LETTER TO THE EDITOR

To The Editor

Steroid-Responsive Acute Left-Arm Chorea as a Presenting Symptom of Moyamoya Disease

Keywords: Chorea, Moyamoya disease, Secondary, Steroid-responsive

Moyamoya disease is a rare occlusive intracranial arteriopathy characterized by progressive narrowing of the supraclinoid internal carotid artery and other main intracranial arteries.[1] Ischemic stroke, recurrent TIAs, headache, and seizures are the characteristic symptoms of the disease.[1]

Chorea is one of the less common presenting signs of moyamoya disease, occurring only in 3–6% of patients (mainly pediatric) and almost always together with the other symptoms of the disease.[1–3]

A 44-year-old Chinese woman acutely developed intermittent choreic and choreoathetoid movements restricted to the left upper limb (Video, segment-1). Occasionally, abnormal dystonic postures of the left hand were also present. There was no family history of neurological diseases, and the patient was not taking any medication that could induce chorea. Moreover, her medical history was unremarkable except for type II diabetes, which was well compensated. During the hospitalization, the patient underwent several investigations, including the control of glycemic profile (that showed values within the normal limits), electrolytes, autoimmunity panel (rheumatoid factor [RF]; antinuclear antibody [ANA]; phospholipid antibody [aPL]; antinuclear antibody specific for extractable nuclear antigens [ENA]; thyroperoxidase antibody [TPO]; lupus anticoagulants [LA]; anti-cyclic citrullinated peptide [CCP] antibody), antistreptolysin O (ASO) titer, neoplastic markers, onconeural antibodies (GAD65, CV2, amphiphysin, Ma2; Hu, Yo, Ri), autoantibodies against neuronal surface antigens (LG1, CASPR2, AMPAR, NMDAR), and total-body CT scan that were all negative. Brain-MRI Axial FLAIR scan (Figure 1A) at the basal ganglia level with corresponding DWI (Figure 1B) and ADC map (Figure 1C) sequences showed the lack of acute vascular lesion and the presence of a silent brain infarction at the genu of left internal capsule. The axial FLAIR scan (Figure 1D) at the centrum semiovale level shows few

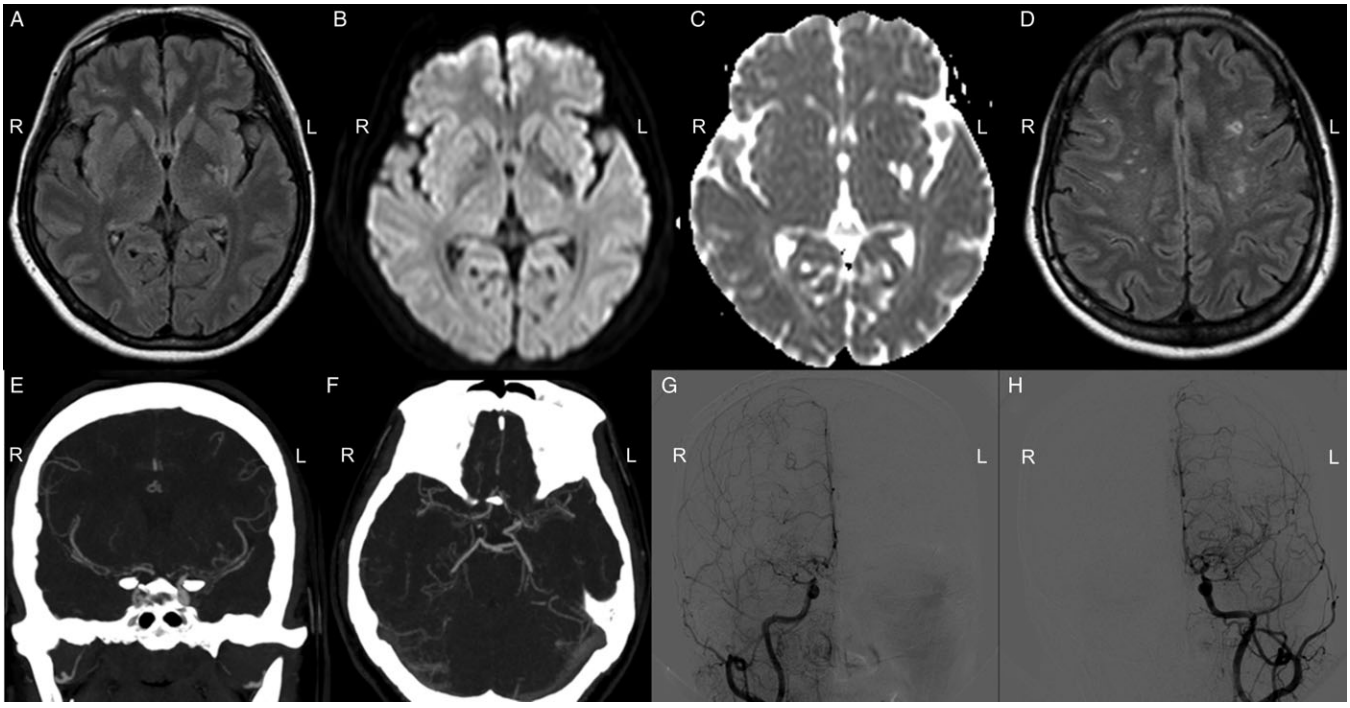

Figure 1: *Brain MRI, CT angiography, and DSA. Brain-MRI Axial FLAIR scan (A) at the basal ganglia level with corresponding DWI (B) and ADC map (C) sequences showed the lack of acute vascular lesion and the presence of a silent brain infarction at the genu of left internal capsula. The axial FLAIR scan (D) at the centrum semiovale level shows few scattered white matter hyperintensities and the "Ivy Sign" (which is a radiologic indicator of developed leptomeningeal collaterals) more evident in right leptomeningeal sulci. The findings of CT angiography reconstructed on coronal (E) and axial (F) planes were the bilateral steno-occlusion at internal carotid arteries (ICA) terminus involving both anterior cerebral arteries (ACA) and middle cerebral arteries (MCA) (right > left) and the classical evolution of the deep collateral circulation with a network of lenticulostriatal perforators. DSA by injection of right (G) and left (H) ICA confirming the occlusive arteriopathy at ICA terminus involving both ACA and MCA with an occluded right ICA and a more pronounced deep collateral network on left-hand side.*

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scattered white matter hyperintensities and the "Ivy Sign" (which is a radiologic indicator of developed leptomeningeal collaterals) more evident in right leptomeningeal sulci. CT angiography, reconstructed on coronal (Figure 1E) and axial (Figure 1F) planes, showed the presence of bilateral steno-occlusion at internal carotid arteries (ICA) terminus involving both anterior cerebral arteries (ACA) and middle cerebral arteries (MCA) (right > left) and the classical evolution of the deep collateral circulation with a network of lenticulostriatal perforators. Digital subtraction angiography (DSA) by injection of right (Figure 1G) and left (Figure 1H) ICA confirmed the occlusive arteriopathy at ICA terminus involving both ACA and MCA with an occluded right ICA and a more pronounced deep collateral network on left-hand side. The neuroradiological picture was compatible with moyamoya disease. No clinico-radiological signs that suggest fibromuscular dysplasia were found and a diagnosis of idiopathic moyamoya disease were made. The patient was treated with high dosage of IV-methylprednisolone (1gr/day for 5 days) with a prompt reduction of involuntary movements in few days (Video, segment-2). To date, 6 months after the onset of chorea, the patient's conditions are stable with no involuntary movements.

In the few cases reported, chorea was the presenting symptoms of moyamoya disease mainly in the form of acute/subacute hemichorea or diffuse chorea.[1,3] On the contrary, our patient developed only a more subtle left-arm mild intermittent chorea expanding the possible manifestations of chorea in moyamoya disease. This is an important point considering that a timely diagnosis of moyamoya disease despite atypical presenting signs is of paramount importance because of the potential for neurological worsening if treatment is delayed.[3] The etiology of chorea in moyamoya disease is still debated and little evidence suggesting causality. Some authors have speculated that chorea in moyamoya disease might occur as a result of the ischemic changes affecting the excitatory–inhibitory circuits connecting the basal ganglia and the cerebral cortex.[1,2,4] It has also been suggested that the presence of collateral vessels, with a subsequent physical disruption of normal basal ganglia physiology, could play a role.[1] Surgical revascularization led to symptoms reduction or resolution in the majority of cases. It is speculated that the clinical amelioration might be related to the improvement of basal ganglia perfusion and the regression of collateral vessels.[1] To date, there are only four pediatric case reports of moyamoya-related chorea responsive to steroids treatment that may act through the restoration of perfusion and the modulation of neurotransmitters within the basal ganglia.[2,4,5] Compared to the few previous descriptions, our case differs for two main aspects: it concerns a much older patient, and steroid-responsive chorea was the first and the only presenting symptom of the disease. Considering the lack of acute vascular lesion and the response to steroid treatment, our report supports the hypothesis that chorea in moyamoya disease could be secondary to dynamic ischemic changes and not to a structural ischemic lesioning. This could help to better understand the relationship between chorea and moyamoya disease. Furthermore, in the setting of moyamoya-related chorea, a short course of pulse steroids treatment should be tried representing a possible curative treatment of the involuntary movement disorders rather than only a temporary treatment pending definitive surgical intervention.

Conflict of interest

The authors declare that there is no conflict of interest relevant to this work.

Statement of authorship

FC: conception, organization, and execution of the research project; writing of the first draft, review, and critique of the manuscript; MZ: conception and organization of the research project; writing of the first draft, review, and critique of the manuscript; FA: conception and organization of the research project; writing of the first draft; FV: conception and organization of the research project; review and critique of the manuscript.

Ethics Statement

Written informed consent was obtained from the patient to be videoed for publication.

Supplementary material

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