Acute Subdural Hematoma in Young Patient with Moyamoya Disease
—Case Report—

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Abstract

A 17-year-old boy with known moyamoya disease developed an acute subdural hematoma after a mild head trauma. He had been confined to a wheelchair with contracture in the upper and lower extremities due to juvenile rheumatoid arthritis since age 1 year. He had undergone encephalo-duro-arterio-synangiosis (EDAS) on the right and encephalo-myo-synangiosis (EMS) on the left at 13 years of age. He was admitted with headache, nausea, and vomiting after a fall from his wheelchair at age 17. Computed tomography on admission showed a large acute subdural hematoma in the right frontotemporal region but no bleeding at the EDAS or EMS sites. Cerebral angiography 12 weeks after the head trauma revealed a remarkable reduction in the spontaneous transdural external-internal carotid anastomoses in the right frontal region. The acute subdural hematoma was probably caused by rupture of the spontaneous transdural anastomoses.

Key words: moyamoya disease, subdural hematoma, head injury, encephalo-duro-arterio-synangiosis, encephalo-myo-synangiosis, anastomosis

Introduction

Moyamoya disease is characterized angiographically by progressive occlusion of cerebral arteries, abnormal vascular networks in the basal ganglia, leptomeningeal anastomoses, and multiple transdural external-internal carotid anastomoses. Moyamoya disease usually manifests clinically as ischemic episodes in children and intracranial hemorrhage in adults. Modern computed tomographic (CT) scanning clearly shows that most intracranial hemorrhages are intracerebral or intraventricular. Despite the presence of multiple transdural arterial anastomoses, associated acute or subacute subdural hematoma rarely occurs. Here, we describe a young adult with an acute subdural hematoma due to the rupture of spontaneous transdural external-internal carotid anastomoses after a mild head trauma.

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Case Report

This 17-year-old boy had suffered from a juvenile type of rheumatoid arthritis in the upper and lower extremities since 1 year old, requiring a wheelchair because of gradual contracture in the hip, knee, and ankle joints. On August 26, 1978 (at 9 years old), he suddenly developed clonic convolution of the left upper extremity for over 15 minutes, followed by hemiparesis on the left, dysarthria, alexia, and dyscalculia. He was slowly recovering from these symptoms when he presented at the Department of Neurosurgery, Brain Research Institute, Niigata University on July 12, 1982. Neurological examination revealed mental deterioration, weakness of the left upper extremity, and contracture of the upper and lower extremities. A CT scan showed a large low-density lesion in the right frontal lobe. Cerebral angiograms demonstrated typical findings of moyamoya disease, including moderate transdural external-internal carotid anastomoses in the right frontal lobe.
On January 5, 1983 (at 13 years old), he was admitted to Niigata University Hospital for surgical management of the moyamoya disease. CT scans showed low-density lesions in the bilateral frontal lobes (Fig. 1). Encephalo-duro-arterio-synangiosis (EDAS) was performed on the right on January 14, and encephalo-myo-synangiosis (EMS) on the left on January 27. EMS was the surgical treatment of choice for child patients with moyamoya disease at that time, but the right EDAS was performed to avoid injuring spontaneous transdural arterial anastomoses. General convulsion, transient pseudobulbar paresis, and transient weakness of the right upper extremity occurred a few days postoperatively. He became independent gradually, and attended a disabled children's school. Cerebral angiograms on August 5, 1983 showed slight revascularization of the brain via the right EDAS and marked revascularization via the left EMS (Fig. 2). He received 1500 mg acetylsalicylic acid daily to treat the rheumatoid arthritis pre- and postoperatively. He continued to be relatively well without convulsion or ischemic attack for 4 years.

On February 3, 1987 (at 17 years old), he fell from a wheelchair and hit the left temporal region on the floor without loss of consciousness. He complained of only slight left temporalgia. He fell again and hit the right frontotemporal region on the floor without loss of consciousness on February 5, 1987. He was admitted to the Department of Neurosurgery, Kuwana Hospital with complaints of headache, nausea, and vomiting on the same day. Neurological examination revealed mental deterioration, contracture, and deformity in the upper and lower extremities which were present before the head trauma, but no new deficits. Skull x-ray films showed no fracture except the operative craniotomy. CT scans demonstrated a large acute subdural hematoma in the right frontotemporal region and a small cerebral contusion in the right temporal region with a slight mass effect (Fig. 3). There was no bleeding or cerebral contusion at the right EDAS or the left EMS sites. He was treated conservatively with symptomatic improvement, and was discharged on March 12, 1987. Cerebral angiograms on April 27, 1987 showed that revascularization via the operative sites was unchanged, but the spontaneous transdural arterial anastomoses in the right frontal region were remarkably reduced compared with the angiograms on August 5, 1983 (Fig. 4).
Children with moyamoya disease usually develop spontaneous transdural external-internal carotid anastomoses. Acute or subacute subdural hematoma has occurred in a few adults with this disease. However, only one child patient with acute subdural hematoma has been reported. Taveras reported an adult with a subdural hematoma following ventriculography with considerable subdural air. He suspected that separation of the brain caused by the subdural air resulted in rupture of the transdural arterial channels. Mori et al. described an adult case of moyamoya syndrome with acute subdural and intracerebral hematomas from different bleeding sources. They thought that distortion of the brain by the intracerebral hematoma caused rupture of an aneurysmal dilatation in a transdural anastomosis. These authors emphasized the importance of shearing of enlarged transdural arterial channels as a significant source for subdural bleeding. In our case, there was a low-density lesion in the right frontal lobe, where the transdural anastomoses were located. Probably, the transdural anastomoses at the old cerebral infarction site were easily injured by a shearing force even in mild head trauma because of the brain atrophy.

Nakagawa et al. and Sonobe et al. reported juvenile cases of chronic subdural hematoma developing after EMS for moyamoya disease. They pointed out that postoperative administration of acetylsalicylic acid was a factor in the development of the chronic subdural hematoma. In our case, acetylsalicylic acid was administered for rheumatoid arthritis. The maximum rate of secondary platelet aggregation with addition of 10 μM adenosine diphosphate and 2 μg collagen/ml of plasma was 61 and 23%, respectively, using platelet-rich plasma with 300 × 10^4 platelets on admission to Kuwana Hospital. Although other coagulation tests were normal on admission, this reduction in platelet aggregation may have been a factor enlarging the acute subdural hematoma after the head trauma.

Sayama et al. reported the autopsy of a 16-year-old boy with moyamoya disease who received an external carotid-internal carotid bypass and EMS, and died of a severe cerebral contusion and an acute subdural hematoma caused by a traffic accident. The traumatic damage and hemorrhage were most drastic in the area corresponding to the EMS. In our case, there was no bleeding or cerebral contusion at the
EMS and EDAS sites. Impact is unlikely to damage indirect revascularization in moyamoya disease such as EMS and EDAS easily, because the muscle and the galea spontaneously attach to the brain surface postoperatively. The spontaneous transdural external-internal carotid anastomoses are more likely to be injured.

References


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