A Pediatric Case of Carotid Rete Mirabile

Isao Fuwa, MD

Background Carotid rete mirabile (CRM) is a pathological network between the external carotid and internal carotid systems in lower mammals. Very rarely, these arterial channels are observed in humans.

Case Description We describe a 13-year-old girl with CRM who presented with acute hemiplegia after an operation for Dieulafoy's ulcer, a submucosal vascular anomaly of the stomach. Angiogram revealed hypoplasia of the bilateral internal carotid arteries. The abnormal network from the external carotid system was seen around the cavernous portion of the internal carotid arteries. The ophthalmic arteries were developed as the collateral pathway.

Conclusions To our knowledge, this is the first pediatric case reported in the literature. CRM sometimes accompanies intracranial or systemic vascular disorders such as cerebral aneurysm and pseudoxanthoma elasticum. CRM is a unique pathological condition that presents as a hemorrhagic or ischemic cerebrovascular disorder.

Key Words carotid rete mirabile • carotid stenosis • cerebral infarction • child • hemiplegia

Case Report

A 13-year-old girl was referred to us after suffering the sudden onset of right hemiparesis just after undergoing a gastrectomy. She received the operation because of gastrointestinal hemorrhage from Dieulafoy's ulcer, a submucosal vascular anomaly of the stomach. On admission she was parietic, and computed tomographic scan demonstrated a small low-density area in the left basal ganglia.

On the carotid angiogram, the right internal carotid artery (ICA) was gradually tapered at the cavernous portion. A small sinuous arterial network was seen around the lesion. The dilated ophthalmic artery was connected with small arteries originating from the maxillary artery. An accessory meningeal artery also connected with the ophthalmic artery (Fig 1). On the carotid angiogram, the left ICA was stenotic at the supraclinoid segment. Small arterial channels from the maxillary artery connected with the dilated ophthalmic artery. The abnormality was less prominent than on the right side (Fig 2). No abnormal vessels were seen on the vertebral angiogram. The patient was treated medically, and no ischemic attack has occurred in the past 6 years.

Discussion

Carotid rete mirabile is a very rare pathological condition; to our knowledge, only six cases have been reported in the English and Japanese literature (Table). All previously reported case subjects were adults (average age, 39.0 years). To our knowledge, ours is the first patient who presented with symptoms of CRM in childhood.

Collateral channels exist normally between the cavernous portion of the ICA and the external carotid artery. These anastomoses may enlarge and become roentgenographically visible in instances of hypoplasia, surgical ligation, or arteriosclerotic occlusion of the carotid artery. However, CRM has unique characteristics. Araki et al1 noted six specific points on angiograms: (1) hypoplastic ICA, (2) arterial plexus between the maxillary artery and the cavernous portion of the ICA, (3) dilated ophthalmic artery, (4) supraclinoid ICA not hypoplastic and fed by the arterial plexus and the ophthalmic artery, (5) bilateral lesions, and (6) no abnormal vessels such as moyamoya in the intradural portion. CRM is distinguished from moyamoya disease in that moyamoya vessels are not present at the base of the brain and the occluded site is not at the carotid bifurcation. This reported case was not typical according to the aforementioned criteria. The left ICA was stenotic at the supraclinoid portion, and the meningeal network was not developed around the cavernous portion.

Three of the previously described patients had a subarachnoid hemorrhage (SAH); however, aneurysm was found in only one patient. In the other two patients, the cause of SAH was unknown. An abnormal network around the cavernous portion of the ICA may lead to SAH. Three of the patients in earlier reports had experienced an ischemic cerebrovascular episode. In our case, an episode of acute infantile hemiplegia occurred just after the patient had received general

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FIG 1. The right internal carotid artery (ICA) is gradually tapered at the cavernous portion on the carotid angiogram (arrowheads). A small sinuous arterial network is seen around the cavernous portion of the ICA (arrow). The dilated ophthalmic artery is connected with small arteries originated by the external carotid artery (arrowhead). The anterior and middle cerebral arteries are not involved.

FIG 2. The left internal carotid artery is stenotic at the supraclinoid segment on the carotid angiogram (small arrow). Small arterial channels from the maxillary artery connect with the dilated ophthalmic artery (arrowhead). The abnormality is less prominent than on the right side.

Reported Cases of Carotid Rete Mirabile

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age, y</th>
<th>Sex</th>
<th>SAH</th>
<th>Infarction</th>
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<td>43</td>
<td>M</td>
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<td>M</td>
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<td>M</td>
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<td>20</td>
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<td>CCF, PXE</td>
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<tr>
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<td>1993</td>
<td>13</td>
<td>F</td>
<td>-</td>
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<td>Dieulafoy's ulcer</td>
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</table>

SAH indicates subarachnoid hemorrhage; CCF, carotid cavernous fistula; and PXE, pseudoxanthoma elasticum.

*Same case as that reported by Rios-Montenegro et al.
anesthesia. CRM is an important cause of ischemic cerebrovascular disorder in pediatric patients.

This unique abnormality is associated with some systemic or intracranial vascular disorders. Two reported cases involved pseudoxanthoma elasticum,1,7 which is a very rare congenital disease associated with intracranial aneurysms or carotid cavernous fistulas caused by vascular wall fragility.1,7 A splenic vascular anomaly was present in the patient described by Rios-Montenegro et al.7 Our patient had Dieulafoy's ulcer, a vascular anomaly in which the artery is exposed on the mucous surface of the stomach.10,11 Considering the association of these vascular abnormalities, CRM may be seen in patients with a congenital systemic anomaly of the arterial system. The prognosis is not bad in these patients: only one of the described patients died as a result of severe SAH.8 SAH and ischemic episodes did not recur in the other patients, and no patients with CRM underwent surgical anastomosis or synangiosis.

References
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