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Case Report

Moyamoya syndrome in a pediatric patient with congenital human immunodeficiency virus type 1 infection resulting in intracranial hemorrhage☆

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ABSTRACT

In the era of Antiretroviral Therapy (ART) in which human immunodeficiency virus type 1 (HIV-1) infection affected children can expect a better prognosis, the importance of careful follow up of pediatric HIV-1 cases for neurological complications has been growing. We present a case of hemorrhagic Moyamoya syndrome in a child with congenital HIV-1 infection. A 10-year-old girl was referred to our hospital for the treatment of *Pneumocystis Jirovecii* Pneumonia (PCP: Pneumocystis pneumonia). Her HIV-1 control was poor and Moyamoya syndrome was found during the opportunistic infection screening at admission. Despite subsequent successful treatment of PCP and HIV-1 infection, we could not save her life due to the intracranial hemorrhage caused by Moyamoya syndrome. A few reported cases of Moyamoya syndrome associated with HIV-1 infection have shown negative outcomes when the control of HIV-1 infection is unsuccessful. Recently "HIV-associated vasculopathy" has been used to describe the cerebrovascular disorder related to HIV-1 infection that is caused by the endothelial dysfunction induced from chronic inflammation and cytokine imbalances due to HIV-1 infection. We assumed that "HIV-associated vasculopathy" may have contributed to the development of collateral vessels impairment related to the bleeding, although the mechanism of vascular damage with HIV-1 infection is not yet well defined. Therefore proper management of the HIV-1 infection is crucial for Moyamoya syndrome with HIV-1 cases. Furthermore it is better to take into account the risk of intracerebral hemorrhage when considering the indication and timing of the revascularization surgery, although generally hemorrhaging is rare in Moyamoya disease in children.

1. Introduction

Although the number of new patients in children has decreased by 50% compared to the year 2000 due to the expansion of the Prevention of Mother to Child Transmission of HIV (PMTCT) program, about a hundred fifty thousand newly diagnosed HIV-1 infections in children were still reported worldwide in 2015 [1]. The introduction of ART has enabled HIV-1 infected children to achieve long-term survival, while various neurological complications have also become apparent, such as HIV encephalopathy, developmental disorders, behavior problems, central nervous system (CNS) infections due to opportunistic infections, cerebrovascular disorder, convulsions, central nervous system tumors, neuropathy and so on [2,3]. One of the cerebrovascular disorders associated with HIV-1 infection is Moyamoya syndrome and several cases have been reported [4–9]. Moyamoya syndrome is the Moyamoya-like vasculopathy associated with underlying disease such as systemic infections, autoimmune diseases, metabolic diseases, blood diseases, endocrine diseases and others [10–12]. Moyamoya disease is defined as stenosis or obstruction of the proximal part of the intracranial carotid artery, the anterior cerebral artery and the middle cerebral artery, and abnormal blood vessels in the vicinity. Symptoms, pathology, and cerebrovascular findings vary

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depending on the underlying disease [10,12]. A typical symptom in pediatric cases is ischemic attack such as hyposthenia, sensorial impairment, loss of consciousness, seizure and headaches, while adult cases often present sudden onset of intracranial bleeding. Advancing age is considered to be an influence on the damage of collateral vessels [10,12]. Intracranial bleeding in children with Moyamoya disease is uncommon. Previous reports have described that only three percent of children with Moyamoya disease have presented hemorrhagic symptoms, which is significantly lower than the 25%–30% adult incidence [13]. It rarely occurs in older children and worsens their life prognosis.

We present Moyamoya syndrome in a pediatric patient with congenital HIV-1 infection that resulted in intracranial hemorrhage. As it is known that onset of intracranial hemorrhage in pediatric Moyamoya disease is rare, we assume that it was probably due to the HIV-associated vasculopathy damaging the collateral vessels.

2. Case report

A 10-year-old girl with mother-to-child transmission of HIV-1 was referred to our hospital because of dyspnea. She was born via selective caesarean section because of her mother's HIV-1 positive status. However, she was not brought in for follow up till one year of age. Even after she was diagnosed with HIV-1 infection, her treatment was difficult due to allergies to drugs, including antiretroviral drugs. Thus she had been treated with single-agent didanosine for a long period and her HIV-1 infection control was not successful as her HIV-1 viral load was quite high (2.5×10^4 copies/ml) and her CD4 count was extremely low (2/μl) when she reached our hospital. She was diagnosed with PCP and admitted to our hospital for treatment. During the opportunistic infections (OIs) screening at the admission, we found the Moyamoya syndrome with prior cerebral infarction and HIV encephalitis, while we did not find any other common OIs such as varicella-zoster virus (VZV), syphilis, tuberculosis, cytomegalovirus or fungal infections. With respect to neurological symptoms, she often experienced headaches and hyposthenia when crying since four years of age. Magnetic resonance angiography (MRA) and magnetic resonance image (MRI) examination revealed bilateral findings of middle and anterior cerebral arterial occlusion and collateral formation around the circle of Willis and old brain infarction at the right temporal occipital lobe and diffuse brain atrophy (Fig. 1). After the successful treatment of the PCP, we started the ART with a combination of stavudine, lamivudine, lopinavir, ritonavir. After two months of ART, HIV-1 viral loads became undetectable and CD4 counts

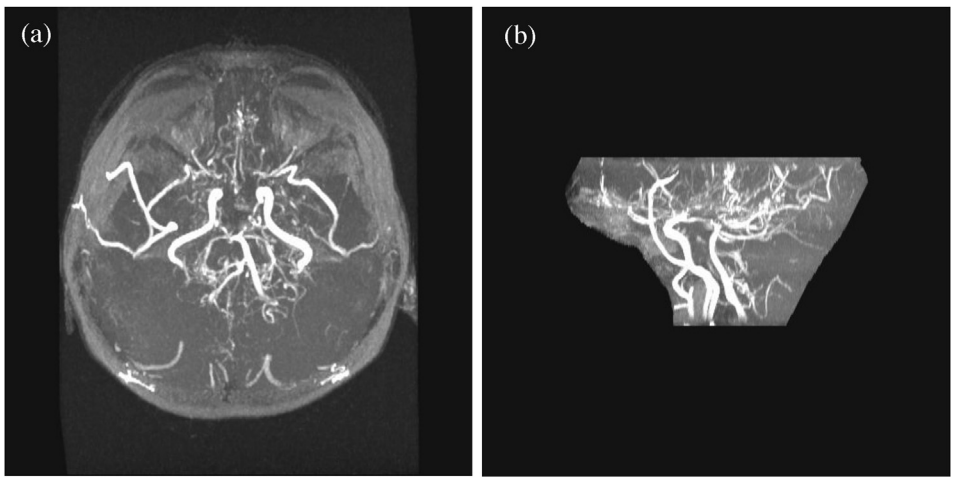

Fig. 1. a) b) MRA shows bilateral Moyamoya syndrome; complete occlusion of bilateral internal carotid artery, anterior/middle cerebral artery, and collateral formation around the circle of Willis.

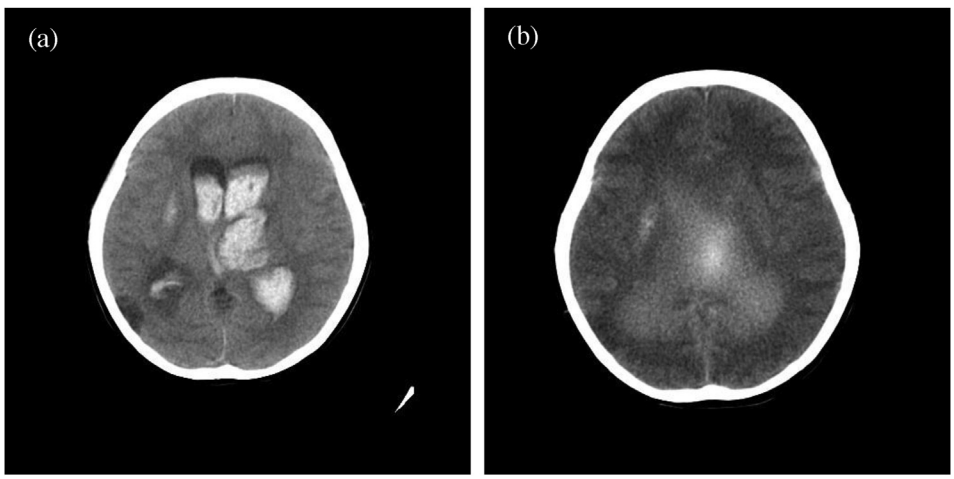

Fig. 2. a) CT image shows the left thalamic bleeding spread into ventricle, the hydrocephalus, the right putamen calcification and old cerebral infarction on right occipital lobe. b) CT image shows the residual hematoma and diffuse hypoxic encephalopathy.

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