

Angiographically verified progression of moyamoya disease in an adult

Case report

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✓ The authors present the case of a 37-year-old man with definite moyamoya disease in whom angiographic findings drastically changed. The patient presented with left hemiparesis due to lacunar infarction. Angiography initially disclosed a narrow right carotid artery (CA) siphon and severe stenosis of the horizontal segment of the left middle cerebral artery. Four years later, the patient experienced right-central facial paresis, which developed because of a small putaminal hematoma. Angiography results demonstrated occlusion of the internal CA siphons bilaterally, with moyamoya vessels. It therefore appears that in some adults, moyamoya disease is accompanied by very progressive vascular changes.

KEY WORDS • moyamoya disease • progressive disease • angiography

MOYAMOYA disease—spontaneous occlusion of the circle of Willis—is progressive. It is generally believed that in children moyamoya disease advances in angiographically visible stages as classified by Suzuki,¹² whereas in adults the disease rarely progresses in stages.^{2,12} Whether an angiographically documented progression of changes from Suzuki's early stage occurs in adult as well as in pediatric cases is unknown. In this article we report an adult case of definite moyamoya disease with drastic angiographically documented changes progressing from Suzuki's early stages.

Case Report

This 37-year-old man presented with left hemiparesis in May 1994. Magnetic resonance T₂-weighted imaging demonstrated a small high-intensity lesion in the man's right corona radiata. An angiogram of the CA revealed a narrow right CA siphon (Fig. 1 *upper*) and severe stenosis of the horizontal segment of the left MCA (Fig. 1 *center*). An angiogram of the vertebral artery demonstrated leptomeningeal anastomoses in both temporoparietal areas (Fig. 1 *lower*). The patient's hemiparesis improved in response to conservative therapy. Results of single-photon emission computerized tomography scanning performed in April 1996 indicated a low level of perfusion in the frontal lobes bilaterally. Follow-up magnetic resonance imaging performed in November 1996 revealed elimination of the bilateral horizontal segments of the MCA flow void. The patient complained of mild dysarthria while at

work in January 1998. His Glasgow Coma Scale score was 15, with minimal right-central facial paresis. Computerized tomography scanning results revealed a small hemorrhage in the patient's left putamen. An angiogram demonstrated complete obliteration of the CA siphons bilaterally with typical moyamoya vessels (Fig. 2). The patient's facial paresis improved in response to conservative treatment.

Discussion

This patient had no history of cranial insult or causative systemic disease and his ICAs were occluded bilaterally at their terminal portions with moyamoya vessels. Therefore, the diagnosis in this case was definite moyamoya disease in accordance with the guidelines published by the Research Committee on the Spontaneous Occlusion of the Circle of Willis of the Ministry of Health and Welfare, Japan.⁴

Progressive angiographically verified vascular changes in adults with moyamoya disease are thought to be extremely rare. Suzuki and Kodama¹² suggested that in children the disease is dynamic and progressive, whereas in adults it is static and arrested. Ezura, et al.,² reported that moyamoya disease progresses in angiographically evident stages until adolescence and that this progression stabilizes or slows significantly after the patient reaches adulthood. Houkin, et al.,⁷ have speculated that moyamoya disease progresses very quickly during early childhood, especially in children younger than 2 years of age, may progress slowly after the early pediatric stage, and become stable later in childhood and adolescence.

Although a few cases of moyamoya disease with angio-

Abbreviations used in this paper: CA = carotid artery; ICA = internal CA; MCA = middle cerebral artery.

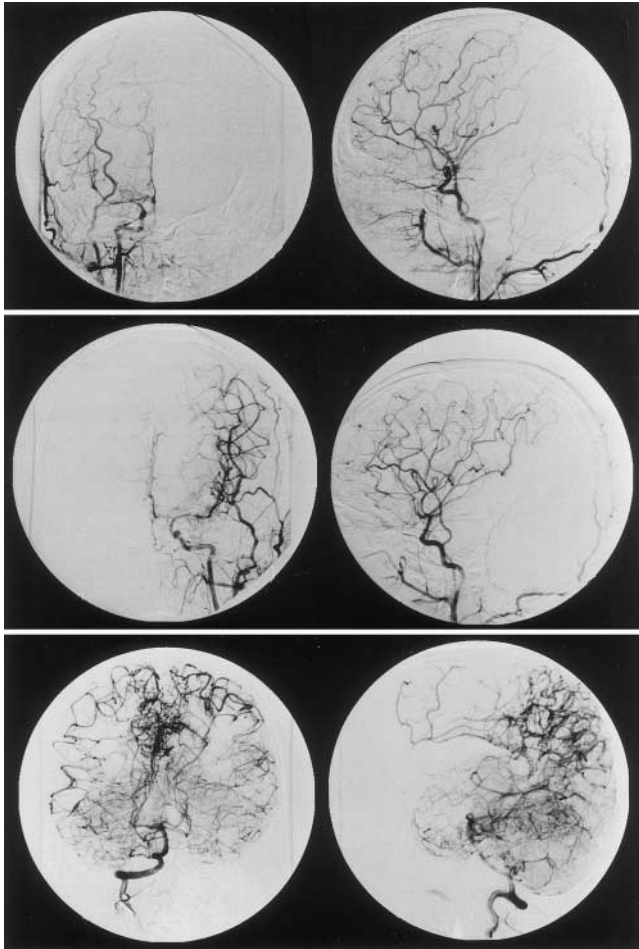


FIG. 1. Angiograms obtained in May 1994. *Upper:* Angiograms of right CA revealing narrowing of the entire right CA siphon. *Center:* Angiograms of left CA demonstrating severe stenosis of the middle portion of the horizontal segment of the left MCA. *Lower:* Angiograms of left vertebral artery demonstrating leptomeningeal anastomoses from the posterior cerebral artery to the temporoparietal area.

graphically evident changes have been reported in adults, in most reported cases this is believed to have developed from probable moyamoya disease.^{3,8,9,14} Most probable adult cases are considered different from definite moyamoya disease in that they are thought to exhibit no changes in angiography studies.⁶ However, Wanifuchi, et al.,¹⁴ have strongly suggested that probable moyamoya disease, in which ischemic stroke is the initial manifestation, may develop into definite moyamoya disease. We reviewed three reported definite adult cases in which there were angiographically evident changes that did not develop in cases formerly deemed probable.^{1,10,11} However, in those cases the changes observed in angiography studies were slight compared with those in the present case.

It remains unknown when angiographically visible changes in the CA siphon begin in adults with moyamoya disease. In our case, at the time of the initial ischemic attack, angiography revealed no evidence of moyamoya vessels on either side and the lesions of both CA siphons were not specific to moyamoya. In most cases of child-

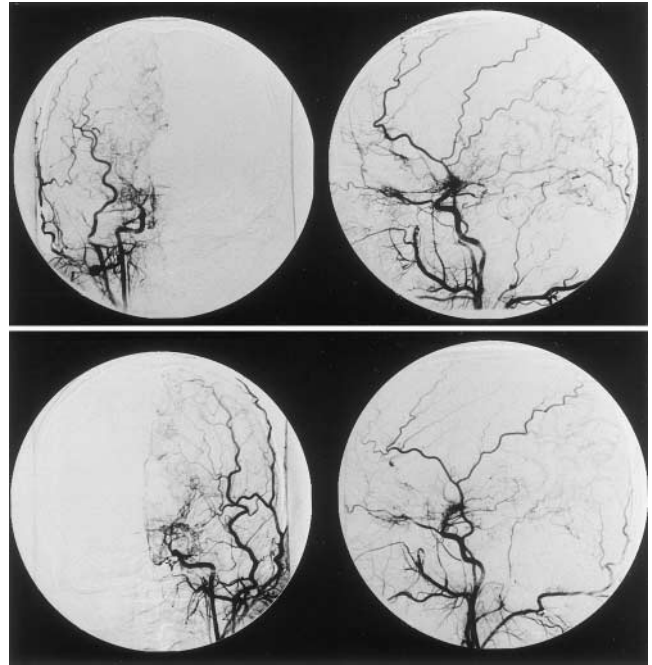


FIG. 2. Angiograms obtained in January 1998. *Upper:* Angiograms of right CA indicating complete occlusion of the terminal portion of the ICA with moyamoya vessels. *Lower:* Angiograms of left CA indicating total occlusion of the ICA and basal moyamoya vessels.

hood-onset moyamoya disease, obstruction of the CA siphon rapidly progresses with moyamoya vessels within 2 or 3 years after the first attack.⁵ The changes observed on angiograms in our case were like those seen in childhood-onset cases. Thus, the findings in our case suggest that some angiographically documented changes in the CA siphon begin in adulthood in adult-onset moyamoya as in child-onset moyamoya. Takahashi, et al.,¹³ have speculated that there may be cases of angiographically verified cases of adulthood-onset moyamoya disease, based on the fact that among those who suffer from this disorder the incidence of mental retardation is lower in the adult than in the pediatric population.

It is generally believed that stenosis may begin from the terminal portion of the ICA or the proximal portion of the MCA and/or the anterior cerebral artery.¹² In our case, the stenosis of the left CA siphon began in the middle portion of the MCA. This suggests that, in some cases, stenosis may begin at more peripheral sites of the CA siphon than previously believed.

The present case demonstrates that moyamoya disease with onset in adulthood, although very rare, includes drastic angiographically evident changes. Even in adult cases of moyamoya disease, clinicians should consider the possibility of an early stage in which lesions occur in the CA siphon with abundant collateral flow from the posterior circulation, even if angiographic studies fail to disclose moyamoya vessels.

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Moyamoya disease with vascular change in an adult

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