Progressive Diffuse Arteriovenous Malformation

—Case Report—

Yasuhiro KOJIMA and Nobumasa KUWANA

Department of Neurosurgery, Yokohama Minami Kyosai Hospital, Yokohama

Abstract

A rare diffuse arteriovenous malformation (AVM) occurred in an 11-year-old boy with a 4-year history of transient ischemic attacks and weakness of the left extremities. Postcontrast computed tomography (CT) showed cord-like enhancement in the corpus callosum and pineal region. Magnetic resonance (MR) imaging demonstrated atrophy of the right hemisphere and multiple signal voids in the sulci. Cerebral angiography showed a diffuse AVM fed by the bilateral anterior cerebral and right middle cerebral arteries. The AVM nidus consisted of scattered nidi. Several draining veins appeared as multiple signal voids on MR images. No surgery was possible because of the scattered nidi. Left hemiparesis and mental retardation worsened, with progressive atrophy of the right hemisphere on serial MR images. Follow-up radiological examinations showed that the number of nidi increased, the draining vein enlarged, and the gyri calcified. The gyriform calcification on CT scans strongly resembled that seen in Sturge-Weber syndrome.

Key words: arteriovenous malformation, magnetic resonance imaging, angiography

Introduction

Diffuse cerebral arteriovenous malformation (AVM) is rare, and the computed tomographic (CT), magnetic resonance (MR) imaging, and angiographic characteristics have not been clarified. We report a patient with diffuse AVM, describe the angiographic and MR imaging characteristics, and discuss the definition of diffuse AVM.

Case Report

A 7-year-old boy presented with transient motor weakness of the left extremities in August, 1986. Examination at our hospital found transient hemiparesis and no intellectual impairment. Diffuse AVM was diagnosed based on cerebral angiographic findings. In October, 1988, he developed permanent motor weakness on the left, and intellectual impairment became conspicuous. In April, 1990, the left hemiparesis had progressed, causing stiffness of the left upper extremity. His familial history was non-contributory.

At third admission in April, 1991, he was alert,

but with moderate intellectual impairment. Motor functions were 3/5 in the left upper extremity and 4/5 in the left lower extremity by the manual muscle test, and the upper extremity was stiff. He was barely able to walk unaided. No cranial nerve dis-

Fig. 1 Postcontrast CT scan in August, 1986, showing irregular enhancement in the corpus callosum and pineal region.

turbance was observed.

CT scans at first admission in August, 1986 demonstrated abnormal blood vessels coinciding with the cerebral sulci. In addition, the vein of Galen was abnormally enlarged in the pineal region (Fig. 1). MR images in April, 1990 revealed multiple signal voids

Fig. 2 Axial (*left*) and sagittal (*right*) T₁-weighted MR images in April, 1990, showing multiple signal voids in the right frontoparietal region indicating AVM.

Fig. 3 A: Right carotid angiogram, arterial phase, showing scattered nidi (*arrow*) along the middle cerebral artery. B: Right carotid angiogram, venous phase, showing several draining veins into the superior and inferior sagittal sinuses. C: Left carotid angiogram, arterial phase, showing scattered nidi (*arrow*) along the bilateral anterior cerebral arteries. D: Left carotid angiogram, venous phase, showing enlarged inferior sagittal sinus and vein of Galen.

Fig. 4 Serial CT scans, showing a small calcification in the frontal lobe appearing first in 1988 and progressing to a gyriform calcification as in Sturge-Weber syndrome. A: August, 1986, B: September, 1988, C: April, 1990, D: August, 1992.

coinciding with the cerebral sulci in the right frontotemporal and parietal regions. A tortuous signal void was also observed from the corpus callosum to the pineal region (Fig. 2). Carotid angiograms showed dilation of the right middle cerebral and bilateral anterior cerebral arteries, and numerous small scattered nidi along the arteries. The draining vein was not visible in the arterial phase, first appearing in the capillary phase. The vein drained into the superior sagittal sinus, inferior sagittal sinus, and vein of Galen (Fig. 3). Vertebral angiograms revealed development of the posterior choroidal artery and similar scattered nidi. The posterior fossa appeared normal.

He was conservatively treated, but left hemiparesis and mental retardation worsened. Serial CT scans during 6-year follow-up showed atrophy of the frontal lobe and gyriform calcification similar to Sturge-Weber syndrome (Fig. 4). MR images showed progressive enlargement of the cerebral sulci and signal void areas (Fig. 5). Angiograms revealed an increasing number of scattered nidi.

Fig. 5 Serial MR images, showing progressive atrophy of the frontoparietal cortex and increased signal void areas. A: September, 1986, B: April, 1990, C: April, 1992.

Discussion

The angiographic characteristics of AVM include the size of the nidus and superficial or deep draining veins. In our case, the AVM had scattered nidi, so the size could not be measured. The draining vein drained into both superficial and deep regions. Spetzler and Martin⁶ described diffuse AVM with similar angiographic findings, but no details were reported. Some authors proposed the term diffuse AVM^{1,6,7,9} for lesions with such angiographic findings, while others suggested telangiectatic vascular mass,⁵ diffuse angioma,⁴ or angiodysplasia based on the involvement of impaired blood vessel formation.⁸

The angiographic findings of diffuse AVM can be defined as: 1) diffuse nidus without a clearly identifiable margin; 2) the scattered nidus located in one lobe to differentiate from angioma; and 3) the superficial or deep draining veins. We think that the scattered nidi demonstrated by angiography distinguish diffuse AVM from multiple or large AVM. Our case showed that the number of nidi increases, the draining vein enlarges, and the gyri calcify. The gyriform calcification on CT scans strongly resembles that seen in Sturge-Weber syndrome.

The origin of diffuse AVM may be a fetal vascular circuit disorder at 3–6 weeks age.² Impairment of capillary formation causes abnormal enlargement of the capillaries at numerous locations due to an arteriovenous shunt, resulting in formation of scattered nidi similar to diffuse AVM.

Ten cases of diffuse AVM have previously been reported.^{1,3–9} The reported ages ranged from 19 to 60 years (mean, 38.1 yrs), with no sex predominance.

Our patient was 7 years old, and is the youngest reported. Initial symptoms reported were subarachnoid hemorrhage in two patients,^{1,9} transient ischemic attack in three,^{3,7,8} and epilepsy in two.⁹ Angiographic findings were described for nine patients: diffuse nidus in five,^{1,6,9} and scattered nidi,⁴ racemose AVM,³ irregular pooling,⁷ and moyamoya vessels³ in one each. No CT or MR imaging information has previously been reported.

All patients have so far been treated conservatively. Yasargil *et al.*⁹ has suggested radiation therapy for diffuse AVM, but this has never been used. Deaths have occurred due to status epilepticus and dysphagia.^{5,7} Lichtor and Mullan³ found no changes in the angiographic findings 3 years later, but follow-up of other patients was not reported. We considered vascular embolization and radiation therapy for our patient, but the diffuse nature of the nidus makes such procedures difficult. At present we are monitoring our patient, but as the symptoms are progressive, we hope to find some effective treatment.

References

- 1) Appel L, Dochez CH: Diffuse arterio-venöse Mißbildung der beiden Gehirnhemisphären und der Schädelhaut. *Der Radiologe Heft* 1: 10–12, 1969
- 2) Kaplan HA, Aronson SM, Browder EJ: Vascular malformation of the brain on anatomical study. *J Neurosurg* 18: 630–635, 1961
- 3) Lichtor T, Mullan S: Arteriovenous malformation in moyamoya syndrome. Report of three cases. *J Neurosurg* 67: 603–608, 1987
- 4) Luessenhop AJ, Gennarelli TA: Anatomical grading of supratentorial arteriovenous malformations for determining operability. *Neurosurgery* 1: 30–35, 1977
- 5) Sadoshima S, Tanaka K, Ogata J, Fushimi K:

- Capillary telangiectasis of the brain. Case report and the literature. *Rinsho Shinkei* 16: 617-624, 1976 (in Japanese)
- 6) Spetzler RF, Martin NA: A proposed grading system for arteriovenous malformations. *J Neurosurg* 65: 476-489, 1986
- 7) Terao A, Morimoto K, Araki S, Fukai H, Nakagawa S: Diffuse extra- and intracranial malformation. Report of an autopsied case with mental change, epileptic seizures and exophthalmus. *Rinsho Shinkei* 12: 601-608, 1972 (in Japanese)
- 8) Voigt K, Beck V, Reinshagen G: A complex cerebral vascular malformation studied by angiography. Multiple aneurysms, angiomas and arteriolectasia. *Neuroradiology* 5: 117-123, 1973
- 9) Yasargil MG, Teddy PJ, Valavanis A: AVM of the brain. History, embryology, pathological considerations, hemodynamics, diagnostic studies, microsurgical anatomy. *Microsurgery* 4: 76-81, 1981
-
- Address reprint requests to:* Y. Kojima, M.D., Department of Neurosurgery, Yokohama City University School of Medicine, 3-9 Fukuura, Kanazawa-ku, Yokohama 236, Japan.