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## Posterior Circulation Intracranial Arterial Occlusive Disease in Neurofibromatosis

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It is well known that neurofibromatosis is one apparent etiology for the type of intracranial arterial occlusive disease known as moyamoya disease [1-4]. All previously reported cases of neurofibromatosis with moyamoya disease have had involvement only of the intracranial carotid circulation (supraclinoid internal carotid artery, middle cerebral artery, anterior cerebral artery) [1-4]. We report a case of neuro-

fibromatosis with moyamoya disease demonstrating progressive posterior circulation involvement.

### Case Report

An 11-year-old girl presented with left hemiparesis, mild mental retardation, and numerous café-au-lait spots. Her father and sister

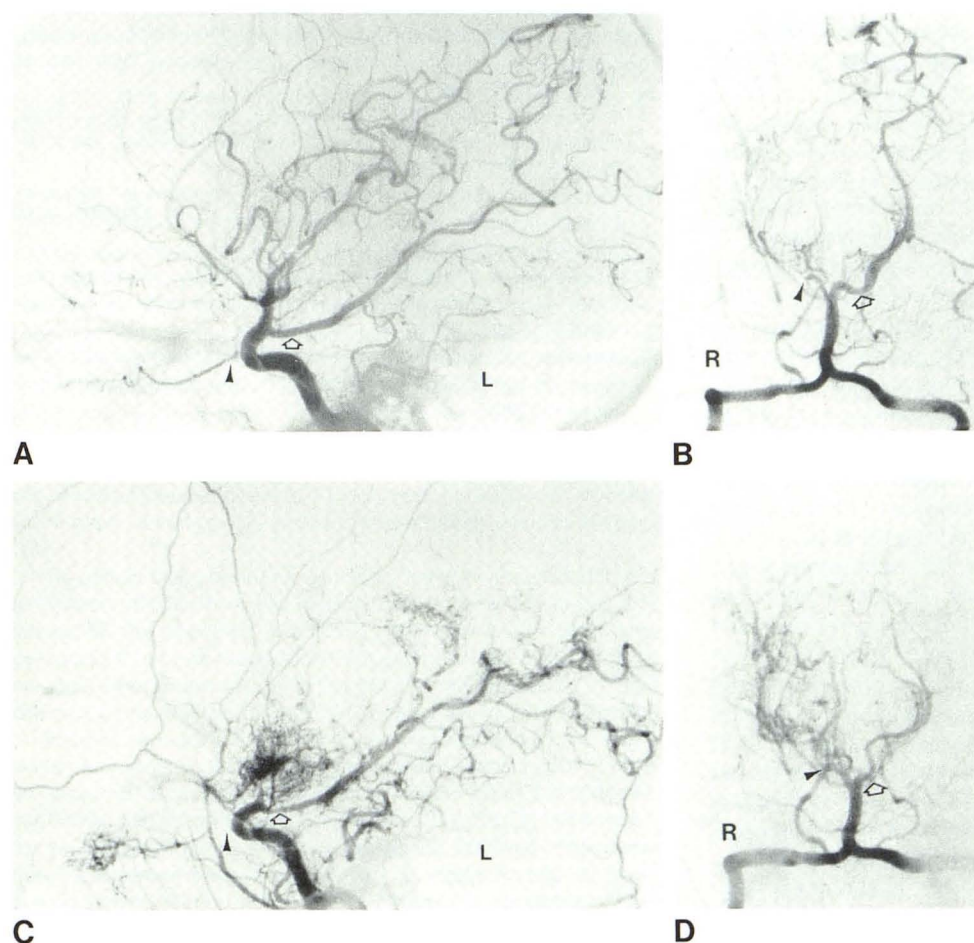


Fig. 1.—Lateral film of left internal carotid angiogram at 11 years of age. Minimal supraclinoid internal carotid artery stenosis. Collateralization is caused by marked proximal anterior and middle cerebral artery stenoses apparent on anteroposterior (AP) views (not illustrated). Left ophthalmic artery (arrowhead) and left posterior communicating artery (arrow) are normal. B, AP film of right vertebral angiography at 11 years of age. Stenosis of proximal right posterior cerebral artery (arrowhead). Proximal left posterior cerebral artery (arrow) is normal. Caliber of left posterior cerebral artery proximal to posterior communicating artery is smaller than that distal to posterior communicating artery, consistent with presence of congenitally large posterior communicating artery on this side. C, Lateral film of left common carotid angiography at 15 years of age. Marked progression of supraclinoid internal carotid artery stenosis and further collateralization. Interim development of occlusion of proximal left ophthalmic artery (arrowhead) with collateral opacification from external carotid artery and irregular stenosis of left posterior communicating artery (arrow). D, AP film of right vertebral angiography at 15 years of age shows probable progression of right posterior cerebral arterial stenosis (arrowhead), more apparent when serial films are inspected. Interim development of proximal left posterior cerebral artery occlusion (arrow). Vessel appearing to extend from left posterior cerebral artery stump actually is superimposed left superior cerebellar artery (cf. B).

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had also experienced episodes of transient hemiparesis and had café-au-lait spots. Cranial computed tomography (CT) before and after intravenous administration of contrast material showed changes consistent with a recent right middle cerebral artery territory infarction. Complete cerebral angiography revealed marked stenoses of the right supraclinoid internal carotid, both anterior cerebral arteries proximally, and the left proximal middle cerebral artery. Minimal stenoses of the left supraclinoid internal carotid artery and the right proximal middle cerebral artery were also present. The left ophthalmic artery and posterior communicating artery were normal (fig. 1A), but the proximal part of the right posterior cerebral artery was involved (fig. 1B). Anticoagulation therapy was started, and the left hemiparesis cleared completely in about 4 weeks. However, at age 15 years, the patient developed a mild right hemiparesis and marked expressive aphasia. Cranial CT before and after intravenous administration of contrast material showed changes consistent with recent left and old right middle cerebral artery territory infarctions. Repeat complete cerebral angiography demonstrated marked progression of moyamoya disease including lush, increased development of collateral channels. There was interim development of an occlusion in the proximal part of the left ophthalmic artery, moderate stenosis of the left posterior communicating artery (fig. 1C), occlusion of the proximal part of the left posterior cerebral artery, and probable progression to a more severe stenosis in the proximal part of the right posterior cerebral artery (fig. 1D). The right hemiparesis resolved within 7 days, but the expressive aphasia remained unchanged during a month of follow-up.

### Discussion

In cases of neurofibromatosis, moyamoya disease changes are thought to be secondary to angiodysplasia. All previously reported cases of neurofibromatosis with moyamoya disease have shown curious, apparent sparing of the posterior circulation. Specifically, the posterior communicating artery, posterior cerebral artery, basilar artery, and cerebellar arteries have not been reported to be involved with moyamoya disease in neurofibromatosis [1-4]. In other

cases of moyamoya disease without neurofibromatosis, posterior circulation involvement generally is seen only in later stages [5-7]. Our case suggests that the apparent sparing of the posterior circulation in neurofibromatosis with moyamoya disease may be caused by the relatively few cases reported and the rarity of follow-up angiography in these cases, rather than by a unique feature of moyamoya disease in neurofibromatosis not found in other cases.

It is also interesting to note the dramatic development of a left ophthalmic artery occlusion in our case. Although ophthalmic artery occlusive disease is well documented in moyamoya disease [5, 6], it has been reported previously in only a single patient with neurofibromatosis and moyamoya disease. That patient had received radiation therapy for an ipsilateral optic glioma before angiography [1].

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