Moyamoya syndrome as a complication of radiation therapy

Case report

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A case is reported with occlusion and stenosis of the internal carotid arteries in association with basal telangiectasia. Fifteen years after postoperative irradiation for an optic glioma, radiological signs typical of the moyamoya syndrome were observed. Radiation therapy is discussed as the cause of the vascular damage in this case.

Key Words: moyamoya syndrome, occlusive vascular disease, radiation therapy, carotid artery occlusion, optic glioma

The etiology of the disease of multiple progressive intracranial arterial occlusion, also known as the moyamoya syndrome, is unclear. A congenital etiology has been suggested, but acquired forms have also been shown to exist. In the neurocutaneous syndromes of children, radiographic features similar to those in moyamoya are found. Intracranial arterial occlusive disease in combination with neurofibromatosis has previously been reported in 14 cases. Nine of these had the moyamoya appearance, and three of the nine patients had received previous radiation therapy for optic gliomas. Intracranial occlusive disease secondary to radiation has been reported. We report an additional case of neurofibromatosis with cerebral arterial occlusive disease after radiation therapy.

Case Report

This 17-year-old boy presented for neurological evaluation. At the age of 1 1/2 years, he had received a course of irradiation, with a total of 4200 rads, postoperatively for an optic glioma. The irradiation was given over a period of 4 weeks through two opposing temporal ports and one anterior port. During the operation before irradiation, the anterior cerebral arteries were identified on both sides and seen to be normal. The glioma compressed the optic chiasm and extended below the arteries. After irradiation the patient recovered well, except that the left eye was blind and vision in the right eye was impaired. Ten years later, at the age of 12 years, the patient ceased to grow. Panhypopituitarism was diagnosed and substitution therapy was started.
For the past 4 years before his present admission, the patient had had periodic attacks of headache, nausea, and vomiting. He also had numerous café-au-lait spots. He had never suffered hemiparesis.

On admission, 15 years after radiation therapy, left common carotid angiography showed that the internal carotid artery was occluded at the origin of the ophthalmic artery (Fig. 1 right). A dense network of small ethmoidal arteries arose from the ophthalmic artery, which was thick, tortuous, and displaced inferiorly. There was early opacification of transdural meningocortical anastomoses from the middle meningeal and superficial temporal arteries (Fig. 1 left). These anastomoses and the ethmoidal network together supplied the parietal branches of the anterior cerebral artery. Since the middle cerebral artery was not visualized, the artery obviously originated from the basilar artery by collaterals.

Right common carotid angiography showed that the internal carotid artery had a stenosis at the origin of the ophthalmic artery. An ethmoidal arterial network was seen here, too, but there were no transdural meningocortical anastomoses. Above the stenotic carotid siphon originated an intricate and dense vascular network, which involved the whole region of the basal nuclei (Fig. 2). The posterior cerebral artery was thick and the posterior choroidal artery participated in this basal arterial network. A rapid collateral circulation to both pericallosal arteries emerged from this network (Fig. 3 left), and there was collateral circulation to the
branches of the middle cerebral artery from the posterior cerebral artery (Fig. 3 right). These findings are typical of the moyamoya syndrome.

Discussion

Vascular lesions of the cerebral arteries in neurocutaneous syndromes are well known. In neurofibromatosis, occlusive lesions are found mostly in the peripheral leptomeningeal arteries, but the internal carotid artery may also be occluded. Table 1 summarizes five previously reported cases with typical radiographic pictures of moyamoya syndrome. All five patients had received radiation therapy several years before the radiological examination was made. No angiograms were made on these patients before irradiation; the diagnosis of optic or hypothalamic gliomas was based on echoencephalography. In three of the patients the occlusions may have resulted from neurofibromatosis as well as irradiation.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Diagnosis</th>
<th>Neurofibromatosis</th>
<th>Irradiation Dosage (rads)</th>
<th>Time Interval*</th>
<th>Angiogram Side</th>
<th>Occlusion</th>
<th>Basal Telangiectasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Debrun et al., 1975</td>
<td>hypothalamic glioma</td>
<td>−</td>
<td>5500</td>
<td>15 mos</td>
<td>right</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Hilal et al., 1971</td>
<td>optic glioma</td>
<td>+</td>
<td>3000</td>
<td>10 yrs</td>
<td>right</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Klatte et al., 1976</td>
<td>optic glioma</td>
<td>+</td>
<td>2400</td>
<td>7 yrs</td>
<td>left</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Lee &amp; Hodes, 1967</td>
<td>optic glioma</td>
<td>+</td>
<td>5500</td>
<td>3 yrs</td>
<td>left</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Servo &amp; Puranen, 1978</td>
<td>optic glioma</td>
<td>−</td>
<td>4200</td>
<td>15 yrs</td>
<td>right</td>
<td>−</td>
<td>+</td>
</tr>
</tbody>
</table>

*Time elapsed between irradiation and radiological examination.
Radiation therapy and moyamoya syndrome

In the two patients without neurofibromatosis, the carotid occlusion was probably secondary to radiation therapy.

There have been reports of arterial stenosis following irradiation in infancy and carotid thrombosis after therapeutic radiation in adults. Postirradiation myelomalacia has also been reported, where a selective spinal cord arteriography was made before and after radiotherapy, and spinal cord arterial occlusion was seen in the postirradiation arteriogram. In young patients, the basal telangiectasia has been considered to be a nonspecific response to a slow occlusive process in the circle of Willis. Although in our case no angiogram was made before radiation therapy, at the operation of the glioma the anterior cerebral arteries on both sides had been identified and regarded as normal. The occlusive process may therefore have been caused by irradiation.

Opinions differ on the advisability of irradiation of optic gliomas in children. In two recent reports radiation therapy was considered contraindicated in anterior chiasmal gliomas.

References


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