An extremely rare cause of thalamic tremor: MOGAD

Mert Altıntaş*, Miraç Yıldırım*, Ömer Bektaş*, Serap Teber*

*Department of Pediatric Neurology, Ankara University Faculty of Medicine, Ankara, Turkey

OBJECTIVES

Thalamic tremor, also known as Holmes tremor, first described by Gordon Holmes in 1904, is a rare movement disorder with rest, intention and postural components that predominantly affects the proximal upper extremities unilaterally or asymmetrically, is characterized by high amplitude and low frequency (<5 Hz).[1, 2] Etiologically, the most common causes are vascular events (ischemic or hemorrhagic) and head trauma.[3] Although the occurrence of Holmes tremor in demyelinating diseases has been previously reported in the literature, there is no data on the occurrence of Holmes tremor in cases with myelin-oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOGAD).[3] In this report, we aimed to present a case who was diagnosed with MOGAD and developed Holmes tremor in the right arm during the course of the disease.

CASE PRESENTATION

A three-year-old male patient was admitted to our pediatric emergency unit due to febrile status epilepticus. Following clinical stabilization, computed tomography (CT) performed for etiological investigation and evaluation of the suitability of lumbar puncture revealed a bilateral multiple nodular hypodense areas with asymmetric distribution, more dominantly in the right frontotemporal and frontal, both insulae, and left occipital areas. In the cerebrospinal fluid (CSF) analysis performed with a suspected diagnosis of meningoencephalitis, glucose was 67mg/dL (simultaneous capillary blood glucose was 118mg/dL), protein was 22mg/dL (20-45mg/dL), and meningitis PCR panel and CSF culture were negative. MRI for differential diagnosis, revealed hyperintense multifocal lesions on T2/FLAIR sequences consistent with demyelinating diseases involving the bilateral basal ganglia, cortical and subcortical regions, and left thalamus. (Figure 1.)

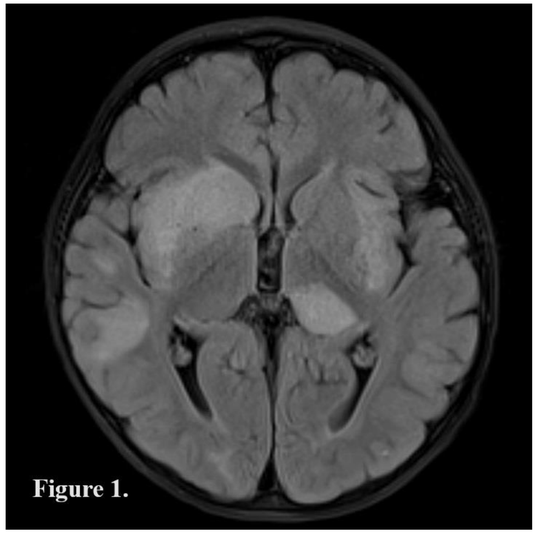

Figure 1.

Electroencephalography (EEG) demonstrated diffuse slow background activity consistent with moderate encephalopathy. The serum anti-MOG titer was positive, 1:1000. Subsequently, the patient received a diagnosis of MOGAD and underwent treatment with intravenous pulse steroid for five days and then intravenous immunoglobulin (IVIG) at a dose of 400mg/kg/d for five days. During the follow-up, it was observed that the rigidity and muscle weakness appeared in his extremities, and a slow and large-amplitude tremor appeared in the right arm, potentially attributed to thalamic involvement. After the completion of his treatment regimen, he was discharged with maintained oral methylprednisolone (MPZ) treatment and physiotherapy. Additionally, the patient was considered to be at high risk for relapse because he was male, the disease presented with acute disseminated encephalomyelitis (ADEM), and occurred at a young age, and monthly IVIG preventive treatment was planned.

DISCUSSION

MOGAD is an autoimmune disorder characterized by inflammatory demyelination of the central nervous system caused by MOG antibody-associated myelin and oligodendrocyte damage.[4] MOGAD can occur at any age, and in contrast to other inflammatory demyelinating diseases, no marked sex or racial predominance has been demonstrated.[5] Its incidence and prevalence are 1.6-3.4 per million people per year and ~20 per million, respectively.[5] It has broad and heterogeneous phenotypic spectrum with optic neuritis, myelitis, optic neuromyelitis, brainstem syndromes, encephalomyelitis (ADEM), acute disseminated encephalomyelitis, encephalitis, and seizures. Phenotypic presentations differ somewhat between pediatric and adult patients; MOGAD predominantly presents with ADEM in pediatric cases and optic neuritis and myelitis in adult cases.[4,6] Cases tend to present with lesions in the cortical and subcortical deep gray matter (including thalamus), white matter, brainstem, cerebellum, and spinal cord.[6]

However, despite its broad clinical spectrum, the occurrence of Holmes tremor in patients with MOGAD has not been previously reported in the literature. Our case is noteworthy in that MOGAD has not been previously reported as an underlying cause of Holmes tremor and contributes to the etiological diversity of Holmes tremor.

REFERENCES

1. Holmes G. On certain tremors in organic cerebral lesions. Brain 1904;27:327-375. doi: 10.1093/brain/27.3.327
2. Lenka A, Jankovic J. Tremor Syndromes: An Updated Review. Front Neurol. 2021 Jul 26:12:684835. doi: 10.3389/fneur.2021.684835.
3. Raina GB, Cersosimo MG, Silvia S Folgar SS et al. Holmes tremor: Clinical description, lesion localization, and treatment in a series of 29 cases. Neurology. 2016 Mar 8;86(10):931-8. doi: 10.1212/WNL.0000000000002440.
4. Zhuo Z, Duan Y, Tian D et al. Brain structural and functional alterations in MOG antibody disease. Mult Scler. 2021 Aug;27(9):1350-1363. doi: 10.1177/1352458520964415.
5. Banwell B, Bennett JL, Romain Marignier R et al. Diagnosis of myelin oligodendrocyte glycoprotein antibody-associated disease: International MOGAD Panel proposed criteria. Lancet Neurol. 2023 Mar;22(3):268-282. doi: 10.1016/S1474-4422(22)00431-8.
6. Rechtman A, Brill L, Zveik O et al. Volumetric Brain Loss Correlates With a Relapsing MOGAD Disease Course. Front Neurol. 2022 Mar 24:13:867190. doi: 10.3389/fneur.2022.867190.