Unilateral Moyamoya Phenomenon Due to MCA Occlusion in a Child Presenting with Intracerebral Hemorrhage

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Summary

Spontaneous middle cerebral artery (MCA) occlusion leading to moyamoya phenomenon is different from classical moyamoya disease. Previous studies have reported such phenomena in adults with ischemic lesions, except for a solitary case in a child. We report a case of a ten-year old girl who presented with a deep intracerebral hematoma and a normal ipsilateral middle cerebral artery on initial evaluation by CT. Subsequently, on follow-up angiography, the ipsilateral MCA was occluded with evidence of unilateral basal lenticulostriate and transdural collateral supply causing a 'moyamoya' phenomenon.

This report discusses a rare and interesting disease from the perspective of 'moyamoya' phenomenon and angiogenesis.

Introduction

Spontaneous MCA occlusion is associated with moyamoya phenomenon around the occlusion site in the proximal MCA¹⁻⁹. Unlike the classical moyamoya disease, the occlusion is unilateral and does not involve the internal carotid artery (ICA). While the most common presentation of classical moyamoya disease in children is that of recurrent episodes of cerebral ischemia manifesting clinically as focal deficits, parasthesiae and seizures, hemorrhagic presentation is more common in adults.

This case presents a hemorrhagic event as an early clinical manifestation in a child with spontaneous MCA occlusion and a ipsilateral 'moyamoya' phenomenon. The etiology of this rare entity is unknown, although a congenital origin is proposed². We hypothesise that the arterial occlusion and 'moyamoya' phenomenon are due to vascular remodelling and neovascularisation.

Methods and Results

A ten-year-old child presented with headache, vomiting and dizziness of five days' duration. Historically, there was no loss of consciousness or seizures. Clinically, neck stiffness, right pronator drift and mild right upper motor neuron type of facial palsy were noted. A non-contrast Computed Tomographic (CT) examination showed intracerebral hematoma in the left basal ganglia (figure 1A). An intravenous contrast study (CECT) showed a normal circle of Willis and no abnormal enhancement (figure 1B). The child was followed-up and a repeat CT examination after four weeks showed resolution of the hematoma (figure 2A). However, on CECT, the left MCA enhancement was conspicuously absent (figure 2B). A diagnostic angiogram was performed the next day to rule out a vascular malformation.

The angiogram showed occlusion of left MCA with basal lenticulo-striate collaterals re-

Figure 1 Non-contrast (A) and CECT (B) when the child presented show left lentiform hematoma with intraventricular extension and enhancing bilateral MCAs.

forming the M2 segment ('moyamoya'). Collaterals were seen to MCA territory via the ipsilateral anterior and posterior cerebral arteries, and transdural collaterals from external cerebral artery branches.

The contralateral anterior and middle cere-

bral arteries and bilateral internal carotid arteries were normal. No aneurysms were seen on the collateral vessels or the circle of Willis (figure 3-5).

The routine biochemical investigations and hemogram were normal except for a raised ery-

Figure 2 Non-contrast (A) and CECT (B) after four weeks show resolution of the hemorrhage with non-enhancing left MCA.

Figure 3 DSA of left ICA, AP (A) and lateral (B), shows occlusion of left MCA at its origin.

Figure 4 A) DSA of late arterial phase of left ICA (lateral) shows collaterals from ACA to MCA territory and basal collaterals ('moyamoya' vessels) with no evidence of aneurysms. B) Right ICA (AP) angiogram appears normal.

Figure 5 Lateral view DSA of left vertebral (A) and left ECA (B) show leptomeningeal, dural and transdural collaterals to left MCA territory.

throcyte sedimentation rate (ESR). No serological markers of vasculitis were present. The child has improved and is on close follow-up.

Discussion

Spontaneous MCA occlusion leading to 'moyamoya' phenomenon was first described by Zulch et Al. in 1974¹. The disease is rare and patients reported with this disease are usually elderly who present with cerebral ischemia or hemorrhage in their fifth or sixth decade³. There is no involvement of the supraclinoid segment of the ICA on cerebral angiogram as evident in moyamoya disease. The etiology remains obscure. In the only case in which autopsy was performed, the outer diameter of MCA was thin, the internal elastic lamina was frayed and the smooth muscle layer was almost absent. There was no evidence of inflammatory or atheromatous changes. The multilayered eccentric intimal fibrous thickening characteristic of moyamoya disease was absent⁴. An accessory MCA and other associated anomalies have been reported².

'Moyamoya' phenomenon is known to present in adults with intracerebral hemorrhage. The hemorrhage may be due to aneurysms in the collaterals or proximally in the circle of Willis, pseudo-aneurysms or arterial dissections. Nijdam JR et Al. described a similar case in a thirty-four year old female presenting with left parietal hemorrhage and assigned to unilateral 'moyamoya' phenomenon⁶.

In our case, it is not known whether the occlusion preceded the hemorrhage or followed it. It is likely that the hemorrhage was the initial event. The MCA occlusion and the 'moyamoya' collaterals are probably a result of angiogenesis. Postnatal angiogenesis engages the endothelial cells, muscle cells and matrix and is involved in the maintenance, repair, regeneration, adaptation to needs and response to injury. The process includes arterial remodelling and neovascularisation¹⁰ and is mediated by factors such as nitric oxide synthase and transforming growth factor β 1¹⁰. Triggers such as shear stress, trauma or biological agents activate genetic transcription for such factors and alters their balance.

Transforming growth beta factor 1 (TGF β 1), a factor involved in angiogenesis and expression of connective tissue genes, is shown to be elevated in 'moyamoya' disease¹¹. It is per-

haps likely that similar mechanisms play a role in this rare entity.

The occlusion of MCA is also intriguing. The left MCA is seen as normal enhancing structure on the CT performed immediately after the hemorrhage, but is non-visualised on the CECT performed four weeks later, just prior to the angiogram (figure 2A,B). While this could represent a partial volume effect, it is less likely as further objective evidence of an absent MCA and developed collaterals was demonstrated on the subsequent angiogram. The absence of occlusion of other arterial segments in the contralateral anterior and bilateral posterior

or circulation is also noteworthy. The selective MCA involvement could represent that segment's vulnerability to a triggering factor or event. This arterial segmental vulnerability could be related to the differences in phylogenetic age besides differing anatomical and physiological characteristics that may co-exist in two consecutive arterial segments¹⁰. It could also represent an isolated event in an otherwise evolving process with potential temporal vulnerability of other arterial segments. Further data and serial studies, perhaps with histopathological correlation, may help in understanding the pathogenesis better.

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