Carotid Rete Mirabile Associated With Subarachnoid Hemorrhage

—Case Report—

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Abstract

A 47-year-old man presented with carotid rete mirabile manifesting as subarachnoid hemorrhage (SAH). Computed tomography showed SAH, and angiography disclosed an abnormal vascular network around the petrous and cavernous portions of the internal carotid artery. Single photon emission computed tomography (SPECT) with technetium-99m methyl cysteinate dimer revealed reduced regional cerebral blood flow (CBF). Twelve months later, he was leading a normal life without neurological problems, hemorrhage, or ischemic manifestations. SPECT with iodine-123 N-isopropyl-p-iodoamphetamine and the acetazolamide challenge test showed the CBF had normalized.

Key words: carotid rete mirabile, subarachnoid hemorrhage, single photon emission computed tomography

Introduction

Carotid rete mirabile is defined as an arterial network located at the cavernous portion of the internal carotid artery.3) The intracranial internal carotid arteries are supplied mainly through the internal maxillary artery, a branch of the external carotid artery.10,23) This configuration is common in lower vertebrates such as cats, goats, sheep, and pigs.1,3,18) Carotid rete mirabile was first reported in humans in 1953.9) This abnormality is not seen in any developmental stage in humans, but some primitive anastomotic channels in the human embryo may reappear in the adult as collateral vessels circumventing an occluded carotid artery.8) The exact pathogenesis and clinical significance of carotid rete mirabile in humans remain unknown.

Carotid rete mirabile is associated with hypoplasia or non-persistence of the internal carotid artery in humans,3,11) and may be accompanied by other vascular disorders such as carotid-cavernous arteriovenous fistula, splenic vascular malformation, Dieulafoy ulcer, and aortic system,5,12,13) Carotid rete mirabile is important due to its hemorrhagic or ischemic manifestations in patients.5,18) However, changes in the cerebral blood flow (CBF) associated with carotid rete mirabile have not been investigated.

We treated a patient with carotid rete mirabile manifesting as subarachnoid hemorrhage (SAH), and discuss the probable cause of the hemorrhagic disorder.

Case Report

A 47-year-old male presented with sudden onset of severe headache and general feeling of discomfort. On admission, no focal neurological abnormality was observed. Computed tomography demonstrated SAH centering around the left sylvian fissure (Fig. 1). Bone window computed tomography of the skull base showed hypoplasia of the left internal carotid canal. Left carotid angiography demonstrated stenosis of the internal carotid artery at the petrous portion (Fig. 2), dilation of the external carotid artery, and an arterial network around the cavernous portion of internal carotid artery (Fig. 3). Right carotid angiography showed no vascular...
Computed tomography scan demonstrating subarachnoid hemorrhage mainly in the left sylvian fissure.

Lateral (A) and anteroposterior (B) views of the left internal carotid angiogram in the early phase showing stenosis of the left internal carotid artery at the petrous portion, and anastomosis of the dense plexus vessels.

Lateral (A) and anteroposterior (B) views of the left external carotid angiogram in the early phase showing tortuous collateral vessels (rete carotis) originating from the internal maxillary artery and the ascending pharyngeal artery.

Technetium-99m methyl cysteinate dimer single photon emission computed tomography scans showing reduced regional cerebral blood flow in the left middle cerebral artery territory.

Iodine-123 N-isopropyl-p-iodoamphetamine single photon emission computed tomography scans in the resting state (A) and acetazolamide challenge test (B) 12 months after the onset.

abnormality. Repeated angiography detected no cerebral aneurysm or any other vascular disorder. Single photon emission computed tomography (SPECT) using technetium-99m methyl cysteinate dimer at the time of onset revealed the regional CBF was 39.7 ml/100 g/min in the left middle cerebral artery (MCA) territory and 51.8 ml/100 g/min in the right MCA territory, calculated by the Patlak plot method (Fig. 4).
Table 1 Summary of reported cases of carotid rete mirabile

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age</th>
<th>Sex</th>
<th>Ischemic stroke</th>
<th>SAH</th>
<th>ICH</th>
<th>Associated lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Quain et al. (1844)*</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td>multiple scleroses</td>
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<td>2</td>
<td>Fields et al. (1965)</td>
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<td></td>
<td></td>
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<td>BA-top aneurysm</td>
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<td>3</td>
<td>Minagi and Newton (1966)</td>
<td></td>
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<tr>
<td>4</td>
<td>Hawkins and Scott (1967)</td>
<td></td>
<td>M</td>
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<tr>
<td>5</td>
<td>Rockett and Johnson (1968)</td>
<td></td>
<td>M</td>
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<tr>
<td>6</td>
<td>Smith et al. (1968)</td>
<td></td>
<td>F</td>
<td></td>
<td></td>
<td></td>
<td>myocardial infarction</td>
</tr>
<tr>
<td>7</td>
<td>Occleshaw and Garland (1969)</td>
<td></td>
<td>F</td>
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<td>8</td>
<td>Jones and Wetzel (1969)</td>
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<td>M</td>
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<td>9</td>
<td>Austin and Stears (1971)</td>
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<td>M</td>
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<td>10</td>
<td>Danziger et al. (1972)</td>
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<tr>
<td>11</td>
<td>Koo and Newton (1972)</td>
<td></td>
<td>M</td>
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<tr>
<td>12</td>
<td>Araki et al. (1986)</td>
<td></td>
<td>F</td>
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<td>13</td>
<td>Morimoto et al. (1987)</td>
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<td>14</td>
<td>Quint et al. (1992)</td>
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<td>F</td>
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<td>15</td>
<td>Itoyama et al. (1993)</td>
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<td>M</td>
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<td>16</td>
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<td>17</td>
<td>Tanaka et al. (1996)</td>
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<td>M</td>
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<td>18</td>
<td>Hyogo et al. (1996)</td>
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<td>19</td>
<td>Karasawa et al. (1997)</td>
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<td>20</td>
<td>Meder et al. (1997)</td>
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<td>M</td>
<td></td>
<td></td>
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<td>cerebral tumor</td>
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<td>21</td>
<td>Present case</td>
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The patient was treated conservatively and was discharged without neurological deficit. There has been no recurrence of the symptoms for 12 months. SPECT using iodine-123 N-isopropyl-p-iodoamphetamine at 1 year after the onset showed the regional CBF was 42.3 ml/100 g/min in the left MCA territory and 48.0 ml/100 g/min in the right MCA territory, calculated by the autoradiographic method (Fig. 5A). The cerebrovascular perfusion reserve in the left MCA territory was 71.2%, and that in the right MCA territory was 52.3% measured by the acetazolamide challenge test (Fig. 5B).

Discussion

Carotid rete mirabile has six characteristic angiographic features: 1) hypoplasia of the internal carotid artery beginning from the carotid bifurcation, 2) an arterial plexus between the internal maxillary artery and the cavernous portion of the internal carotid artery, 3) dilation of the ophthalmic artery and anastomosis from the internal maxillary artery, 4) the supraclinoid internal carotid artery is not hypoplastic and is fed by the arterial plexus and ophthalmic artery, 5) bilateral lesions, and 6) no anastomosing vessels such as moyamoya disease in the intradural portion. Some atypical features have been described in previous cases. The angiographic frequency of carotid rete mirabile is reportedly 0.01%. Table 1 summarizes the 26 reported cases including our present case. The age of the patients ranged from 2 to 68 years (mean 41.1 years). Twelve of these cases occurred in Japanese patients. Eight cases of carotid rete mirabile were associated with ischemic stroke, 11 cases with SAH, and three cases with cerebral hemorrhage. However, an aneurysm was detected in only three cases with SAH. Repeated angiography found no aneurysm in any other case, and the origin of the SAH was not defined.

The causes of SAH are thought to be rupture of an aneurysm, rupture of anastomosing vessels, and arterial and anastomosis from the internal maxillary artery, 4) the supraclinoid internal carotid artery is not hypoplastic and is fed by the arterial plexus and ophthalmic artery, 5) bilateral lesions, and 6) no anastomosing vessels such as moyamoya disease in the intradural portion. Some atypical features have been described in previous cases.

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and hemodynamic stress. The abnormal vessels of carotid rete mirabile tend to occur in the extradural space, but some may also be found in the intradural space, and the small anastomosing vessels comprising
the carotid rete mirabile may rupture. Hemodynamic stress at the MCA territory through the posterior circulation may cause SAH. Microaneurysms which are angiographically undetectable may also be responsible for hemorrhage.

In our patient, carotid angiography demonstrated abnormal vessels in the extradural space, but no intradural vascular abnormality, so the cause of the SAH was unclear. Regional CBF was reduced in the territory of the left MCA, possibly due to the SAH, but we cannot exclude the possibility that the SAH was caused by the hemodynamic stress associated with the carotid rete mirabile. There is still no consensus regarding the management of carotid rete mirabile. In general, the prognosis is not bad, but one patient died of severe SAH. SAH or ischemic episodes do not always recur, so surgical anastomosis or synangiosis is not required.

References


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Neurol Med Chir (Tokyo) 45, April, 2005