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Letter to the Editor

Rare association of moyamoya syndrome with myelin oligodendrocyte glycoprotein encephalitis

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Dear Sir,

Moyamoya angiopathy (MMA) is characterised by slowly progressive occlusion of the terminal portion of the internal carotid artery (ICA) and the development of collaterals.[1] The primary is an idiopathic called moyamoya disease. Secondary is associated with other causes known as moyamoya syndrome (MMS). Myelin oligodendrocyte glycoprotein (MOG) spectrum diseases manifest with optic neuritis, myelitis and encephalitis.[2] We present a rare association of MMS due to MOG encephalitis.

A 7-year-old girl, previously a normal child, presented with a fever for 2 days followed by drowsiness for the past 2 months. On examination, normal anthropometry, vitals, quadriparesis (power 3/5) and exaggerated reflexes were noted. Investigations showed normal blood counts, peripheral smear, coagulation profile, homocysteine, liver and renal function, ammonia, and lactate. Magnetic resonance imaging (MRI) of the brain showed T2/FLAIR hyperintensities in asymmetric bilateral cerebral white matter, numerous serpiginous blood vessels are seen in bilateral basal ganglia and central corona radiata, and magnetic resonance angiography (MRA) revealed non-visualisation of the circle of Willis arteries suggestive of MMA. MRI orbit and spine were normal. The vasculitis profile, echocardiogram and carotid doppler were normal. Serum anti-MOG immunoglobulin G was strongly positive hence started on intravenous methylprednisolone, followed by tapering oral steroids. Digital subtraction angiogram showed, tapering with moderate stenosis of the right ICA, mild stenosis of right anterior and middle cerebral arteries (MCAs), tapering of communicating segment of left ICA, non-visualisation of A1 and marked tapering of M1 segment of MCA suggestive of bilateral MMA with Suzuki stage 3. She underwent surgical intervention in the form of a left superficial temporal artery-MCA direct bypass and encephaloduroarteriomyosynangiosis followed on the right side.

To date, there are no descriptions of MMS with MOG encephalitis. Further studies are needed to know whether the association is by chance or an effect of the former.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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