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Moyamoya Disease in Patient with Multiple Sclerosis

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Moyamoya disease is a rare progressive disease characterized by irreversible cerebrovascular occlusion. The occlusion would lead to an opening of transcortical, transdural collateral vessels. As a sequelae of this occlusion, ischemic and rarely hemorrhagic stroke happen leading to significant brain damage. We present a unique case of a 16-year-old female with a history of relapsing-remitting multiple sclerosis presented with recurrent ischemic stroke secondary to Moyamoya disease. MRI/MRA of this patient shows the typical features of Moyamoya disease.

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Moyamoya disease is a chronic occlusion of a major vessel of the circle of Willis leading to opening of cerebral collaterals. Moyamoya is common in Japan, but cases from Europe and America were reported[1]. Its etiology could be idiopathic (primary Moyamoya), secondary (inherited), hematological and post infections[2]. It has a special characteristic appearance in the angiography described as "puff of smoke"[3].

Multiple sclerosis (MS) is an inflammatory disease of the white matter of the brain characterized by relapsing neurological symptoms. The masquerade between those diseases is high, but the clinical, radiological and laboratory investigations confirm that both pathologies were present in our patient, although this is not reported before[4]. The studies emphasize early consideration of Moyamoya in the differential diagnosis of MS-spectrum disorders, which shares many features[4,5].

The aim of this presentation is to present a rare possible clinical and radiological association between Moyamoya disease and multiple sclerosis.

THE CASE

A sixteen-year-old single female had a history suggestive of relapsing, remitting MS. A year ago, she had three episodes of numbness where she recovered completely and her diagnosis was supported by radiological and laboratory findings. Recently, the patient presented with a 3-day history of left upper limb weakness and facial asymmetry. CT brain and MRI were performed to confirm the diagnosis and rule out MS relapse. CT scan and MRI confirmed the

presence of right MCA ischemic stroke on top of her demyelinating lesions, see figures 1, 2 and 3.

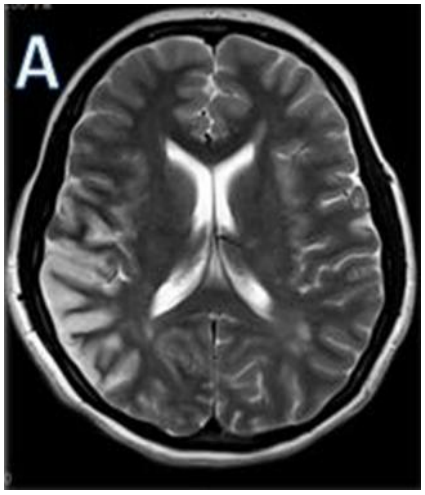

Figure 1: Brain MRI: T2 Axial Image Shows Multiple Deep White Matter, Abnormal High Signal Intensities Typical for MS along with Abnormal Right Parietal Gray Matter Signal Intensity

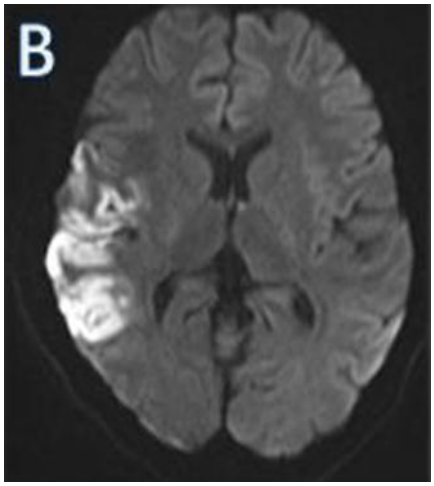

Figure 2: Axial Diffusion Weight Image Showed Restriction at the Right Parietal Gray Matter Representing Acute Ischemic Infarction

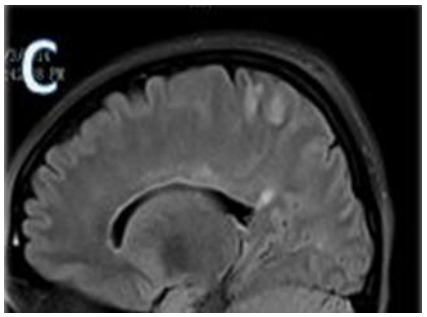

Figure 3: Sagittal Flair Image Showed Typical MS Periventricular Abnormal Signal Intensity Perpendicular to the Wall of the Lateral Ventricular

The patient already started on interferon beta 1a (Rebif) 44 Mcg SQ 3 times a week 6 months before her presentation in our facility. She denied any similar illness or presence of ischemic stroke in young members of her family.

Examination revealed that the left side facial weakness was of upper motor neuron type. The tone was decreased; the power was 3/5 with exaggerated reflex and up going planter on the left side. The following investigations were performed: autoimmune profile, lactic acid level, hypercoagulable state profile, carotid Doppler, Echo, Holter monitor and Sickle cell screening. All investigations were normal. CSF revealed the presence of oligoclonal bands (OCB) with normal protein and cell count.

Two days after her admission, the power improved significantly to 4+/5 with only very mild weakness. The patient was discharged on Aspirin 81 mg BD, physiotherapy and her previous DMT.

Two weeks later, she presented with right-sided weakness and aphasia; the CT revealed an evidence of new left MCA ischemic stroke.

Conventional angiogram revealed obliteration in supra-clinoid portion of both internal carotid arteries and bilateral MCA stenosis more on the left side associated with an opening of lenticulostriate arteries giving the appearance of "puff of smoke", narrowing of right ACA as well as right PCA, see figure 4.

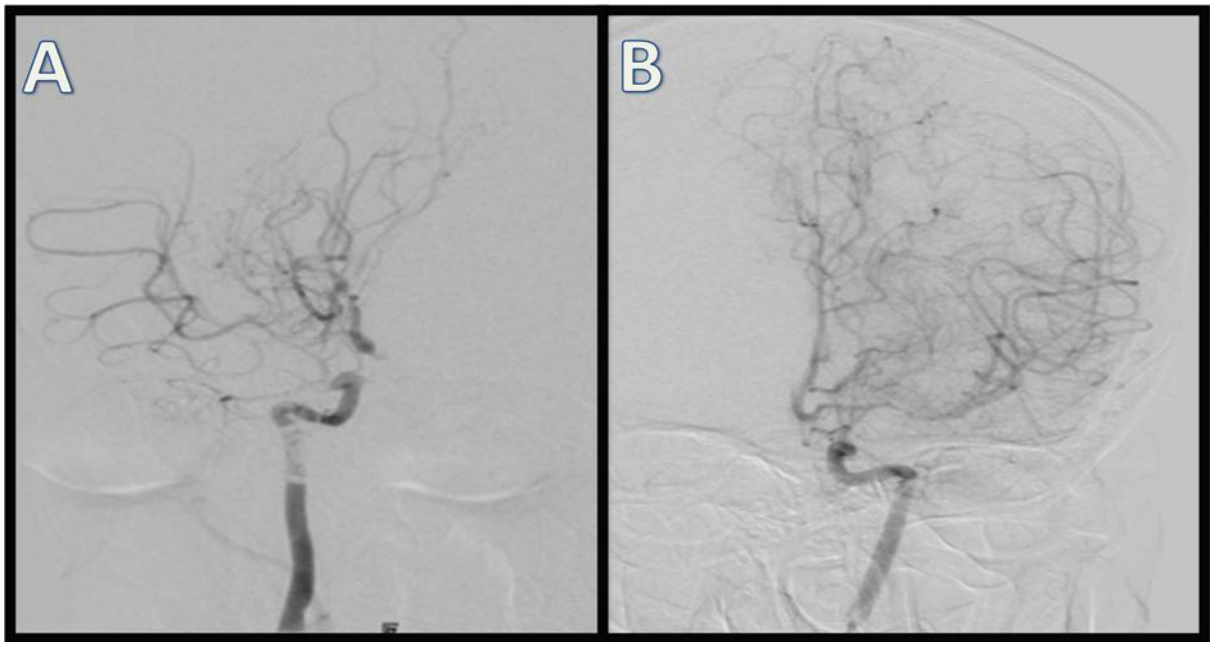

Figure 4: Cerebral Digital Subtraction Angiogram Shows Right (A) and Left (B) Supraclinoid ICA/MCA Stenosis with Hypertrophied Lenticostriated Vessels – Puff of Smoke Appearance

The patient was diagnosed with primary Moyamoya disease apart from her previous Multiple sclerosis. The patient was referred to neurosurgery for revascularization surgery to prevent further deterioration and to restore blood flow.

DISCUSSION

Moyamoya disease is a condition associated with prominent telangiectasia especially in the region of basal ganglion blood vessels and compensating occlusive arteries at the base of the

brain involving the circle of Willis. It may complicate SCD, meningitis neurofibromatosis type I, Down syndrome or as sequelae of radiotherapy. It is common in children and females[6,2,7]. It is a chronic and progressive disease, but the rapid presentation could happen. The common presentation of this disease is ischemic stroke and seizure[8-10]. Hemorrhagic stroke could happen in these cases due to rupture of the collaterals. Although Moyamoya disease and MS have a different pathophysiology, both of them could share the same presentation of recurrent neurological symptoms leading to misdiagnosis as reported in the litreature[4].

In our case, the diagnosis of MS was confirmed clinically, radiologically and by the presence of OCB in CSF and significant improvement after steroids therapy.

It was found that most cases initially are considered being compatible with multiple sclerosis; stroke and the typical angiography finding change the diagnosis to Moyamoya and this is a unique about our case report[4,5,11].

MRA is a modality of choice to diagnose Moyamoya disease where it shows the occlusion of the proximal territories and the extensive collateral demonstrating "puff of smoke appearance"[12,13]. The Treatment of this condition is a surgical revascularization with EDAS (encephalo duroarteriosynangiosis) which is considered to be the most preferred procedure. Many procedures have been done, and they have been individualized according to each case. Those procedures show some symptomatic benefit along with blood flow improvement[1,14-15].

CONCLUSION

Misdiagnosis of MS and Moyamoya disease is not uncommon; however, a careful evaluation clinically, radiology and laboratory is necessary to confirm both diseases. In our case, it was a rare association. Although future reports could reveal more cases with similar associations, giving possible linkage between MS and Moyamoya disease.

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