Rete Mirabile in Humans
—Case Report—

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Abstract
Carotid rete mirabile is a physiological vascular network between the external carotid and internal carotid systems present in some vertebrate species, but rarely observed in humans. We describe a 17-year-old girl with rete mirabile who presented with subarachnoid hemorrhage. Angiography disclosed the bilateral internal carotid arteries (ICAs) ended at the cavernous portion, and abnormal arterial networks visualized via the ICAs and the external carotid arteries in the paracavernous region. The distal ICAs were visualized via the abnormal arterial networks. After 18 years of follow-up she is leading a normal life without neurological problems. Rete mirabile in humans may present with hemorrhage or ischemic symptoms, but the prognosis appears to be good.

Key words: arterial rete mirabile, abnormalities, carotid artery

Introduction
Rete mirabile, a physiological vascular network between the external carotid and internal carotid systems, was first described by Herophilus of Alexandria (335-280 B.C.). Rete mirabile occurs in some vertebrates including ungulates and cats. The intracranial internal carotid arteries (ICAs) are supplied via the arterial network (rete mirabile) at the base of the skull mainly through the internal maxillary artery, a branch of the external carotid artery (ECA). The proximal ICA is usually hypoplastic or undeveloped, whereas the ECA is large. Carotid rete mirabile is a very rare pathological condition in humans, with only 11 reported cases. Rete mirabile may manifest as hemorrhage or ischemic cerebrovascular disorders, but the prognosis is unknown.

We report a patient with rete mirabile manifesting as subarachnoid hemorrhage (SAH).

Case Report
A 17-year-old girl suffered sudden onset of severe headache, nausea, and vomiting. Lumbar puncture demonstrated bloody cerebrospinal fluid at a local hospital. She had suffered headache from 1965 to 1975. She was referred to our department on December 14, 1977.

On admission, she was somnolent with severe nuchal rigidity and exaggerated patellar tendon reflex. No motor dysfunction, speech disturbance, or cranial nerve dysfunction was observed.

Right internal carotid angiography (Fig. 1) demonstrated that the right ICA was slightly small and ended at the cavernous portion (C4), where an abnormal network was noted. Portions distal to the anterior knee (C3) of the right ICA were supplied via the abnormal network. The middle cerebral artery (MCA) was normal. The anterior cerebral artery (ACA) and posterior cerebral artery (PCA) were not visualized. Right external carotid angiography (Fig. 2) demonstrated that the right ECA was larger than the right ICA. The distal portion of the ipsilateral ICA received blood supply via the deep temporal artery, the internal maxillary artery, and the middle meningeal artery, and as a result the MCA was well visualized. The MCA and ophthalmic artery were both better visualized by right external than right internal carotid angiography. Left internal carotid angiography (Fig. 3) demonstrated that the left ICA ended at the C4, where the abnormal network was noted. The C3 portion of the left ICA was supplied via the
abnormal vascular network. The left ophthalmic and posterior communicating arteries were not visualized. The ACAs were visualized bilaterally via the left ICA. Left external carotid angiography (Fig. 4) demonstrated that the large ophthalmic artery and the C3 portion of the left ICA were supplied via a
number of abnormal arteries through an abnormally large internal maxillary artery and middle meningeal artery. The left ICA was better visualized by left external than left internal carotid angiography. Left vertebral angiography (Fig. 5) showed that the left PCA was not visualized in the peduncular segment, and portions distal to the ambient segment of the left PCA were supplied via the abnormal arterial network. The abnormal arterial network was fed by the anterior and posterior thalamoperforating arteries.

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The left ICA and left MCA were normograderely visualized via the left posterior communicating artery, which was supplied via the left PCA. A portion of the right ACA was retrogradely visualized via the posterior pericallosal artery. Right vertebral angiography showed the right vertebral artery was normal.

The bilateral ICAs ended at the C4 portions, and an abnormal arterial network (carotid rete mirabile) was visualized via the ICA and the ECA in the paracavernous region. C3 portions were visualized via the abnormal arterial network. These findings were similar bilaterally. Cerebral angiography detected no aneurysm or arteriovenous malformation (Fig. 6).

Four days after admission, she developed mild left hemiparesis which improved within 4 days. She was alert without neurological deficits at discharge on February 2, 1978. In December 1995, after a 18-year follow-up period, she was a mother with a child and leading a normal life without neurological symptoms.

**Discussion**

Previous cases of rete mirabile in humans are summarized in Table 1. In most cases of carotid rete mirabile, the ICA ended in the ganglionic portion (C5), in which the rete mirabile was present, and portions distal to the supraclinoid portion of the ICA were supplied via the rete mirabile. The abnormal portion of the ICA was 0.5 to 1 cm long, and the rete mirabile was located between the base of the skull and dura mater. The circle of Willis was normal. The ascending pharyngeal artery and occipital artery are occasionally associated with rete mirabile.10) In our case, the bilateral ICAs ended at C4, and the distal portions were supplied via collateral circulation from the carotid rete mirabile. These anastomotic channels were supplied via the ICAs and via the internal maxillary artery of the ECA.

Five patients with rete mirabile, including ours, presented with SAH, and two patients presented with episodes of ischemia. Two of the five patients with SAH harbored aneurysms, and the cause of SAH was unknown in three patients. In our case, angiography detected no aneurysm. The cause of SAH was considered to be related to the abnormal vascular networks. The left hemiparesis noted 4 days after admission was thought to be due to vasospasm. The prognosis for patients with rete mirabile appears to be good, although one patient with rete mirabile died as a result of severe SAH.31 No recurrence of SAH or ischemic episodes was noted in the other patients. Our patient is presently leading a normal life without neurological problems 17 years after the diagnosis of rete mirabile.

Carotid rete mirabile in lower mammals is thought to provide heat exchange to prevent overheating of the brain,1) and protection of the brain by regulation of the pressure and flow of the cerebral circulation.4,14) These effects are of minor importance in humans. Rete mirabile does not occur in any stage of normal human development,1) so is thought to be an anomalous atavistic development. However, the exact pathogenesis and clinical significance of rete mirabile in humans remain unknown. The five most recent patients were all Japanese. Moyamoya disease, a cerebrovascular occlusive disease, is also prevalent in the Japanese.

**Table 1 Rete mirabile in humans**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age</th>
<th>Sex</th>
<th>Side</th>
<th>Diagnosis</th>
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<tbody>
<tr>
<td>1</td>
<td>Quain (1844)*</td>
<td>ND</td>
<td>ND</td>
<td>lt</td>
<td>ND</td>
</tr>
<tr>
<td>2</td>
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<td>ND</td>
<td>bil</td>
<td>ND</td>
</tr>
<tr>
<td>3</td>
<td>Minagi and Newton (1966)</td>
<td>43</td>
<td>M</td>
<td>lt</td>
<td>hypoplastic ICA</td>
</tr>
<tr>
<td>4</td>
<td>Hawkins and Scott (1967)</td>
<td>37</td>
<td>M</td>
<td>bil</td>
<td>SAH with aneurysm</td>
</tr>
<tr>
<td>5</td>
<td>Rockett and Johnson (1968)</td>
<td>40</td>
<td>M</td>
<td>bil</td>
<td>SAH</td>
</tr>
<tr>
<td>6</td>
<td>Rios-Montenegro et al. (1972) and Koo and Newton (1972)</td>
<td>20</td>
<td>M</td>
<td>bil</td>
<td>CCF, PXE</td>
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<tr>
<td>7</td>
<td>Danzinger et al. (1972)</td>
<td>39</td>
<td>F</td>
<td>bil</td>
<td>fainting</td>
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<td>8</td>
<td>Araki et al. (1986)</td>
<td>55</td>
<td>M</td>
<td>bil</td>
<td>SAH, PXE</td>
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<td>F</td>
<td>bil</td>
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<td>10</td>
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<td>40</td>
<td>M</td>
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<td>12</td>
<td>Present case</td>
<td>17</td>
<td>F</td>
<td>bil</td>
<td>SAH</td>
</tr>
</tbody>
</table>


**References**

4) Edelman NH, Epstein P, Cherniack NS, Fishman AP:

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