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Cyclic headaches in β-thalassemia intermedia case presenting as moyamoya syndrome

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Moyamoya disease is a cerebrovascular disorder with unknown cause characterized by the occlusion of the bilateral internal carotid arteries (ICA) and proximal segments of ICA.[1,2] On the other hand, moyamoya syndrome (MMS) is a rare form of this condition with underlying several pathologies including hematologic disorders, congenital syndromes, vascular malformations or vasculitis after irradiation, infections, and head trauma.[1]

The symptoms of MMS are headache, seizure, and recurrent transient ischemic attacks. MMS frequently presents with the symptoms of occlusion in children, whereas in adults, the symptoms are mainly due to subarachnoid hemorrhage.[3] The collateral vessels which is a compensatory mechanism occur as a result of obstruction that resemble puff of smoke on digital subtraction angiography and magnetic resonance angiography (MRA).[1,2]

A β-thalassemia intermedia patient of 51 with cyclic headaches was investigated using MR imaging which demonstrated focal chronic infarcts and on MRA bilateral ICA were occluded at the level of petrous segment whereas vascular supply was from external carotid artery and by collateral development. At the posterior circulation, microangiopathic collaterals at thalamus and basal ganglia were detected originating from basilar artery and its branches (Figure 1a-c).

Few cases of MMS with β-thalassemia reported were under the age of 20 in our research of the literature. Among hemoglobinopathies, β-thalassemia intermedia is very rarely associated with MMS however, sickle cell anemia is the most frequent type.[4] Although silent strokes could be detected in the young patients, with the progression of moyamoya vessels we did not find any ischemic changes in our 51-year-old patient on MRI.[5] This β-thalassemia intermedia patient is an exclusive MMS case with the findings of cerebrovascular occlusion and collateral vessels demonstrated on MRA which is the preferred noninvasive imaging modality in the diagnosis.

Conflict of Interests

The author declares no conflict of interest in this study.

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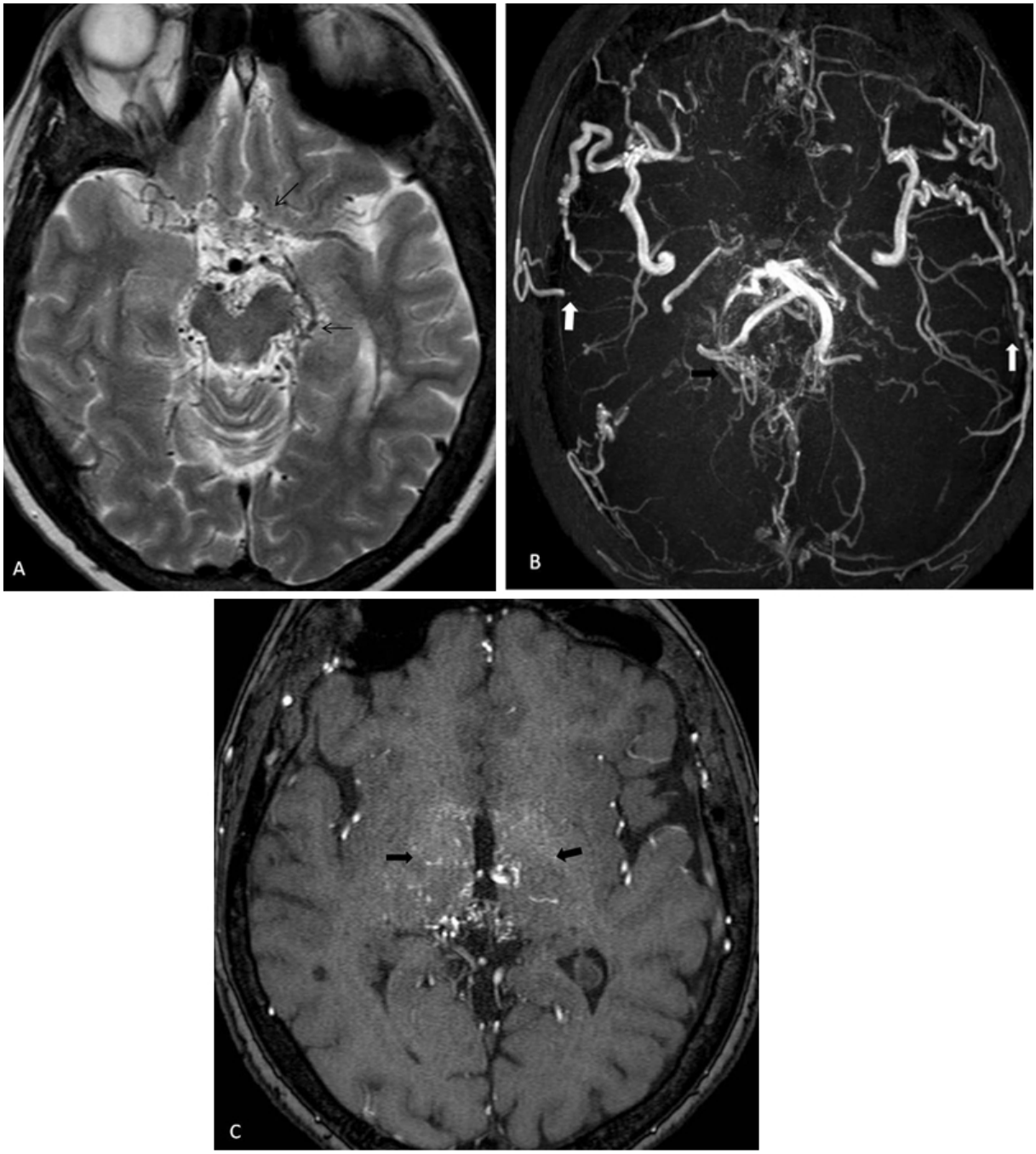

Figure 1. (a) Axial T2-weighted magnetic resonance (MR) image shows microvascular collaterals at the level of perimesencephalic cisterns and vascular supply to anterior cerebral artery and middle cerebral artery at the anterior circulation (arrows). (b) Axial MR angiography image reveals microangiopathic collaterals at the posterior circulation originating from basilar artery and its branches (black arrow). There was vascular supply from an external carotid artery by collateral development (white arrow) and bilateral internal carotid arteries occlusion at the petrous segment. (c) Axial MR angiography image demonstrates microangiopathic collaterals resembling puff of smoke (moyamoya vessels) at thalamus and basal ganglia (arrows).

References

1. Fukui M. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ('moyamoya' disease). Research Committee on Spontaneous Occlusion of the Circle of Willis (Moyamoya Disease) of the Ministry of Health and Welfare, Japan. Clin Neurol Neurosurg 1997; 99(Suppl 2): S238-S240.
2. Houkin K, Aoki T, Takahashi A, Abe H. Diagnosis of moyamoya disease with magnetic resonance angiography. Stroke 1994; 25(11): 2159-64.
3. Suzuki J, Kodama N. Moyamoya disease--a review. Stroke 1983; 14(1): 104-9.
4. Marden FA, Putman CM, Grant JM, Greenberg J. Moyamoya disease associated with hemoglobin Fairfax and beta-thalassemia. Pediatr Neurol 2008; 38(2): 130-2.
5. Goksel BK, Ozdogu H, Yildirim T, Oguzkurt L, Asma S. Beta-thalassemia intermedia associated with moyamoya syndrome. J Clin Neurosci 2010; 17(7): 919-20.