

## Moyamoya disease in a pediatric case: A case report

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### Abstract

Moyamoya disease is extremely rare. In children, it is one of the rare causes of ischemic stroke. We present a 4-year-old female patient diagnosed with Magnetic Resonance Imaging (MRI) and Magnetic Resonance Angiography (MRA). In the case with chronic lacunar infarct in the white matter on magnetic resonance imaging, a cigarette smoke image characterized by diffuse collateral vascular networks was observed in MRA imaging. Since it is rare, we think that MRI and MRA imaging is safe and contributes to the diagnosis of the disease in children with mental and motor development retardation.

**Keywords:** Moyamoya, Magnetic resonance imaging, Lacunar infarct

### Introduction

Moyamoya disease can occlude the Willis Polygon and the internal carotid artery. It is a rare cause of ischemic stroke in children [1, 2]. Collateral circulation networks are formed to provide vascularization of the brain due to occlusion. Collaterals, especially at the level of the basal ganglia, are considered to have a cigarette smoke pattern because they have a typical appearance in Magnetic Resonance Angiography (MRA) images. The disease shows a bimodal course in the age of onset, and there are two peaks: At the age of 5 years in children and at the age of 40 years in adults [3]. Moyamoya disease may be associated with other diseases such as neurofibromatosis, Down syndrome and sickle cell anemia. Radiological examinations are very useful in diagnosing this disease. Here, we present a case of a 4-year-old who was diagnosed incidentally by magnetic resonance imaging and magnetic resonance angiography.

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### Informed Consent

The authors stated that the written consent was obtained from the parents of the patient presented with images in the study.

### Conflict of Interest

No conflict of interest was declared by the authors.

### Financial Disclosure

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## Case presentation

A 4-year-old girl was brought to the emergency room by her family with seizures. She had had seizures before and did not use her left hand much. Physical examination revealed somewhat retarded mental and motor development compared to her peers. No diffusion restriction in favor of acute ischemic infarct was detected in the diffusion Magnetic Resonance Imaging (MRI). In the brain MRI examination, there were sequelae of lacunar infarct in the deep white matter and adjacent to the caudate nucleus body on the right (Figure 1A). Laboratory tests, such as a complete blood count, Protein C-S, fibrinogen, homocysteine, were requested from the patient to reveal coagulation disorders that may cause thrombosis. The results were normal. In the Magnetic Resonance Angiography examination, the cavernous segment of the internal carotid artery was occluded on the right (Figure 1B). Widespread collaterals were also seen at this level and at the basal ganglia levels. On the 3D images, a typical appearance in the form of cigarette smoke, secondary to the collaterals, was observed (Figure 2 A-B). In the light of these findings, we diagnosed the patient with Moyamoya disease. Permission was obtained from the patient's father to publish this case and the MR images.

Figure 1: Cranial MRI imaging A) Sequelae of lacunar infarct area in Flair sequence, adjacent to the caudate nucleus body on the right (red arrow) B) Occlusion in the cavernous segment of the right internal carotid artery in MR Angiography examination (red arrow).

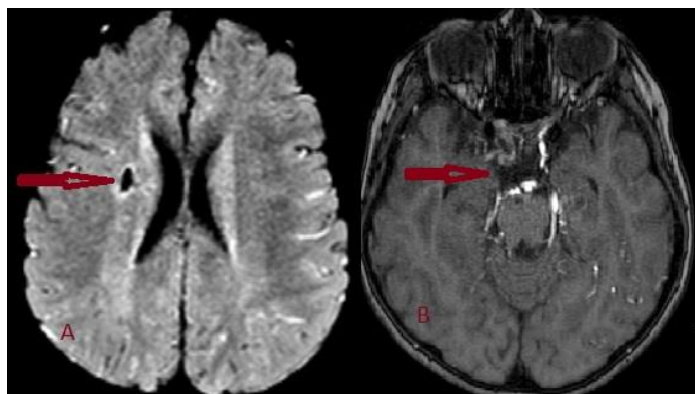
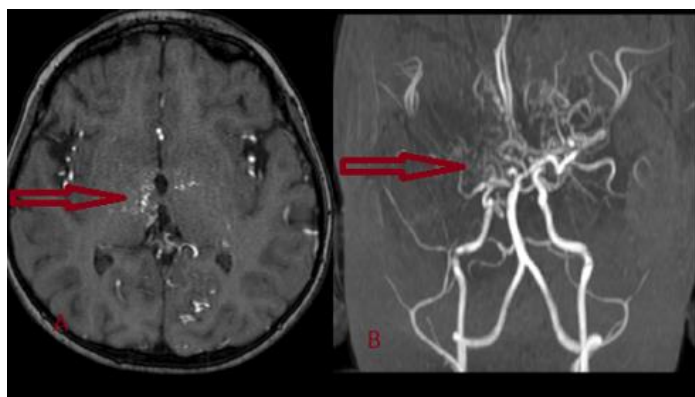


Figure 2: Cranial MR Angiography A) Axial MR Angiography image shows the collateral vascular network at the level of the basal ganglia, more prominent on the right (red arrow) B) 3D MR Angiography image shows typical cigarette smoke appearance characterized by collateral networks (red arrow).



## Discussion

The etiopathogenesis of Moyamoya disease is not fully known. However, a genetic predisposition is presumed. The disease is especially common in Japan, and its incidence is higher in families with relatives with the same disease [5]. HLA

B 40 antigen is considered suspicious for the disease, especially in those under 10 years of age. In addition, in some studies, fibroblast growth factor was high in the cerebrospinal fluid of these patients [6], which may lead to intimal thickening, smooth muscle proliferation and elastin deposition, causing suprasellar internal carotid artery stenosis. In addition, thinning of the tunica media layer and tortuous changes in the internal elastic lamina are observed [7]. However, since these are pathological findings, there is no data on this in our case. We observed multiple anastomoses in the Willis polygon region on magnetic resonance angiography images. Treatment unfortunately targets the symptoms in the acute phase. Antiepileptic drugs can be used to reduce seizures. In our case, symptomatic treatment was administered, and antiepileptic drugs were not needed. The prognosis of the disease changes with age and course, and the prognosis of the form with epileptic attacks is better than that with infarcts [8]. In our case, the prognosis was poor because of her young age and progression with infarct. MR Angiography examination plays an important role in the early diagnosis and follow-up of the disease and provides an advantage to detect even the smallest stenoses beforehand. In our case, the disease was diagnosed incidentally by Magnetic Resonance Angiography [9].

## Conclusion

Moyamoya disease is very rare. Moreover, it does not have typical clinical findings. Therefore, it is usually at the end of the differential diagnosis list of clinicians. However, it has a specific appearance in radiological MR Angiography, which is of great use in diagnosing the disease. In this case, we wanted to draw attention to the importance of radiological imaging by presenting a case that was diagnosed incidentally.

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