MoyaMoya disease is one of the main causes of childhood ischemic stroke and is characterized by spontaneous progressive stenoocclusive changes of the terminal ICA and fine collateral vessel development at the base of the brain. These conditions are also reported as secondary phenomena of vascular occlusion in several underlying diseases. Glycogen storage disease Type 1a is an autosomal recessive inborn error of carbohydrate metabolism caused by glucose-6-phosphatase deficiency, and the clinical manifestations include growth retardation, rounded doll-like face, hepatomegaly, hypoglycemia, and lactic acidosis. We report on a case of a child with MMD and GSD-1a with a focus on perioperative management.

Case Report

History and Examination. This 7-year-old Japanese girl was examined at her local hospital due to repeated transient ischemic attacks, right hemiparesis, and dysphasia. Magnetic resonance imaging revealed severe stenosis at the terminal portions of the bilateral internal carotid arteries, with typical moyamoya vessels. The patient underwent superficial temporal artery–middle cerebral artery anastomosis and encephalomiosynangiosis bilaterally, in 2 staged procedures at an interval of 4 months. Despite perioperative administration of glucose, hypoglycemia and metabolic acidosis occurred after both surgeries. The symptoms were milder after the second surgery, in which an increased dose of glucose was used. The patient tolerated the perioperative conditions well under intensified medical treatment, and no further ischemic symptoms occurred. (DOI: 10.3171/2010.10.PEDS10175)

Key Words • moyamoya disease • glycogen storage disease Type 1a • perioperative complication • lactic acidosis

Abbreviations used in this paper: CBF = cerebral blood flow; GSD-1a = glycogen storage disease Type 1a; ICA = internal carotid artery; MMD = moyamoya disease.
(263 mg/dl), triglycerides (1321 mg/dl), lactate (48.3 mg/dl), and uric acid (7.5 mg/dl). The patient’s fasting blood glucose level was 74 mg/dl. An MR imaging study was performed; the T2-weighted images showed right frontoparietal cortical atrophy and the previous left frontal subcortical infarction (Fig. 1A). Cerebral angiography demonstrated severe stenosis at the terminal portions of bilateral ICAs, with typical moyamoya vessels at the base of the brain (Fig. 1B and C). There was no obvious abnormality on the posterior circulation or extracranial vasculature. A decrease of CBF in the bilateral frontoparietal region was revealed on SPECT images.

**Surgical Treatment and Outcome.** Direct revascularization with superficial temporal artery–middle cerebral artery anastomosis and indirect revascularization with encephalomyosynangiosis were performed to prevent further ischemic injury. Bilateral surgery was performed in 2 stages (initially on the left side) with an interval of 4 months. Postoperative angiography revealed a patent bypass; adequate collateral vessels were seen bilaterally (Fig. 2). Postoperative SPECT images showed marked improvement of CBF in regions that preoperatively demonstrated impaired perfusion.

**Perioperative Course.** Despite continuous pre-, intra-, and postoperative intravenous administration of glucose, hypoglycemia and metabolic acidosis occurred after the first operation. The patient’s blood glucose level was 73 mg/dl, the lactate level was 13.0 mmol/L, and the base excess value was −9.1 mmol/L at 3 hours postoperatively; compensatory hyperventilation gradually developed with a respiration rate of 36 breaths per minute and arterial carbon dioxide pressure of 25.6 mm Hg. Hyperventilation ceased following an increased dose of glucose with intravenous infusion of sodium bicarbonate. The patient’s respiration rate was 24 breaths per minute and PaCO₂ was 35.5 mm Hg at 9 hours postoperatively (Fig. 3). After the second operation, lactic acidosis and compensatory hyperventilation were milder than the first postoperative period because a larger dose of glucose and sodium bicarbonate were administered during the operation and postoperative period. The minimum blood glucose and maximum lactate levels were 82 mg/dl and 10.0 mmol/L, respectively; the minimum base excess was −4.5 mmol/L; and the minimum PaCO₂ was 30.0 mm Hg. The patient tolerated each postoperative period well; no further ischemic neurological symptom developed.

**Discussion**

Moyamoya disease is a cerebrovascular condition that results in the narrowing of vessels at the circle of Willis and collateral vessel formation at the base of the brain. Although these vascular conditions have been reported in association with various diseases, only 2 cases concurrent with GSD-1a have been reported in the literature. The pathogenesis of MMD in patients with GSD-1a has not yet been clarified. In an autopsy case of a patient with...
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GSD-1a and concurrent pulmonary hypertension, Pizzo\(^9\) described pathological changes suggestive of a chronic vasoconstrictive process with intimal fibrosis of the pulmonary vasculature. Similar histopathological findings of intra- and extracranial vessels had been observed in patients with MMD,\(^3\) thus GSD-1a may be a possible cause of MMD. Further studies involving more patients with concurrent MMD and GSD-1a are necessary to elucidate the relationship between these 2 conditions.

The inherent deficiency of glucose-6-phosphatase activity in patients with GSD-1a leads to the inhibition of free glucose release from liver glycogen into the bloodstream, resulting in hypoglycemia. Production of lactate increases, given the normal glycolysis of glucose-6-phosphate. Therefore, patients with GSD-1a tend to exhibit lactic acidosis and hyperventilate to compensate for metabolic acidosis. Lactic acidosis becomes more severe when the glucose supply is exhausted.\(^7\) On the other hand, cerebral cortical vessels in patients with MMD may dilate in response to decreased perfusion pressure caused by bilateral carotid artery stenocclusive lesions.\(^6,8\) Therefore, cerebrovascular ischemic symptoms of MMD are usually precipitated by hyperventilation via decreased PaCO\(_2\). Because the most serious perioperative ischemic complications related to cerebral bypass surgery for MMD are more often caused by the occurrence of perioperative hypo- and hypercapnia than the surgical procedure itself,\(^4,5\) meticulous perioperative management to prevent potentially hazardous lactic acidosis is mandatory.

Goutières et al.\(^2\) reported a case of a surgically treated patient with MMD associated with GSD-1a. The patient underwent right encephaloduroarteriomyosynangiosis (EDAMS) and developed complete left hemiplegia at 36 hours postoperatively, but specific causes of ischemic complications were not mentioned. An important lesson learned from the present case is that successful perioperative management after bypass surgery in patients with MMD and GSD-1a depends on meticulous prevention of hypoglycemia and lactic acidosis by sufficient administration of glucose and sodium bicarbonate. Another case, reported by Sunder,\(^11\) involved a 17-year-old boy, who was treated without surgery and suffered episodic headache and transient ischemic attacks. Our case is distinct from previously reported cases in that revascularization surgery under intensified perioperative medical treatment effectively improved CBF and prevented ischemic stroke.

Conclusions

We report on a patient with concurrent MMD and GSD-1a who was managed successfully during her perioperative period. Although surgical risk for MMD may be higher in patients with GSD-1a, surgical revascularization under intensified perioperative management should be considered to prevent future ischemic events.

Disclosure

The authors report no conflict of interest concerning the mate-
Author contributions to the study and manuscript preparation include the following. Conception and design: Egashira. Reviewed final version of the manuscript and approved it for submission: Miyamoto. Administrative/technical/material support: Ohnishi, Kawasaki, Higashigawa. Study supervision: Takahashi, Iihara.

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