Moyamoya disease in monovular twins

Case report

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The first reported instance of identical twins with moyamoya disease is presented. The involved portions of the main intracranial arteries were similar, but the formation of the moyamoya vessels was somewhat different. The cause of obstructive changes in large intracranial arteries remains obscure, but the frequent familial occurrence suggests that the initial stages of this disease and the formation of the moyamoya vessels occur in the prenatal period.

KEY WORDS: moyamoya disease, cerebral angiography, congenital disorder, cerebrovascular disease

Once considered to be a specifically Japanese entity, moyamoya disease or syndrome has been described in patients of other races. Two types of moyamoya syndrome have been differentiated; the infantile form, characterized clinically by recurring transient hemiparetic attacks, both uni- and contralateral, and the adult form, usually made manifest by subarachnoid hemorrhage.

In 1975, Kudo reviewed the moyamoya syndrome in Japan. He found seven instances of familial occurrence among 363 cases of moyamoya disease. Kitahara, et al., collected 23 families from the literature, and added three familial cases of their own. We have found two other reports of familial involvement. However, to our knowledge, there has been no previous report of moyamoya syndrome in monovular twins.

Case Report

These patients were 4-year-old identical twin girls, born at term after a normal pregnancy and delivery. The 31-year-old father and the 28-year-old mother were Japanese, in good health, and of normal intelligence. An 8-year-old brother was normal. There was no family history of vascular disease or other congenital anomalies. The babies presented no feeding problems, sat at 6 months, walked at 13 months, and had no serious illness.

Case 1

This patient developed normally until 4 years of age, when she had episodes of headache, and exhibited weakness of the left side of the body, which cleared within 1 hour. Convulsions were not noted. She was admitted to Nagoya University Hospital.

Physical examination on admission was normal, and routine laboratory data were within normal limits. Radiographs of the skull were normal. Left carotid angiography revealed marked stenosis of the supraclinoid portion of the internal carotid artery, the proximal portion of the anterior cerebral artery, and the middle cerebral artery. The distal branches of the left anterior cerebral artery were opacified through an abnormal network of anastomotic vessels that arose from the posterior cerebral arteries (Fig. 1). Right brachial angiography showed that the internal carotid artery was very narrow from its origin, and was occluded at the level of the posterior communicating artery (Fig. 2). The right anterior and middle cerebral arteries were not opacified, even in the late stages of the angiograms. Significant numbers of small vessels arose from the posterior communicating and posterior...
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FIG. 1. Case 1. Left carotid angiogram, anteroposterior (left) and lateral (right) projections. The anterior cerebral and middle cerebral arteries are stenotic at their origin. The distal branches of these vessels are opacified by the anastomotic network of vessels arising from the posterior communicating and posterior cerebral arteries.

FIG. 2. Case 1. Right brachial angiogram, lateral projection, of the cervical portion, showing the bilateral carotid and right vertebral arteries and hypoplasia of the internal carotid artery, the diameter of which is smaller than that of the vertebral or external carotid arteries. There is stenosis of the supraclinoid internal carotid artery, and major filling of the posterior communicating artery.

cerebral arteries. These vessels formed an intricate and dense vascular network that involved the whole region of the basal nuclei (Fig. 3).

Case 2

At 2 years of age, this little girl had her first episode of right hemiconvulsion and a transient right hemiparetic attack. After that, she had three similar episodes with complete recovery. At 3 years and 10 months of age, she was admitted to Nagoya University Hospital.

On examination, she had no motor deficits, and neurological examination was essentially normal. Right brachial angiography disclosed marked stenosis of the supraclinoid carotid artery, and very poor filling of the cerebral branches of the internal carotid artery. A dense network of vessels was seen, filled mainly by branches of the posterior communicating and posterior cerebral arteries (Fig. 4). Left carotid angiography was performed with visualization of only the external carotid artery, which showed dural anastomosis and hypertrophy of the subcutaneous branches of the superficial temporal artery (Fig. 5).

Discussion

Various explanations have been offered for the etiology of this syndrome, but it is still not fully understood. Most authors consider the carotid stenosis to be the primary condition, and the moyamoya vessels to be the result of it. Progressive arterial occlusion has been demonstrated by several authors,
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**Fig. 3.** Case 1. Right brachial angiograms, lateral (*left*) and anteroposterior (*right*) projections. Marked narrowing of the internal carotid artery at the origin of the opthalmic artery and a dense network of small arteries supplied from the posterior cerebral arteries are seen in the central portion of the right hemisphere.

**Fig. 4.** Case 2. Right brachial angiograms, lateral (*left*) and anteroposterior (*right*) projections. The internal carotid artery ends just above the origin of the posterior communicating artery. An extensive network of anastomotic vessels supplied by the posterior communicating, posterior choroidal, and thalamoperforating arteries is seen.
angiographic vascular patterns of these twins were similar but not entirely identical, with some differences in the severity and formation of the moyamoya vessels.

We believe that the cause and initial stages of the stenotic or obliterative changes in the large intracranial arteries, and even the development of the collateral moyamoya vessels, begin in the prenatal period.

with stenosis gradually reaching the anterior and middle cerebral arteries, and then spreading to the communicating and posterior cerebral arteries along with an abnormal vascular network. The neurological status of the infantile type of moyamoya disease also shows a progressive course, with gradual motor deficit and intellectual deterioration.

An acquired type of moyamoya disease has been documented. The collateral network has been described in cases of tumor, basal meningitis, and after radiation therapy. But the angiographic appearance of these cases seems to be somewhat different from those of the "idiopathic" type of moyamoya disease. The acquired moyamoya network is scanty and frequently unilateral. In most of the idiopathic cases, it is usually not possible to find any exogenous factors, such as mass lesion, meningitis, hemorrhage, or obstetrical trauma, as the underlying cause of the moyamoya syndrome. Pecker, et al., stressed a congenital origin for this condition, based upon the bilateral picture, which is more or less symmetrical, the frequent coexistence of other congenital disease, incidence of familial cases, and the fact that more cases are discovered in infancy or childhood. Kitahara, et al., found a relatively high incidence (7%) of moyamoya disease in their totally Japanese series, emphasizing the importance of the genetic factor in the pathogenesis of the disease. In the present case, after toxic and infectious conditions and obstetrical trauma were eliminated as causes, the history and investigations suggested that this was an authentic case of moyamoya syndrome. The

References


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