Moyamoya disease with exaggerated startle response: A rare co-occurrence

Dear Sir,

Moyamoya disease (MMD) presenting with movement disorders in the form of chorea, dystonia, and diskinesias has been reported before; however, MMD with exaggerated startle response has not been reported previously.[1,2] Recently, we came across a case of MMD featuring exaggerated startle response.

A 7-years-old boy, presented with the sudden jerky movements of body in the form of blink, flexion of neck and trunk, and abduction of arm especially in response to loud noise since the age of 6 years [Video 1]. It often resulted in fall and injuries. There was no history of loss of consciousness with these jerks. He had past history of recurrent strokes at the age of 3 years. No other family members or siblings were affected with similar illness. At the time of presentation he was conscious, alert, and was able to understand commands. His speech was limited to speaking bi-syllables only. Residual right hemiparesis with spasticity was present. Involuntary movement in the form of exaggerated startle response to sudden, loud noise was present. No spontaneous involuntary movements were observed. Computed tomography (CT) scan of head showed bilateral cerebral hemisphere infarcts in middle cerebral artery territory. He was diagnosed to have MMD on the basis of angiographic findings which showed occlusion of bilateral supraclinoid internal carotid arteries and proximal middle cerebral arteries with bilateral basal collaterals as well as right posterior cerebral artery (P1) stenosis [Figure 1].

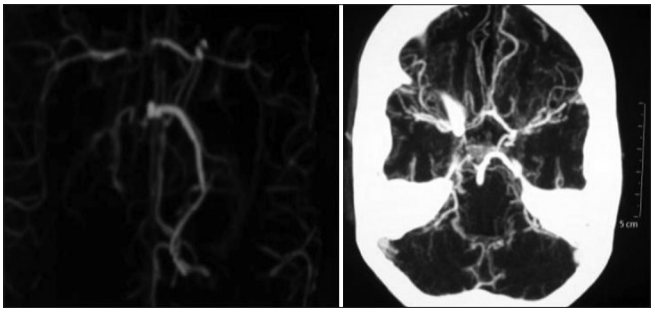

Figure 1: Computed tomography angiography brain showing occlusion of bilateral supraclinoid internal carotid arteries and proximal middle cerebral arteries with collaterals at the base of the brain suggestive of moyamoya disease along with stenosis of P1 segment of right posterior cerebral artery

Exaggerated startle syndromes (hyperekplexia) consist of an excessive motor response or jump, to unexpected auditory, somesthetic, and visual stimuli. An exaggerated startle syndrome may be due to local brainstem pathology (anoxia, inflammatory lesions, and hemorrhage) and also occurs as an inherited condition transmitted as an autosomal-dominant trait.[3]

Our patient presented with recurrent ischemic strokes at the age of 3 years and exaggerated startle response since the age 6 years [Video 1]. Development of exaggerated startle response in this patient can be explained by the ischemia of brainstem with the progression of moyamoya

vasculopathy involving posterior circulation. Jayakumar, *et al*.,[4] in their series reported that posterior circulation is frequently involved in MMD as evident by angiography; ischemic events of the posterior circulation are not frequent, as the posterior circulation acts as collateral pathway for the diseased anterior circulation till the later stage. Mugikura, *et al*.,[5] reported high prevalence of clinical strokes and infarctions in patients diagnosed before 4 years of age, associated with advanced steno-occlusive lesions of the posterior cerebral artery (PCA).

To conclude, this is the first report of sporadic exaggerated startle syndrome (sporadic hyperekplexia) in association with MMD. This case suggests that moyamoya syndrome should be considered in differential diagnosis of exaggerated startle response especially on the background of strokes in childhood.

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