Case Report / Olgu Sunumu

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Moyamoya Disease, Which is Rare in Infancy: A Case Report

Bebeklik Döneminde Nadir Görülen Moyamoya Hastalığı: Olgu Sunumu

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Abstract

Moyamoya disease etiology is an undetermined vasculopathy and is mainly thought to affect the Internal Carotid Artery and Wills circle. An 8-month-old patient without any underlying disease was referred to our hospital with sudden extreme right hemiparesis. There was no evidence of meningeal irritation in the neurological examination, decreased tonus on the right side, strength in the upper right extremity 3/5 and strength in the lower right extremity 2/5, deep tendon reflexes were exaggerated. Cranial brain tomography angiography showed in both distal internal cerebral arteries and branches and a significantly curvy appearance, with similar changes present in the veins forming the Willis circle. Low molecular weight heparin, acetylsalicylic acid, and levetiracetam were started. On the fifth day of follow-up, motor activity in the lower right extremity returned to normal. The power loss of 2/5 in the upper right extremity was continuing. The patient was then transferred to a center with Moyamoya surgery, which may be needed. This case highlights the importance of considering Moyamoya disease as a classical etiologies of acute ischemic strokes in children. It also highlights the rare presentation among the Turkish population and the use of neurovascular imaging techniques to facilitate the diagnosis of Moyamoya diseas

Keywords: Infant, Moyamoya disease, cerebral angiography, acute stroke

Öz

Moyamoya hastalığı etiyolojisi belirlenmemiş bir vaskülopatidir ve esas olarak internal Karotis arteri ve Wills çemberini etkilediği düşünülmektedir. Herhangi bir şikayeti olmayan 8 aylık hasta ani aşırı sağ hemiparezi ile hastanemize sevk edildi. Nörolojik muayenesinde meningeal irritasyon bulgusu yoktu, sağda tonus azalması, sağ üst ekstremitede kuvvet 3/5 ve sağ alt ekstremitede kuvvet 2/5, derin tendon refleksleri abartılı idi. Kraniyal beyin tomografisi anjiyografisi hem distal iç serebral arterlerde ve dallarda hem de Willis çemberini oluşturan damarlarda benzer değişikliklerle birlikte önemli ölçüde kıvrımlı bir görünüm gösterdi. Düşük moleküler ağırlıklı heparin, asetilsalisilik asit ve levetirasetam başlandı. Takibinin beşinci gününde sağ alt ekstremite motor aktivitesi normale döndü. Sağ üst ekstremitede 2/5 güç kaybı devam ediyordu. Hasta daha sonra ihtiyaç duyulabilecek Moyamoya ameliyatı olan bir merkeze transfer edildi. Bu olgu, Moyamoya hastalığının çocuklarda akut iskemik inmelerin klasik etiyolojisi olarak düşünülmesinin önemini vurgulamaktadır. Ayrıca, Türk popülasyonu arasında nadir görülen prezentasyona ve Moyamoya hastalıklarının tanısını kolaylaştırmak için nörovasküler görüntüleme tekniklerinin kullanımına vurgu yapmaktadır.

Anahtar Kelimeler: Bebek, Moyamoya hastalığı, serebral anjiyografi, akut inme

Introduction

Moyamoya disease was first described in Japanese literature in 1957, and 12 years later, in 1969, Suzuki and Takaku used "Moyamoya disease". Its etiology is an undetermined vasculopathy and is thought to be mainly influenced by the Internal carotid artery and the Wills circle. Moyamoya disease has been associated with hereditary conditions (sickle cell anemia, neurofibromatosis type 1, Down syndrome) and acquired conditions (chronic meningitis, intracranial mass, cranial radiotherapy, cerebral vasculitis, etc.). We present an 8-month-old moyamoya case with weakness in the right lower and upper extremities.

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Case Report

The 8-month-old girl, who had no previous complaints, presented with complaints of weakness in her lower right and upper extremities. The child was admitted to the intensive care unit because he had 4/5 muscle weakness in the right arm and 3-4/5 in the right leg. Although the patient was conscious at the first physical examination, he was restless and could not use his lower right and upper extremity. Other system findings were normal. The patient has been monitored-with oxygen support. Peripheral vascular pathway open. Initial vital findings: Fever: 37.1 °C, heart rate: 160/minute, blood pressure: 85/40 mmHg measured. Fluid support from 100 cc/kg was started. The patient's lower and upper extremity X-rays were taken and interpreted. Encephalitis, meningitis, and intracranial mass were considered in the differential diagnosis of the patient. Cranial tomography of the patient was taken. There was no intracranial mass. The patient underwent a lumbar puncture. Both biochemical tests and culture were sent in the cerebrospinal fluid (CSF). Brain-neck CT angiography examinations and non-contrast cranial computed tomography (CT) examination revealed a large area suggesting ischemia in the left frontoparietal. CT angiography showed numerous slates in both distal internal cerebral arteries (ICA), branches, and a pronounced curvy appearance, and similar changes were present in the veins forming the Willis circle. In addition, very thin collateral veins-Moyamoya veins were noted in the Willis circle (Figure 1).

Cranial diffusion MRI imaging following CT noted the restriction of diffusion in the area of the left middle cerebral artery (MCA) superior segment and partly acute ischemia in the left anterior cerebral artery (ACA) irrigation area, and thickening of the vortex in this area in line with acute ischemia (Figure 2). In addition to the diffusion study, the FLAIR sequence also had gliotic changes at the centrum semiovale level on the right that suggest the previous ischemic process. No intracranial mass lesions were detected in the etiological examinations of the case. Infection scans did not reveal any culture reproduction. The patient's prior family history was found to have a history of cerebrovascular occlusion (SVO) at a young age in his mother, aunt, and grandfather. Moyamoya disease was considered in line with the imaging findings of the case. Low molecular weight heparin (2 mg/kg/day), aspirin (2 mg/kg/day) and levetiracetam (20 mg/kg/day) were started. In clinical follow-up, the patient had focal seizures in the left arm, and electroencephalography showed no seizure activity. On the fifth day of follow-up, motor activity in the lower right extremity was seen to return to normal. Power loss was continuing in the upper right extremity at 2-3/5. The patient was consulted with pediatric hematology, pediatric neurology, and neurosurgery. It was recommended that the patient be discharged with low molecular weight heparin, aspirin, and levetiracetam. In addition, neurosurgery approved his referral to a place where Moyamoya surgery was performed, and the patient was then transferred to a center with Moyamoya surgery, which may be required.

Discussion

Moyamoya disease is a rare vasculopathy characterized by angiographic findings of abnormal vascular distribution and bilateral occlusion of the internal carotid artery terminal section, whose etiology is unclear.[1] The disease often presents in the form of a transient ischemic attack (TTIA) or ischemic

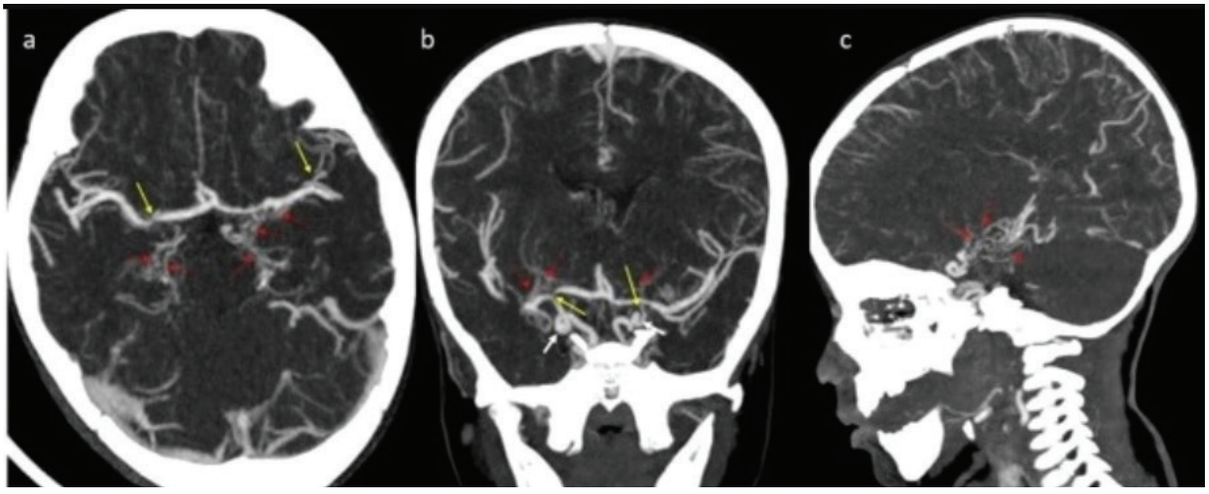

Figure 1. CT angiography examination, axial (a), coronal (b), and sagittal (c) planes; a, occlusions (yellow arrows) in the left MCA superior segment at the right ICA bifurcation level; b, bilateral tortiyoze ICA (white arrows), occlusions in the right ICA bifurcation segment and the left ICA supraklinoid segment (yellow arrows); in all three images, thin curvy collateral veins, Moyamoya veins are shown with red arrows

CT: Computed tomography, MCA: Middle cerebral artery, ICA: Internal cerebral arteries

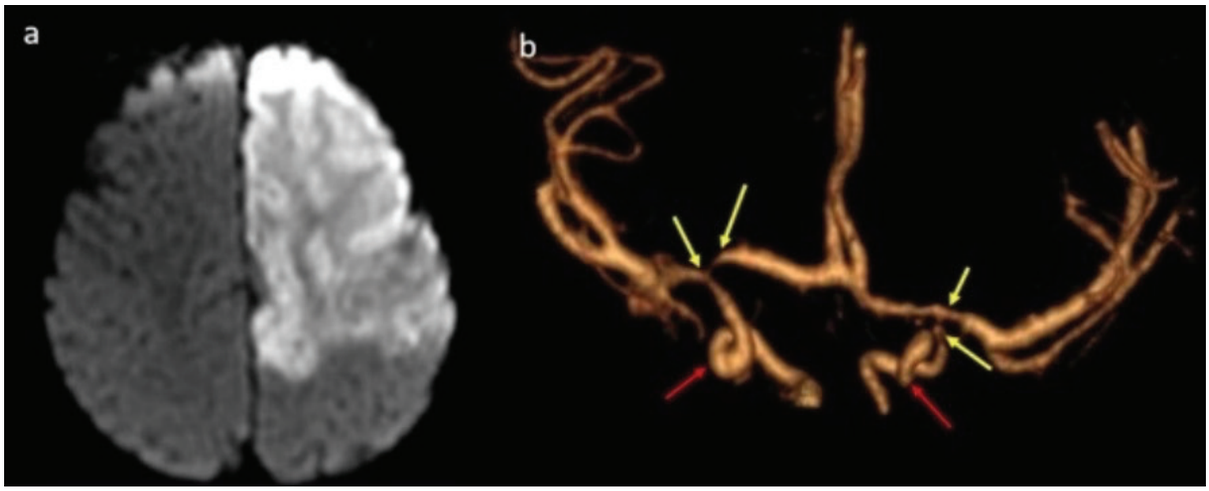

Figure 2. a) Diffusion-weighted MRI imaging, constraint restriction compatible with acute ischemia in the frontal and parietal on the left; b) CT angiography 3D VR imaging shows curvy distal ICA's with red arrows, slacks with yellow arrows

ICA: Internal cerebral arteries, CT: Computed tomography, MRI: Magnetic resonance imaging

infarction.[2,3] However, unlike children, intracranial bleeding was reported in the adult patient group.[4] Seizures occur in child and adult case groups, often complications of ischemic or hemorrhagic events. Children may experience a decline in the cognitive function directly related to the number of ischemic events undergoing and chronic hypoxemia exposed. Although its relationship with hereditary or acquired causes is widely reported in the literature, its etiology is unclear. Moyamoya appearance; neonatal anoxia, trauma, basil meningitis, neurofibromatosis type 1, tuberculosis, Sturge-weber syndrome, brain tumors, Marfan syndrome, Turner syndrome, cerebral dissection, sickle cell anemia, Down syndrome, Alagille syndrome can cause and therefore should be kept in mind in the differential diagnosis.[3] In our case, the perinatal history was normal, and there was no history of trauma, physical examination findings of syndromes, and no clinical findings were detected.

The disease is a bimodal distribution, occurs in children at an average age of 2-17 years, while in adults, it occurs at an average age of 30-40 years. While motor symptoms such as hemiparesis are observed first in ischemic attacks, aphasia, and dysesthesia follow this process.[5] The first symptom was an ischemic cerebrovascular stroke and was an infant patient outside the age range specified in the literature.

As with other cerebrovascular diseases, brain tomography is the first diagnostic imaging method for moyamoya disease suspected. Although the brain is very successful in distinguishing CT ischemic or hemorrhagic stroke, CT or MRI angiography examinations are needed for blocked vessel imaging. In angiographic imaging, moyamoya veins, which give the disease its name, are monitored in very thin collaterals, causing the appearance of clouds or cigarette smoke (Moyamoya).[6] In line with angiographic evaluation, possible or definitive moyamoya disease can be diagnosed.[7] In our case, there were common Moyamoya veins.

Today, an utterly therapeutic approach has not yet been developed. It should be remembered that medical treatments do not stop the progression of the disease but contribute to reducing its complications. Since there is no initial treatment for the disease, standard treatment protocols for stroke or hemorrhage are applied. Each case should be evaluated separately, and the option of surgical treatment should also be considered. Revascularization procedures such as surgical anastomosis from ICA to ECA can reduce the quality of life and ischemic complications.[8] Due to the rapid progression of the clinic in our case, the succession of ischemic attacks over two months, Moyamoya surgery was referred to a center for evaluation of the surgical treatment option. In the literature, there are many patients with moyamoya disease in the older age group. However, moyamoya disease should be kept in mind in patients under the age of 1 who present with muscle weakness and seizures.

Conclusion

The range of differential diagnoses in the child patient presenting with ischemic stroke is vast. Today, there is no optimal treatment for Moyamoya disease, but in fast-moving cases, urgent diagnosis, the necessary anticoagulation treatment should be started, and, where necessary, surgical treatment should be considered. Our case will contribute to the literature in terms of considering vascular pathologies in the differential diagnosis of an infant patient presenting with hemiparesis and hemiplegia.

Ethics

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: E.B., Concept: E.B., M.B., D.B., Design: E.B., M.B., D.B., Data Collection or Processing: E.B., Analysis or Interpretation: E.B., Literature Search: E.B., A.D., Writing: E.B., A.D.

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