*Case Report*

# Efficacy of Extracranial–Intracranial Bypass for Progressive Middle Cerebral Artery Occlusion Associated with Active Sjögren's Syndrome: Case Report

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Sjögren syndrome affecting the major cerebral arteries is rare, and an optimal therapeutic strategy to counteract such a lesion has not yet been established. We herein report a case of a 39-year-old woman with a history of primary Sjögren syndrome, which had previously been treated with immunosuppressive therapy, manifesting with a crescendo transient ischemic attack because of left middle cerebral artery stenosis. Despite the administration of high doses of prednisolone and azathioprine for active Sjögren syndrome, the frequency of crescendo transient ischemic attacks increased with the progression of stenosis and magnetic resonance imaging showed the development of subacute cerebral infarction. Single-photon emission computed tomography with N-isopropyl[$^{123}$I]-p-iodoamphetamine revealed apparent hemodynamic compromise in the affected cerebral hemisphere. In light of the increased risk of further progression of cerebral infarction, we decided to perform surgical revascularization in spite of her active inflammatory condition. The patient underwent extracranial–intracranial bypass without complications and was treated with intensive immunosuppressive therapy during the perioperative period. Based on our findings, we recommend surgical revascularization for occlusive cerebrovascular disease with hemodynamic compromise in combination with intensive immunosuppressive therapy, even in the active inflammatory state of autoimmune diseases, if ischemic symptoms are medically uncontrollable. **Key Words:** Sjögren's syndrome—cerebral artery occlusion—cerebral infarction—immunosuppressive therapy.


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Primary Sjögren syndrome is a chronic autoimmune disorder of the exocrine glands with associated lymphocytic infiltrates in the affected glands. Neurologic manifestations, including the involvement of both the peripheral and the central nervous systems, have been reported in approximately 20% of patients with Sjögren syndrome.[1] Although a recent study demonstrated the association between Sjögren syndrome and the high risk of ischemic and hemorrhagic stroke because of small vessel

vasculitis,[2] only a few cases in which the major cerebral arteries were affected have been reported.[3-5] Therefore, an optimal therapeutic strategy to treat this rare condition has not yet been established, especially during the active stage of Sjögren syndrome.

We herein described a patient with Sjögren syndrome who manifested with a crescendo transient ischemic attack (TIA) caused by the progressive occlusion of the middle cerebral artery (MCA), and was successfully managed by revascularization surgery under the intensive immunosuppressive therapy during the active stage of Sjögren syndrome.

## Case Report

A 39-year-old woman had a 10-year history of primary Sjögren syndrome, which was diagnosed with minor salivary gland biopsy (grade 4), Schirmer test, and positive Sjögren syndrome antigen A and antinuclear antibody. She had previously been treated with prednisolone and azathioprine to control the activity of this syndrome by the department of rheumatology. Eight months before being admitted to our department, magnetic resonance angiography showed the manifestation of asymptomatic bilateral M1 stenosis (Fig 1, A). She developed TIA with right hemiparesis and speech disturbance 2 months later. Because the frequency of TIA increased in spite of intensive immunosuppressive therapy (40 mg prednisolone and 100 mg azathioprine) over 6 months, she was referred to our department for surgical consideration. A neurologic examination on admission revealed mild motor aphasia. A serologic examination was positive for Sjögren syndrome antigen A and antinuclear antibody and showed higher levels of C-reactive protein (5.1 mg/dL), whereas antiphospholipid antibody was negative. These findings suggested that Sjögren syndrome was in the

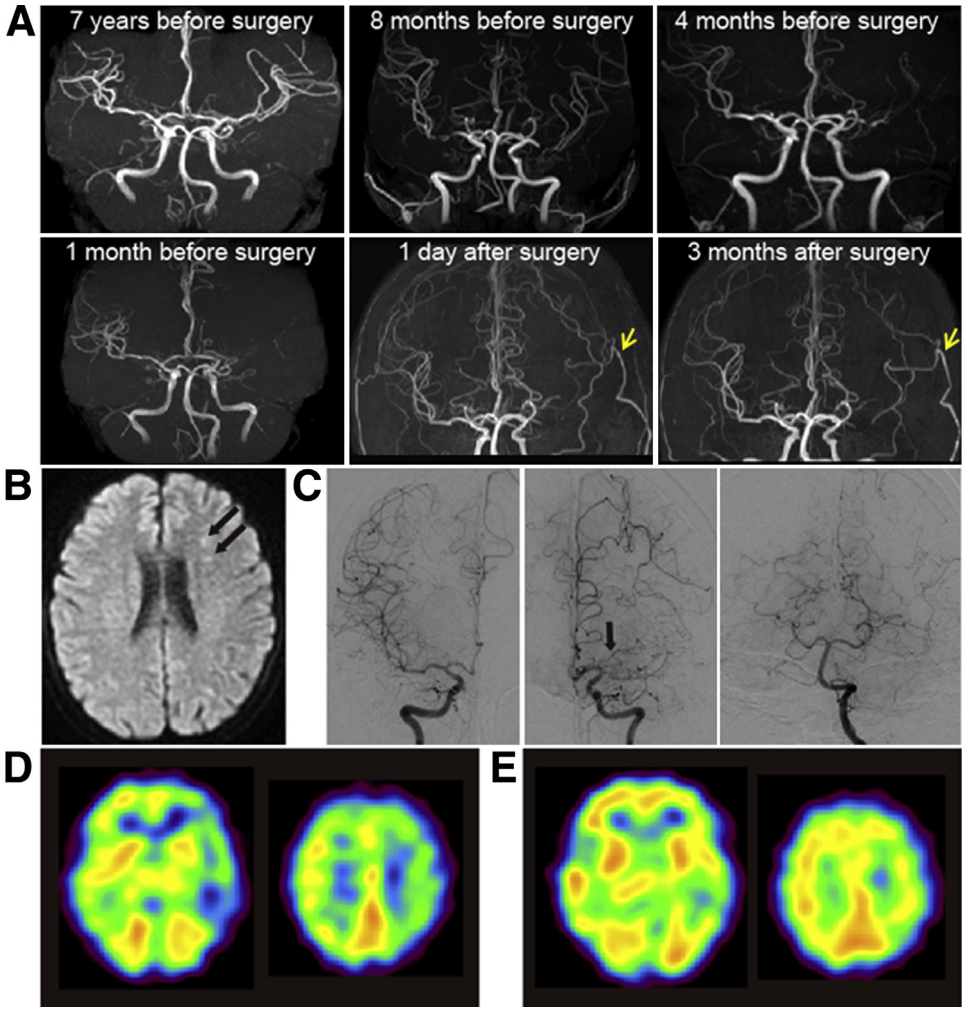


**Figure 1.** *(A) Temporal profile of magnetic resonance angiography showing bilateral middle cerebral artery (MCA) stenosis 8 months before surgery, progression of left MCA stenosis and improvements in right MCA stenosis 4 months before surgery, occlusion of the left MCA 1 month before surgery, and patency of superficial temporal artery–MCA bypass 1 day and 3 months after surgery (arrows). (B) Diffusion-weighted image 1 month before surgery revealing multiple subacute infarction in the left cerebral hemisphere (arrows). (C) Digital subtraction angiography, performed 1 month before surgery, demonstrating occlusion of the left MCA (arrow). Single-photon emission computed tomography with N-isopropyl[$^{123}$I]-p-iodoamphetamine, performed 1 month before surgery (D) and 7 days after surgery (E), showing that the decrease in cerebral blood flow in the left hemisphere significantly improved after revascularization surgery.*