Multiple Cerebral Arteriovenous Malformations
—Case Report—

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Abstract

Multiple cerebral arteriovenous malformations occurred in a 48-year-old male complaining of
headache, after orthopedic treatment for a leg fracture. He was free from neurological deficits and
signs of hereditary hemorrhagic telangiectasia. Postcontrast computed tomography showed two abnor-
amally enhanced lesions in the right occipital and left parietal regions. Magnetic resonance imaging
showed these lesions as tiny vascular flow void signs, with neither new nor old hemorrhages.
Angiography showed these lesions to be arteriovenous malformations. He declined treatment, and was
followed as an outpatient.

Key words: arteriovenous malformation, cerebrum, multiple

Introduction

Multiple cerebral arteriovenous malformations (AVMs) are rare,11 except when associated with
hereditary hemorrhagic telangiectasia (HHT)8,13,19,23 or unilateral retinocephalic vascular malforma-
tion.17,18,21 Eleven such cases have been reported.1,4,8,9,12,14-16,22 Here, we describe another case
of multiple cerebral AVMs and discuss this and previous cases.

Case Report

A 48-year-old male was referred to us on May 10,
1990, with mild occipitalgia. His left tibia and fibula
were fractured in a traffic accident on May 8. The
fractured bones were set under lumbar anesthesia.
Following the operation, he complained of
headache.

On admission, he was fully alert and cooperative
with no neurological deficits. No clubbing, signs of
mucocutaneous telangiectases or nevi were
discovered. There was no history of dyspnea, recur-
rent episodes of epistaxis or gastrointestinal bleed-
ing. His family history showed no predominance

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of hemorrhagic disorders or cerebrovascular dis-
cases.

Laboratory tests demonstrated leukocytosis, but
no other abnormalities. Roentgenograms and com-
puted tomographic (CT) scans of the chest were nor-
mal, as were roentgenograms of the skull and cer-
vical spine. Postcontrast CT scans of the brain showed
two small enhanced lesions in the right occipital and
left parietal regions. Magnetic resonance (MR) imaging demonstrated these lesions as tiny vascular
flow void signs on both T1- and T2-weighted images.
No new or old hemorrhage was observed (Fig. 1).
Angiography showed the lesions to be AVMs. The
right lesion was fed by the temporo-occipital and the
calcarine arteries, and drained into the superior sagit-
tal sinus through the superior cerebral veins and the
transverse sinus through the inferior cerebral vein
(Fig. 2A, B). The left lesion was fed by the paracentral
tery, and drained into the superior sagittal sinus through the superior cerebral vein (Fig. 2C).

His headache, probably caused by low intracranial
pressure following the lumbar puncture, soon disap-
peared. He refused treatment for the AVMs, so was
followed as an outpatient. Up to May, 1992, he was
free from cerebral hemorrhage, ischemia, and convul-
sion.
Multiple Cerebral AVMs

Fig. 1  MR images of the right (A, B) and left AVMs (C, D), showing no hemorrhage. A, C: T1-weighted images; B, D: T2-weighted images.

Discussion

Multiple cerebral AVMs have multifocal niduses completely separated by normal brain. Angiography is essential for diagnosis of AVMs, but multiple AVMs cannot be easily diagnosed by this method alone. A single nidus with multiple angiographic compartments should be distinguished from multiple niduses. Multiple AVMs usually have either superficial or deep-seated niduses, and with involvement of both exceptional. Postcontrast CT and MR imaging provide important information about niduses, the normal brain between, and angiographically occult AVMs which may manifest after surgical treatment or embolization of large AVMs. Therefore, CT, MR imaging, and angiography should be performed.

The incidence of multiple AVMs was about 1% in a cooperative study. However, other series have found 3% and 6%. An autopsy series found the incidence of multiple angiomas, possibly including vascular malformations other than AVMs, as 6%. The increasing incidence is probably due to improved diagnostic modalities and larger patient populations.

Multiple AVMs fall into two major categories: 1) simple, and 2) complicated with known diseases. The second category includes three subgroups: 1) Rendu-Osler-Weber disease or HHT, 2) Wyburn-Mason syndrome or unilateral retinocephalic vascular malformation, and 3) soft tissue vascular malformation.

HHT is characterized by multiple dermal, mucosal, and visceral telangiectases, recurrent bleeding, and inheritance, commonly with pulmonary arteriovenous shunting. Although telangiectasia is the most common cerebral vascular malformation in HHT (40%), cerebral AVMs have occurred in about 20% of definite or possible HHT cases, including more than 10 multiple AVMs. Some of these cases do not fulfill the criteria for HHT, but were associated with pulmonary arteriovenous shunt which may indicate HHT or a variant.

AVMs involving unilateral retina and optic pathways to the midbrain are typical unilateral retinocephalic vascular malformations. Cerebellar

Fig. 2  A: Right carotid angiogram, showing a small AVM fed by the temporo-occipital artery. B: Right vertebral angiogram, showing the same AVM also fed by the right calcarine artery. C: Left carotid angiogram, showing another small AVM fed by the paracentral artery.

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Table 1 Reported cases of simple multiple AVMs

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/ Sex</th>
<th>Clinical symptom</th>
<th>Location</th>
<th>Treatment for AVMs</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Schlaich et al. (1980)</td>
<td>27/M</td>
<td>hemorrhage</td>
<td>rt parasagittal, rt parietal</td>
<td>total removal</td>
<td>good</td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>29/M</td>
<td>hemorrhage</td>
<td>rt parieto-occipital, lt parasagittal</td>
<td>partial removal</td>
<td>fair</td>
</tr>
<tr>
<td>3</td>
<td></td>
<td>45/M</td>
<td>hydrocephalus</td>
<td>rt frontal, lt frontal, midbrain</td>
<td>partial removal</td>
<td>good</td>
</tr>
<tr>
<td>4</td>
<td>Yamada et al. (1980)</td>
<td>12/F</td>
<td>hydrocephalus (hemorrhage)</td>
<td>rt frontal, rt parietal, lt parietal</td>
<td>radiation</td>
<td>died</td>
</tr>
<tr>
<td>5</td>
<td>Berenstein (1981)</td>
<td>17/M</td>
<td>convulsion</td>
<td>bil basal ganglia and thalamus</td>
<td>embolization</td>
<td>fair</td>
</tr>
<tr>
<td>6</td>
<td>Shigemori et al. (1982)</td>
<td>47/F</td>
<td>hemorrhage</td>
<td>rt temporal, rt parietal, lt parietal</td>
<td>partial removal</td>
<td>good</td>
</tr>
<tr>
<td>7</td>
<td>Stone et al. (1983)</td>
<td>24/M</td>
<td>hemorrhage</td>
<td>rt parietal, lt parietal</td>
<td>total removal</td>
<td>fair</td>
</tr>
<tr>
<td>8</td>
<td>Jones et al. (1986)</td>
<td>31/F</td>
<td>hemorrhage</td>
<td>rt parietal, rt temporal</td>
<td>total removal</td>
<td>good</td>
</tr>
<tr>
<td>9</td>
<td>Reddy et al. (1987)</td>
<td>16/F</td>
<td>convulsion</td>
<td>rt frontal, rt temporal, lt parieto-occipital</td>
<td>total removal</td>
<td>excellent</td>
</tr>
<tr>
<td>10</td>
<td>Nishizawa et al. (1988)</td>
<td>42/M</td>
<td>neuralgia (hemorrhage)</td>
<td>lt parietal, lt cerebellum</td>
<td>total removal</td>
<td>excellent</td>
</tr>
<tr>
<td>11</td>
<td>Kohmura et al. (1990)</td>
<td>23/M</td>
<td>hemorrhage</td>
<td>lt basal ganglia, lt splenium</td>
<td>partial removal</td>
<td>good</td>
</tr>
<tr>
<td>12</td>
<td>Present case</td>
<td>48/M</td>
<td>incidental</td>
<td>lt parietal, rt occipital</td>
<td>none</td>
<td>good</td>
</tr>
</tbody>
</table>

vermis (16%) and occipital lobe (4%) may be involved.17,18 Only one case of multiple AVMs associated with a soft tissue vascular malformation has been reported.19

Differential diagnosis of multiple AVMs is important, because the underlying disease should be treated separately. Our case is clearly different from those complicated with known diseases, and is therefore considered as simple multiple AVMs.

Only 12 cases, including ours, of simple multiple AVMs have been reported (Table 1).1,4,6,9,12,14-16,22 There were two lesions in eight cases and three in four cases. Involvement was bilateral in eight cases and unilateral in four. Clinical symptoms were mostly intracranial hemorrhage, followed by convulsion and hydrocephalus. The incidence of hemorrhage is similar to that for single AVM. The most common location was the parietal lobe, followed by the frontal and temporal lobes, and the basal ganglia. The size of the AVMs, where reported, was small.6,9,16

More aggressive treatment may be indicated for small, superficial, symptomatic AVMs with hemorrhage. Total removal of all multiple lesions has been feasible in five cases.4,9,12,14,16 However, good results have also been obtained where AVMs have been partially removed.5,14,15 The principles of treatment for single AVM also apply to multiple AVMs,10,14 but since problems due to multiplicity may occur,14 treatment strategy should be based on evaluation of hemodynamics and possible alterations after removal of one AVM.

References


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