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Moyamoya Case with Stenting for Basilar Stenosis

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What is known on this subject?

Surgical or endovascular interventions can be used in Moyamoya patients with recurrent progressive ischemic events and decreased cerebral perfusion.

What this study adds?

Endovascular treatment, as in Moyamoya disease, is a life-saving treatment option that is successfully applied in many diseases that cause severe cerebrovascular stenosis.

ABSTRACT

Moyamoya disease is a chronic, progressive hereditary disease characterized by narrowing of the vascular lumen because of hypertrophy of smooth muscles in the walls of the arteries that form the circle of Willis. Cerebral vessels may be encountered in bleeding and occlusion clinics. Although it is seen as predominant in Asian races, cases have been reported worldwide. Although its etiopathogenesis is not clear, genetics, some infectious agents, and autoimmune mechanisms are blamed. The gold standard in diagnosis is digital subtraction angiography. Surgical or endovascular interventions can be used in patients with recurrent progressive ischemic events and decreased cerebral perfusion. Here we present a 43-year-old Moyamoya patient who presented with posterior system findings and had a stent implantation with critical stenosis in the basilar artery.

**Keywords:** Moyamoya, stroke, basilar artery, stenosis, stent

Introduction

Moyamoya disease (MMD) is a progressive cerebrovascular disease characterized by narrowing of the lumen because of hypertrophy of smooth muscles in the vascular wall in the terminal part of the intracranial arteries (1). An increase in the collateral circulation in the leptomeningeal vessels is observed due to a decrease in brain perfusion. Because this vascular structure was likened to cigaret smoke by Suzuki and Takaku (2), who first described the disease in 1969 in cerebral angiographies, the disease was named "Moyamoya", which means cigaret smoke in Japanese. In Japan, where the number of cases is the highest, the annual prevalence is 3 in 100,000. The disease can be seen in children and adults and shows a bimodal course. It is two times more common in women than in men. It may present with intracerebral hemorrhage and stroke. Digital subtraction angiography (DSA) is the gold standard for diagnosis. Providing revascularization is important in treatment (3,4). Here, the clinical and radiological findings of the patient who
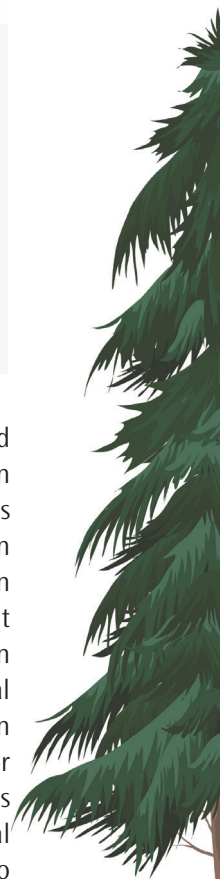
presented with severe cognitive impairment and posterior system stroke findings were discussed.

Case Report

A 43-year-old female patient presented with complaints of diplopia and impaired speech. In her family history, it was learned that her mother died at the age of 30 due to intracranial hemorrhage. In her neurological examination, she was consciously cooperatively oriented, speech dysarthric, with inner gaze limited in the right eye, and horizontal nystagmus in the left eye was observed in the left eye. She had no motor or sensory impairment. In the laboratory examination, low-density lipoprotein was 160 mg/dL (0-100 mg/dL), and no significant pathology was observed in other blood tests. Computed tomography of the brain was normal. In diffusion magnetic resonance imaging (MRI), acute infarction was seen in the left half of the pons (Figure 1a, b). Her electrocardiography was in normal sinus rhythm, and her arrival blood pressure was measured as 150/100 mmHg. His treatment was arranged as acetyl salicylic acid (ASA) 100 mg/day + clopidogrel 75 mg + enoxaparin sodium 0.6 mL 2x1 subcutaneously. In the transthoracic electrocardiogram, the ejection fraction was 60% and no intracardiac mass/thrombus was detected. The thrombophilia panel and vasculitis markers (such as protein C, protein S, antithrombin 3, factor 5 Leiden mutation, prothrombin 2 gene mutation, antinuclear antibody, complement levels, and other vasculitis markers) were normal. In cranial and cervical MR angiography, no flow was observed in both internal carotid arteries (ICA), and

approximately 50% stenosis was observed in the distal of the basilar artery (Figure 2). In DSA, it was found that bilateral ICAs were occluded, and critical stenosis of up to 80% was in the distal part of the basilar artery. It was observed that cerebral circulation was provided through the vertebrobasilar system and leptomeningeal vessels (Figure 3a, b, c). On the 3rd and 7th days of the patient's hospitalization, diffusion MRIs taken due to clinical progression revealed new acute infarct areas in the right half of the pons, right temporal lobe, and centrum semiovale level. In his new neurological examination, consciousness was confused, and taking a single command, she had left hemiparesis (3/5). After the patient's clinical progression, it was decided to stent the basilar artery. The pre-procedure treatment was adjusted as ASA 100 mg/day + ticagrelor 120 mg/day. Under general anesthesia, first angioplasty and then a stent of the appropriate diameter were placed in the spleen in the distal part of the basilar artery (Figure 4). The mini-mental test score performed on the 5th day after the stent was found to be 27/30. Clinically, the patient who had no deficits other than -5/5 in left muscle strength was admitted to the neurology outpatient clinic under ASA 100 mg/day + clopidogrel 75 mg/day and was discharged. Of note, informed consent was signed by the patient for this report.

Discussion

Although the etiopathogenesis of MMD remains uncertain, MMD has been observed in the family in 7-12% of the cases, suggesting that autosomal dominant inheritance plays a
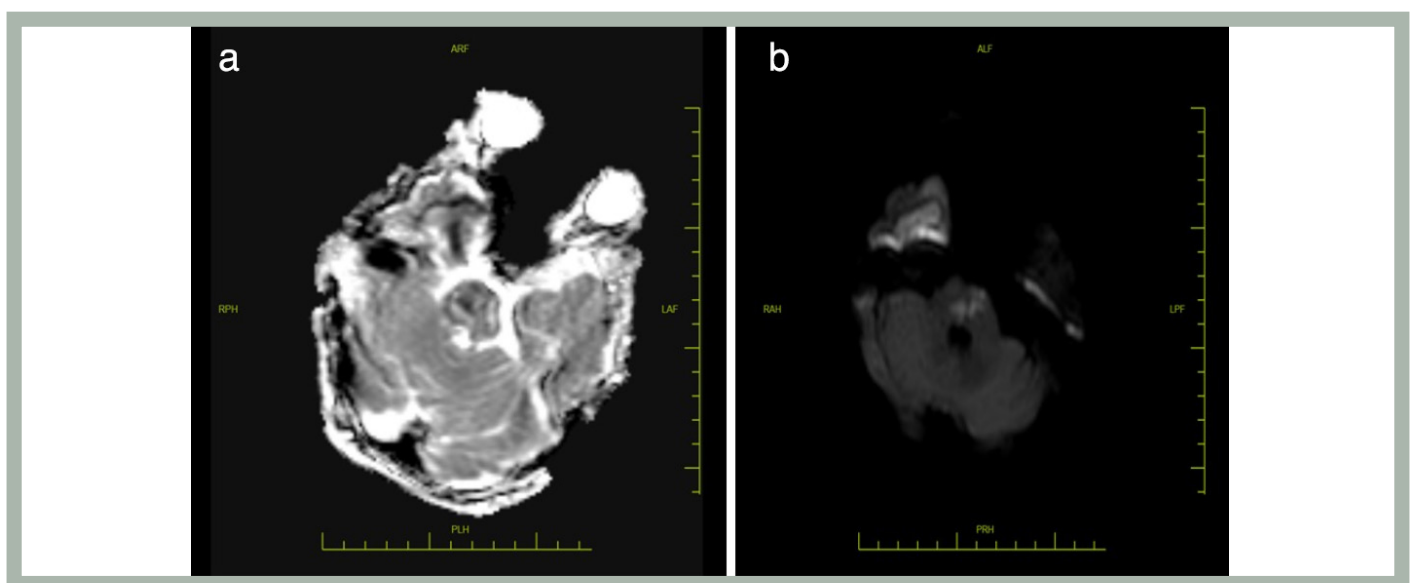

Figure 1. (a, b) ADC-DWI

ADC: Apparent diffusion coefficient, DWI: Diffusion weighted imaging

role in the transmission of the disease (5,6). Studies suggest that the disease may be associated with the *HLA DQB1, B51* genes and 3, 6, 8 and 17 chromosomes (7,8). Considering that the mother of our patient died at the age of 30 due to sudden intracerebral hemorrhage, we had cranial and cervical angiomas in two children of our patient, considering the risk of possible MMD genetic transmission. We found no significant pathology in imaging. We did not perform genetic analysis either. In addition, studies have shown that infectious agents and autoimmunity are effective in pathogenesis (9,10). The presence of hypothyroidism in the history of our patient

Figure 2. MRA

MRA: Magnetic resonance angiography

may suggest this possibility. Studies have shown fibrocellular initial thickening in the affected vessels, proliferation in smooth muscle cells, increased elastin accumulation, and dilated, thin-walled, non-muscular collateral vessels in the subarachnoid space (11). Microaneurysms can often occur as a result of weakening of the vascular media layer. Approximately 25% of individuals diagnosed with MMD may have stenosis in the proximal part of the posterior cerebral arteries (12). Our patient had severe stenosis in the distal part of the basilar artery. MMD may present with migraine-like, medical treatment-resistant, recurrent headaches. Our patient had long-lasting migraine-like headaches. The clinic may differ in children and adults. Ischemia is more prominent in children and bleeding in adults (13,14). It manifests mostly as transient ischemic attacks in children. Hemorrhages may occur in intracerebral, intraventricular, subarachnoid, and subdural areas because of rupture of small microaneurysms and dilated vessels in the posterior circulation (14,15).

In MMD, the specificity of MRI has been reported as 100% and its sensitivity as 73%. When MRI-angiography is included, the sensitivity reaches 92% (16). On MRI, it is observed that the flow areas decrease in the terminal part of the middle cerebral artery, and the flow areas increase due to perforating vessels in the basal ganglion localization (17). The gold standard in diagnosis is DSA. With DSA, stenosis in the distal of the cerebral arteries, increased collateral network ofleptomeningalleptomeningeal arteries, and vascularization near the basal ganglia are observed. In our case, while no flow was observed in both ICAs, it was observed that the circulation of the anterior system was provided from the relative system
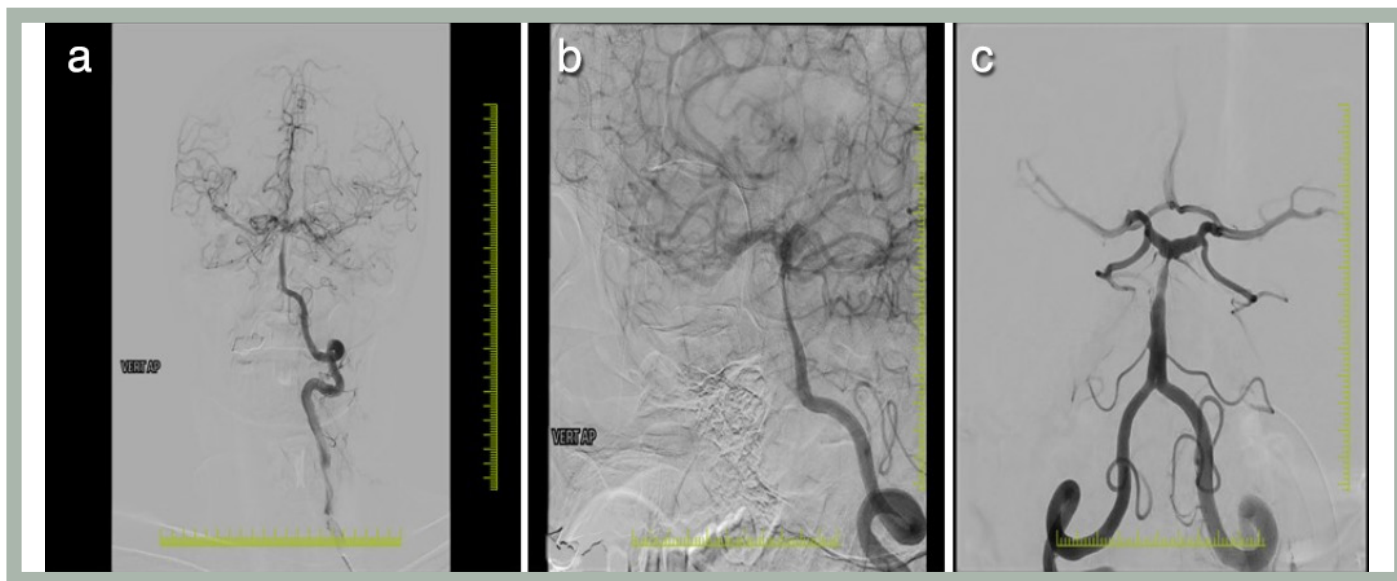

Figure 3. (a, b, c) DSA-before the procedure

DSA: Digital subtraction angiography

Figure 4. DSA-after the procedure

DSA: Digital subtraction angiography

through the Willis polygon and that the superior basilar artery was severely narrowed distally.

There is no definitive treatment for MMD, and the purpose of treatment is to minimize the ischemic process and reduce the risk of bleeding. Steroids, anticoagulants, antiaggregants, and vasodilators can be used in medical treatment, but their efficacy is controversial (3). Our patient continued to have ischemic attacks under dual antiaggregant and anticoagulant therapy. Some studies have shown the positive effects of the calcium channel blockers used in resistant headaches and the reduction in the frequency and severity of transient ischemic attacks (13). Endovascular and surgical treatment options are also used in addition to medical treatment. Few reports have been published detailing endovascular approaches to MMD (18). The stent-assisted angioplasty procedure, which is advantageous due to its less invasiveness, is technically suitable and safe for the treatment of selected patients with intracranial atherosclerotic stenosis. Endovascular embolization of intracranial aneurysms accompanying MMD is also possible. In our patient, time was gained by delaying the development of new collateral network formation in the anterior circulation with the basilar artery stenting strategy.

Surgical interventions to increase cerebral blood flow are used in patients with recurrent progressive ischemic events and reduced cerebral circulation network. Surgical approaches can be applied under direct, indirect, or combined techniques. In the direct surgical approach, bypass of the superficial temporal or middle meningeal artery with the distal parts of the middle cerebral artery or anterior cerebral artery and anastomoses of the occipital artery with the posterior cerebral artery are options (19,20). Dura, temporal muscle, and galea can be used in indirect surgical techniques; thus, it is aimed to induce spontaneous angiogenesis with peduncle tissue (21). The incidence of stroke in the first five years after surgery is almost below 5%. Recurrent episodes of stroke and early onset of the disease are poor prognostic factors. Morbidity is more than 70% in untreated patients (16,22). Progressive course can be seen in patients whose early diagnosis is delayed and appropriate treatment cannot be provided, and due to this situation, irreversible cortical atrophy and mental retardation can be observed in patients. Our patient had recurrent ischemic attacks, cognitive loss, and impaired consciousness. Increasing brain perfusion was achieved by stenting the basilar artery with endovascular treatment. After this treatment, it was observed that the neurological deficit was almost completely resolved and the cognitive loss completely regressed.

As a result, endovascular treatment, as in MMD, is a life-saving treatment option that is successfully applied in many diseases that cause severe cerebrovascular stenosis.

Ethics

Informed Consent: Informed consent was signed by the patient for this report.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Ö.K., U.D., T.G., M.C., Concept: Ö.K., U.D., T.G., M.C., Design: M.Ç., Data Collection or Processing: K.T., A.Ö., Literature Search: K.T., A.Ö., M.Ç., Writing: K.T., A.Ö., M.Ç.

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REFERENCES

1. Huang S, Guo ZN, Shi M, Yang Y, Rao M. Etiology and pathogenesis of Moyamoya disease: an update on disease prevalence. Int J Stroke 2017;12:246-253.

2. Suzuki J, Takaku A. Cerebrovascular "Moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288-299.

3. Gosalakkal JA. Moyamoya disease: a review. Neurol India 2002;50:6-10.

4. Goda M, Isono M, Ishii K, Kamida T, Abe T, Kobayashi H. Long-term effects of indirect bypass surgery on collateral vessel formation in pediatric moyamoya disease. J Neurosurg 2004;100:156-162.

5. Miyatake S, Miyake N, Touho H, et al. Homozygous c.14576G>A variant of RNF213 predicts early-onset and severe form of moyamoya disease. Neurology 2012;78:803-810.

6. Miyawaki S, Imai H, Shimizu M, et al. Genetic variant RNF213 c.14576G>A in various phenotypes of intracranial major artery stenosis/occlusion. Stroke 2013;44:2894-2897.

7. Ikeda H, Sasaki T, Yoshimoto T, Fukui M, Arinami T. Mapping of a familial moyamoya disease gene to chromosome 3p24.2-p26. Am J Hum Genet 1999;64:533-537.

8. Yamauchi T, Tada M, Houkin K, et al. Linkage of familial moyamoya disease (spontaneous occlusion of the circle of Willis) to chromosome 17q25. Stroke 2000;31:930-935.

9. Tanigawara T, Yamada H, Sakai N, Andoh T, Deguchi K, Iwamura M. Studies on cytomegalovirus and Epstein-Barr virus infection in Moyamoya disease. Clin Neurol Neurosurg 1997;99:S225-S228.

10. Horn P, Pfister S, Bueltmann E, Vajkoczy P, Schmiedek P. Moyamoya-like vasculopathy (moyamoya syndrome) in children. Childs Nerv Syst 2004;20:382-391.

11. Kuroda S, Houkin K. Moyamoya disease: current concepts and future perspectives. Lancet Neurol 2008;7:1056-1066.

12. Matsushima Y. Moyamoya Disease. Principles and practice of pediatric nerosurgery. New York: Thieme 1999:1053-1070.

13. Houkin K, Kamiyama H, Abe H, Takahashi A, Kuroda S. Surgical therapy for adult moyamoya disease. Can surgical revascularization prevent the recurrence of intracerebral hemorrhage? Stroke 1996;27:1342-1346.

14. Nijdam JR, Luijten JA, van Gijn J. Cerebral haemorrhage associated with unilateral Moyamoya syndrome. Clin Neurol Neurosurg 1986;88:49-51.

15. Scott RM, Smith ER. Moyamoya disease and Moyamoya syndrome. N Engl J Med 2009;360:1226-1237.

16. Pereira PL, Farnsworth CT, Duda SH, Rose M, Reinbold WD, Claussen CD. Pediatric moyamoya syndrome: follow-up study with MR angiography. AJR Am J Roentgenol 1996;167:526-528.

17. Fujiwara H, Momoshima S, Kuribayashi S. Leptomeningeal high signal intensity (ivy sign) on fluid-attenuated inversion-recovery (FLAIR) MR images in moyamoya disease. Eur J Radiol 2005;55:224-230.

18. Khan N, Dodd R, Marks MP, Bell-Stephens T, Vavao J, Steinberg GK. Failure of primary percutaneous angioplasty and stenting in the prevention of ischemia in Moyamoya angiopathy. Cerebrovasc Dis 2011;31:147-153.

19. Reis CV, Safavi-Abbasi S, Zabramski JM, Gusmão SN, Spetzler RF, Preul MC. The history of neurosurgical procedures for moyamoya disease. Neurosurg Focus 2006;20:E7.

20. Veeravagu A, Guzman R, Patil CG, Hou LC, Lee M, Steinberg GK. Moyamoya disease in pediatric patients: outcomes of neurosurgical interventions. Neurosurg Focus 2008;24:E16.

21. Soriano SG, Cowan DB, Proctor MR, Scott RM. Levels of soluble adhesion molecules are elevated in the cerebrospinal fluid of children with moyamoya syndrome. Neurosurgery 2002;50:544-549.

22. Eicker S, Etminan N, Turowski B, Steiger HJ, Hänggi D. Intracranial carotid artery stent placement causes delayed severe intracranial hemorrhage in a patient with moyamoya disease. J Neurointerv Surg 2011;3:160-162.