, with the decrease of intracranial pressure in the posterior fossa, arteriovenous shunts and early venous filling clarified and a venous angioma was observed in the late arterial phase. The angioma drained into two areas. The main portion went backward and into the torcular while the other portion went forward and passed around the brain stem into the superior petrosal sinus. From these

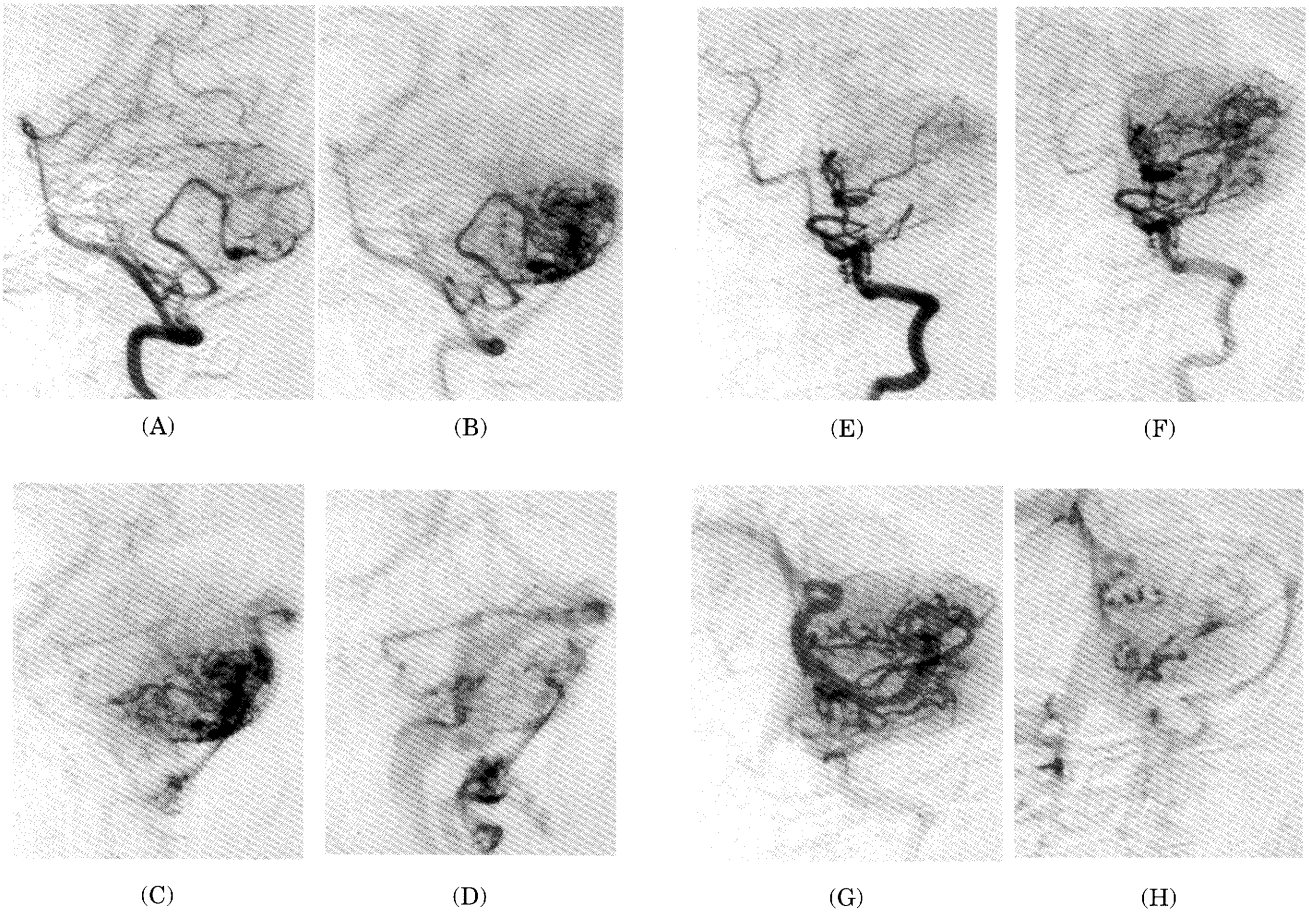

Fig. 4. In the arterial phase (A, E), dilated PICA with distal PICA aneurysm (this aneurysm was misunderstood as an arterial loop on admission) and capillary blush was seen. This time, in the capillary phase (B, F), venous angioma partially appeared reflecting early venous filling.
In the venous phases (C, D and G, H), venous angioma is clearly seen and it drained into the torcular and superior petrosal sinus.

findings, it was diagnosed as venous angioma with arteriovenous shunts. On day 37 emobolization of the distal portion of PICA and PICA aneurysm were performed using liquid coils, and on the following day partial removal of the angioma including the capillary blush was performed. After opening the dura matter of the posterior fossa, the presence of a red vein that served as the main drainer to the torcular was confirmed. Intraoperatively, the distal portion of PICA, the PICA aneurysm and the capillary blush were dissected. Angioma was coagulated and cut in the deep portion and the drainer which drained the brain stem area was preserved. When the dissection of the lesion, except for the main drainer, was completed, the red vein turned to a normal darkish color. However, moderate acute brain swelling with petechial hemorrhage around the peripheral of the right cerebellar hemisphere emerged just after this draining vein was cut away. Internal decompression of the right cerebellar hemisphere was performed while preserving the cerebellar nucleus, and the postoperative course was comparatively good. The histological diagnosis was AVM. Postoperative angiography (Fig. 5) and MRI (Fig. 6) showed subtotal removal of the lesion, and the patient was discharged with mild ataxia.

DISCUSSION

Venous angioma shows the following characteristics. 1. Angiographically normal circulation time 2. No abnormalities in the arterial and capillary phases 3. In the venous phase, many radiating medullary veins converging into a relatively large and centrally located vein, and hence to a dural sinus or, at times, to deep cerebral veins. Although little is known, there is a type of venous angioma that has arteriovenous shunts and or capillary blush in the late arterial phase. This is called a venous angioma with arteriovenous shunts (VA with AVS). This entity occasionally shows dilated feeding arteries[7,18,19] and, owing to the arteriovenous shunts, venous angioma is seen from the arterial or capillary phase[17]. There are many other designations of VA with AVS, as venous angioma with early filling vessels[11,17], medullary venous malformation with an arterial component[5], high-flow medullary venous malfor-

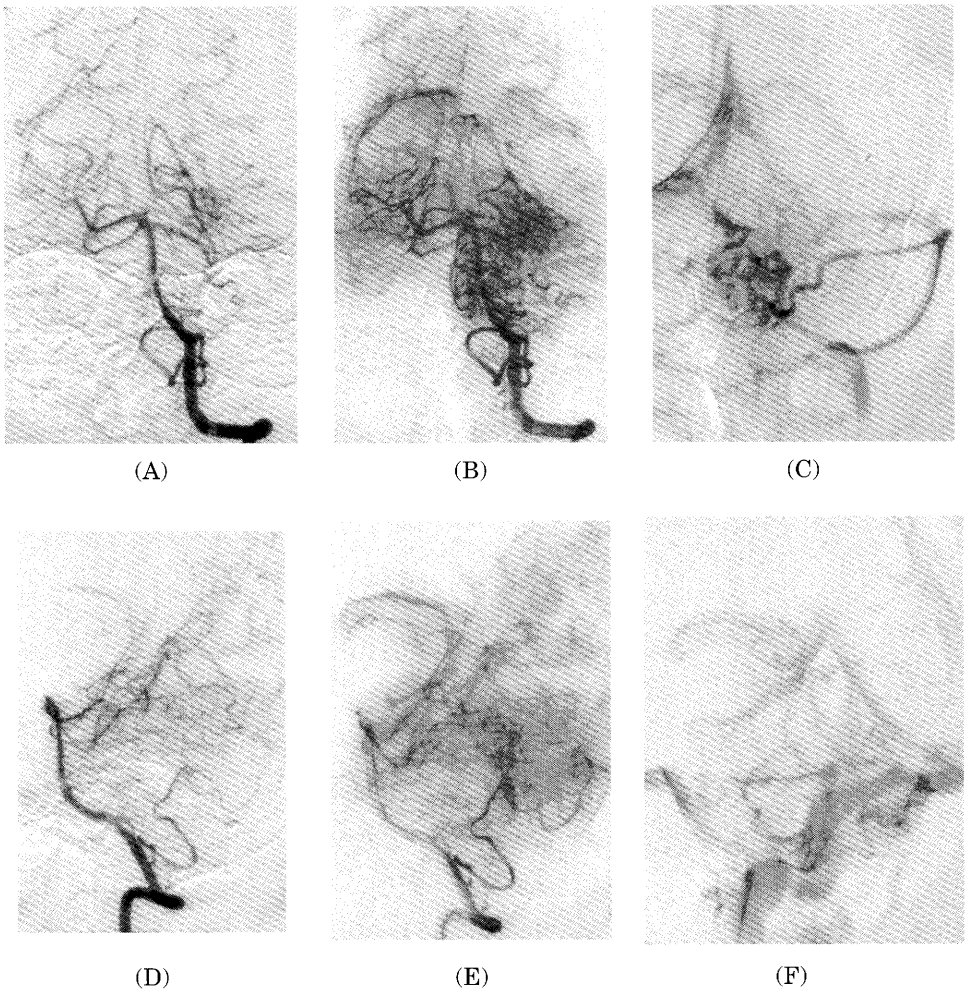

Fig. 5. A post-operative angiogram showed disappearance of PICA aneurysm and subtotal removal of the VA with AVS (A, B, D, E), leaving the draining vein to the superior petrosal sinus (C, F).

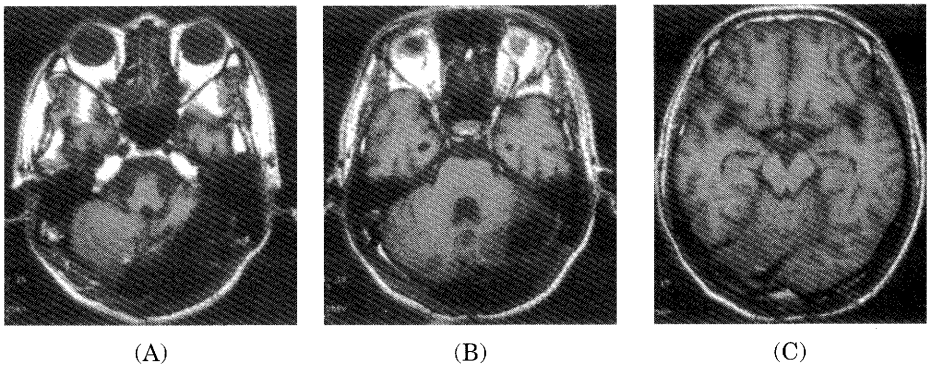

Fig. 6. Post-operative MRI showed partial excision of the left cerebellar hemisphere.

mation[12], arterialized venous malformation[1], and a transitional form between a venous angioma and an arteriovenous malformation[12]. Histology of VA with AVS is reported to be either venous angioma or AVM[11]. Fierstien and associates[3] noted that lack of arteries on histological examination of VA with AVS may be related to the location of the feeding arteries on the periphery of the malformation, outside the area of the pathologist's interest. Moritake et al[11] insisted that histological findings are not necessarily definitive in the diagnosis of vascular malformations and that angiographical findings are important. In the previous literature, there have been 34 patients, including our case, that were diagnosed as VA with AVS (Table 1). There were 20 men and 14 women, ranging in age

Table 1. Clinical information in 34 patients with VA with AVS

No. angioma	Authors for angioma	Age/Sex	Symptoms	Location	Hemorrhage from	Operation	Outcome
1	Wolf et al. (1967)	52//M	ICH	Lt frontal Lt temporal Rt parietal Rt frontal	No	No	Death
2	Wendling et al. (1976)	28/M	Seizure	Rt frontal	No	Lobectomy	—
3	Wendling et al. (1976)	49/M	Seizure	Lt frontal	No	No	Good recovery
4	Preissig et al. (1976)	23/F	Headache	Rt frontal	No	Block resection	Good recovery
5	Michael et al. (1977)	54/F	Epilepsy	Lt frontal	No	No	Good recovery
6	Michael et al. (1978)	29/F	Epilepsy	Rt Cerebellum	No	Extirpation	—
7	Sarwar et al. (1978)	38/M	Hepatic coma	Lt Basal	No	No	Death from hepatic coma
8	Suganuma et al. (1978)	28/M	Seizure	Lt frontal	No	Partial removal	Good recovery
9	Cabanes et al. (1979)	58/F	Headache	Lt frontal	No	Lobectomy	Good recovery
10	Cabanes et al. (1979)	20/F	Cranial nerve palsy	Rt Lt cerebellum	No	No	No change
11	Fierstien et al. (1979)	22/F	SAH	Lt frontal	No	No	—
12	Moritake et al. (1980)	39/M	Incidental	Rt frontal	No	No	Deterioration
13	Pardatscher et al. (1980)	46/M	IVH	Lt occipital	No	No	Good recovery
14	Saito et al. (1981)	27/F	Headache	Lt Basal	No	No	—
15	Saito et al. (1981)	25/M	Incidental	Lt frontal	No	No	—
16	Saito et al. (1981)	41/F	SAH	Rt frontal	No	No	—
17	Huang et al. (1984)	39/M	Tremor	Rt temporal	No	—	—
18	Huang et al. (1984)	52/M	Incidental	Lt parietal	No	—	—
19	Hirata et al. (1986)	18/M	ICH	Rt parietal	Yes	Operation for ICH	Hemiparesis
20	Shiroyama et al. (1986)	34/F	ICH	Lt frontal	Yes	Operation for ICH	Good recovery
21	Yasargil et al. (1988)	27/M	ICH	Rt frontal	Yes	—	—
22	Sonoda et al. (1988)	24/F	IVH	Rt parietal	No	No	Good recovery
23	Kurimoto et al. (1989)	26/M	ICH	Rt frontal Rt parietal	Yes	Operation for ICH	Good recovery
24	Tashiro et al. (1989)	49/M	SAH	Rt frontal	No	No	Good recovery
25	Ochi et al. (1990)	48/F	Headache	Lt frontal	No	No	Good recovery
26	Tomura et al. (1990)	47/M	ICH	Lt frontal	No	No	Good recovery
27	Awad et al. (1993)	39/F	Seizure	Lt frontal	No	Gyrectomy	Good recovery
28	Awad et al. (1993)	36/F	ICH	Parietal	Yes	Ope ration for ICH	Goog recovery
29	Awad et al. (1993)	54/M	ICH	Lt temporal	Yes	Gyrectomy	Good recovery
30	Sagoh et al. (1996)	24/M	Hemiparesis	Rt cerebral Rt cerebellum	No	No	Good recovery
31	Komiyama et al. (1999)	26/M	Incidental	Rt Basal	No	No	Good recovery
32	Komiyama et al. (1999)	24/F	Infarction	Lt parietal	No	No	Good recovery
33	Komiyama et al. (1999)	22/M	IVH	Lt parietal	Yes	No	Good recovery
34	Present case (2003)	35/M	SAH	Lt cerebellum	No	Partial removal	Good recovery

Abbreviations : ICH: intracerebral hemorrhage, Lt: left, Rt: right, SAH: subarachnoid hemorrhage, IVH: intraventricular hemorrhage

from 18 to 58 years (mean 35.4 years). The location of the lesion was frontal lobe in 18 patients, parietal lobe in 8 patients, cerebellum in 4, temporal lobe in 3, basal ganglia in 3, occipital lobe in one. As symptoms, intracranial bleeding was seen in 15 patients, and among these cases, there were 7 where bleeding was thought to be from the VA with AVS (20.6%). Other symptoms were epilepsy in 6, headache in 4, incidental in 4 and others in five. There were twelve operated cases, including evacuation of the hematoma in 4, lobectomy or gyrectomy in 4, partial removal of the angioma in 2, and block resection of the angioma in one. Fierstien et al reported that intracerebral hemorrhage or subarachnoid hemorrhage in patients with VA with AVS might be caused by rupture of the capillary aneurysm[3]. Moritake et al[11] proposed to classify venous angiomas into two subgroups, with or without capillary blush. In this report, 3 cases caused hemorrhage out of 12 venous angioma cases and one case caused hemorrhage out of 12 VA with AVS cases. There were no significant differences in the incidence of hemorrhage between these two groups. However, at that time, there were few cases of VA with AVS reported. Indeed, in the literature, there are now at least 7 cases that caused intracranial hemorrhage out of 34 cases (20.6%). McLaughlin et al[10] reported that the bleeding rate of venous angioma without AVS was 0.61% per year. On the other hand, the natural history of VA with AVS is unknown. From this report, the incidence of bleeding seems to be higher in VA with AVS than in VA without AVS. However, this result may be under a bias. In order to clarify the accurate incidence of bleeding, an accumulation of cases is necessary. For the treatment of venous angioma, it is now widely accepted that angioma does not require surgical resection or, since the venous angioma is frequently associated with a cavernous angioma, it is now recommended to resect an associated, accessible cavernous angioma while preserving the venous angioma in case of hemorrhage[1,10,15]. On the contrary, surgical resection has been reported for venous angioma in the frontal lobe[2,8,14,18], temporal lobe[7,8] and the posterior fossa[6,8,13,16]. Hirata et al[4] emphasized the need for surgical removal of the VA with AVS. In our case, where the cause of SAH and cerebellar hematoma was suspected to be the rupture of a PICA aneurysm, treatment of aneurysm and evacuation of hematoma might be another option for therapy. Indeed, acute brain swelling occurred just after the cutting of main drainer. When VA with AVS is found incidentally, course observation is recommended. However, for cases with intracranial bleeding, it is important to find the precise cause of the hemorrhage. If the hemorrhage is caused by a coexisting AVM or aneurysm, treatment for these lesions is necessary. When the cause of bleeding is thought to be VA with AVS per se, if possible, total resection of the lesion avoiding venous infarction should be aimed at because the existence of red vein in VA with AVS seems that it resembles the AVM.

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