

## Patient Report

**Moyamoya disease-induced hemichorea corrected by indirect bypass surgery**YOUNG OK KIM,<sup>1</sup> TAE SUN KIM,<sup>2</sup> YOUNG JONG WOO,<sup>1</sup> CHAN JONG KIM<sup>1</sup> AND CHAN KYUN OH<sup>1</sup>*Departments of <sup>1</sup>Pediatrics and <sup>2</sup>Neurosurgery, Chonnam National University Medical School, Gwangju, Korea***Key words** chorea, hemiplegia, indirect bypass surgery, moyamoya disease.

Moyamoya disease is a chronic occlusive cerebral vasculopathy of unknown origin that is characterized by progressive steno-occlusion of the proximal intracranial portion of the internal carotid arteries and collateral vessels at the base of the brain. A common clinical finding in children with moyamoya disease is recurrent attacks of ischemic stroke, as opposed to the adult presentation of intracranial hemorrhage. Choreic movements in patients with moyamoya disease have been reported occasionally, and these movements are usually described as an initial manifestation of the disease.<sup>1–8</sup> Only Pavlakis *et al.* have described a 13-year-old boy with steroid-responsive, generalized chorea as a late manifestation following left hemiplegia at an interval of 1 year.<sup>2</sup> We report here a case of surgically corrected left hemichorea in an 8-year-old girl with moyamoya disease who presented with an initial manifestation of right hemiplegia at 1 year of age.

**Case report**

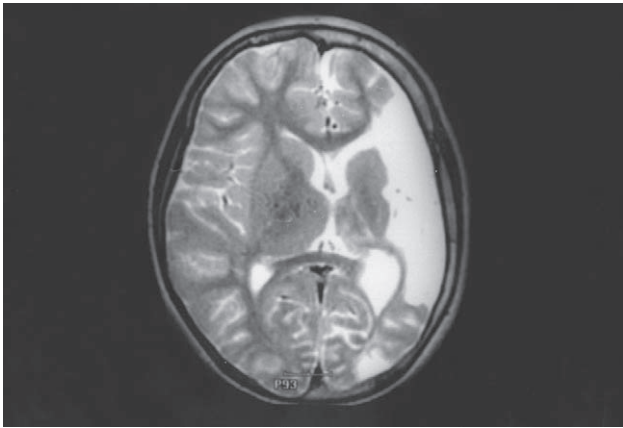
A 1-year-old girl admitted to hospital presented with right hemiplegia and hemisensory impairment following sudden-onset clonic movements of the right arm. The girl's history and the results of a general physical examination were unremarkable. Extensive laboratory evaluations included a complete blood cell count; measurements of the erythrocyte sedimentation rate, antinuclear antibody concentrations, and rheumatoid factor concentrations; a lipid profile; and a cerebrospinal fluid examination. All of these parameters were normal except for mild anemia. Brain magnetic resonance imaging

(MRI) showed high signal intensity within the left fronto-temporoparietal areas on T2-weighted imaging, which was suggestive of acute cerebral infarction. Magnetic resonance angiography (MRA) demonstrated occlusion of the left middle cerebral artery (MCA) and the development of collaterals. Brain [<sup>99m</sup>Tc]-hexamethylpropyleneamine oxime single-photon emission computed tomography (SPECT) revealed a diffuse perfusion defect within the left hemisphere. Cerebral angiography and bypass surgery were required but were not carried out because the parents refused permission. The girl had been treated for seizures with phenobarbital and had been free of seizures for several years. Motor power of the left extremities improved slowly, but a limping gait and small left extremities were observed over several years.

Seven years after the first admission of the patient (i.e., at 8 years of age), involuntary movements developed suddenly in the left extremities. Left hemichoreic movement gradually worsened and was intermittently accompanied by aphasia. Brain MRI revealed a previous infarct lesion with extensive encephalomalacia in the left MCA, but revealed no newly developed infarcted regions (Fig. 1). Magnetic resonance angiography and subsequent four-vessel cerebral angiography revealed severe stenosis and occlusion at the supraclinoid portion of either the right or left internal carotid artery with numerous collateral vessels in the vicinity of the occlusion, which are features that are consistent with moyamoya disease (Fig. 2). The [<sup>99m</sup>Tc]-ethyl cysteinate dimmer SPECT revealed a large perfusion defect within the left hemisphere owing to the previous infarction, but the left basal ganglia were spared (Fig. 3a). Acetazolamide administration demonstrated impaired vascular reserve within the right frontoparietal lobe (Fig. 3b). At 24 days after symptom onset, the patient underwent encephalo-duro-arterio-myosynangiosis on the right side, which ameliorated the chorea. The patient was treated perioperatively with haloperidol, and this treatment was reduced gradually over several months when it became apparent within 2 months

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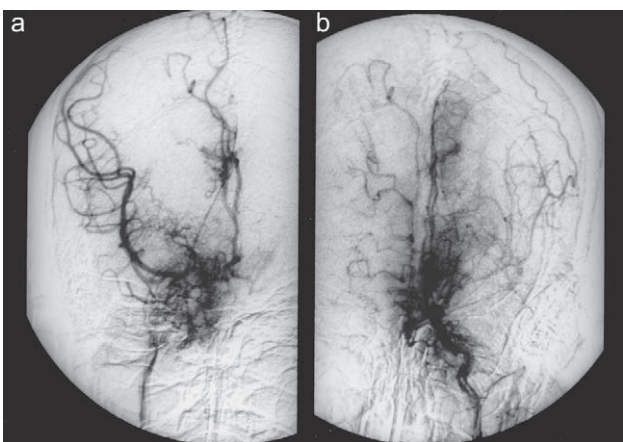


**Fig. 1** Brain magnetic resonance imaging showing a previous infarct lesion with extensive encephalomalacia in the left middle cerebral artery territory. No newly developed infarcted regions were observed.

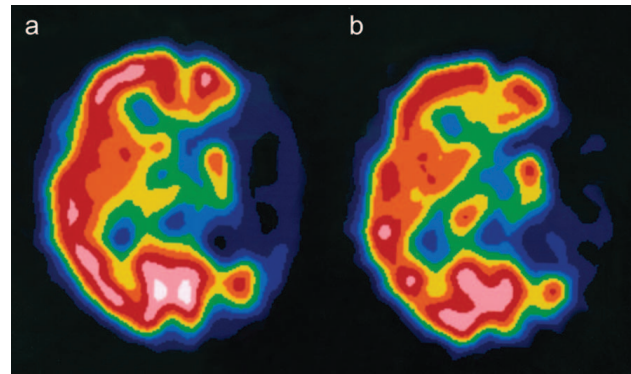
of the operation that she was free of hemichorea and aphasia. The patient has been free of chorea and aphasia for 40 months since the operation, which we believe is attributable to the indirect bypass surgery even though postoperative SPECT was not carried out.

## Discussion

Chorea is an uncommon movement disorder that presents as frequent, brief, sudden, twitch-like movements that flow from one part of the body to others in a chaotic manner.<sup>9</sup> The pathophysiology of chorea is not understood fully. But, in diseases with well-defined neuropathologic characteristics,



**Fig. 2** (a) Right internal carotid angiography demonstrated supraclinoid carotid narrowing with a net-like cluster of blood vessels in the vicinity of the stenotic area. (b) Left internal carotid angiography demonstrated occlusion of the carotid artery at the supraclinoid portion with leptomeningeal collaterals.



**Fig. 3** (a,b) Brain single-photon emission computed tomography revealed a large perfusion defect within the left hemisphere attributable to a previous infarction, but the left basal ganglia were spared. (b) There was impaired vascular reserve in the right frontoparietal lobe following acetazolamide administration.

chorea has been associated with abnormalities in the striatum or subthalamic nucleus.<sup>9</sup> Recently, a variety of different data raised the possibility that input from areas other than the basal ganglia is involved in chorea.<sup>9</sup> In most cases of hemichorea in one side of the body, there are contralateral hemispheric lesions in areas such as the caudate nucleus, putamen, globus pallidus, subthalamic nucleus, parts of thalamus, and subcortical white matter,<sup>9</sup> although there are also several exceptions.<sup>4-7</sup>

Chorea may be either inherited or acquired owing to hypoxia, toxins, infection, antibody-mediated diseases, and metabolic disorders. Even though moyamoya disease is a very rare cause of chorea, moyamoya disease must be considered when diagnosing children who present suddenly with involuntary movements. In contrast, Sydenham's chorea presents late in patients with rheumatic fever (which is the most common cause of acquired chorea), and this is observed rarely today. Choreic movements in patients with moyamoya disease are more prevalent in children than adults.<sup>1-8</sup> But, during childhood, the lesions in the nigrostriatal dopamine neuron and the basal ganglia may be asymptomatic because the striatal indirect pathways are functionally immature.<sup>10</sup> In the present study, choreic movements developed relatively late (7 years later after the initial manifestation), even though hemiplegia and hemiconvulsion developed early at 1 year of age.

The treatment of chorea depends on the pathophysiology of the disease. The most common agents used to treat chorea include antidopaminergic agents such as haloperidol and pimozide, benzodiazepines, and anticonvulsants such as phenytoin, carbamazepine, and valproic acid. In moyamoya-induced chorea, these medications cannot be the definitive treatment because moyamoya disease produces progressive occlusive vasculopathy. Therefore, we recommend bypass surgery, although it should be noted that in some cases improvement was shown without surgery.<sup>1-8</sup>

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