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Case Report

Pseudovascular Revelation of Seropositive Neuromyelitis Optica Spectrum Disorder: A Case Report

K. Haddouali *[1], NEH. Sguiar Lhamdani [1], S. Bellakhdar [1,2,3], H. El Otmani [1,2], B. El Moutawakil [1,2], MA. Rafai [1,3]

[1]Department of Neurology, Ibn Rochd Hospital, Hassan II University, Faculty of Medicine and Pharmacy - Rue Tarik Ibn Ziad, 20000, Casablanca, Morocco.
[2]Genetics Laboratory, Hassan II University, Faculty of Medicine and Pharmacy, Casablanca, Morocco.
[3]Research Laboratory on Diseases of the Nervous System, Neurosensory and Handicap, Hassan II University, Faculty of Medicine and Pharmacy, Casablanca, Morocco.

*Corresponding author: Kamal Haddouali; *kamal.haddouali@gmail.com*

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Abstract

Symptom onset in Neuromyelitis Optica spectrum disorder (NMOSD) is often acute, while sudden onset is rare and suggests vascular causes. We report the case of a 63-year-old woman who had two sudden attacks, blindness in the left eye and weakness of the lower limbs. The presence of a diencephalic hypersignal around the third ventricle and a bright spotty lesion on the MRI had evoked the NMOSD diagnosis which was confirmed by positive anti-aquaporin 4 antibodies in the serum. The patient was treated with cyclophosphamide with no relapses after 6 months of treatment. Few case reports have been published on this subject, testifying to the rarity of this "pseudo-vascular" mode of revelation of NMOSD, which may be a source of diagnostic error or delay.

Keywords: ***Neuromyelitis Optica spectrum disorder, pseudo-vascular, spinal cord infarction, myelitis.***

Introduction

Neuromyelitis Optica spectrum disorder (NMOSD) is an autoimmune inflammatory demyelinating disease of the central nervous system characterized by acute onset of neurological deficits. However, a hyperacute or sudden presentation is considered a red flag for the diagnosis of NMOSD and prompts investigation of arterial vascular etiologies including spinal cord infarction (SCI) and ischemic optic neuropathy [1]. We present a case of a patient with NMOSD manifested with two pseudo-vascular episodes.

Case report

A 63-year-old female patient was admitted to our department in June 2022 with sudden weakness of the lower limbs, low back pain and bladder dysfunction consisting of dysuria without fecal dysfunction. She had a history of sudden blindness in the left eye at the age of 39, the etiological investigation was negative and therefore no treatment was received. Neurological examination showed paraparesis at 3/5 (Medical Research Council strength score), impaired proprioception and hyperreflexia in the lower limbs and bilateral Babinski. Ophthalmological examination revealed negative luminous perception on the left eye and visual acuity at 0.30 on the right eye. Funduscopy showed bilateral papillary atrophy. Upper limbs, bulbar and respiratory functions were normal. Based on these findings, we suggested vascular etiologies, particularly embolic or arteritic mechanisms. Magnetic resonance imaging (MRI) of the spinal cord revealed on sagittal T2 and STIR-weighted (Short Tau Inversion-Recuperation) sections an extensive hyperintense lesion from D2 to D4, occupying more than 50% of the surface area on axial T2-weighted sections with a "bright spotty lesion" sign. No enhancement was found on post-contrast T1-weighted sections. Cerebral MRI displayed on sagittal and axial T2-weighted sections a diencephalic hypersignal around the third ventricle and no signal abnormalities were found on the optic nerves (**Figure 1**). Retinal angiography revealed papillary atrophy without evidence of vascular involvement. Lumbar puncture (LP) testing demonstrated lymphocytic pleocytosis, with normal protein and glucose levels. Intrathecal synthesis and oligoclonal bands were not tested. Screening for aquaporin-4 receptor antibody (AQP4) by immunofluorescence was positive (1/64). Inflammatory, metabolic and infectious tests were negative. The diagnosis of NMOSD was therefore confirmed according to the 2015 Wingerchuk criteria. The

patient received intravenous methylprednisolone followed by oral corticosteroid therapy with a slight improvement in motor deficit and persistent dysuria. Cyclophosphamide was initiated with no new relapse after 6 months of treatment.

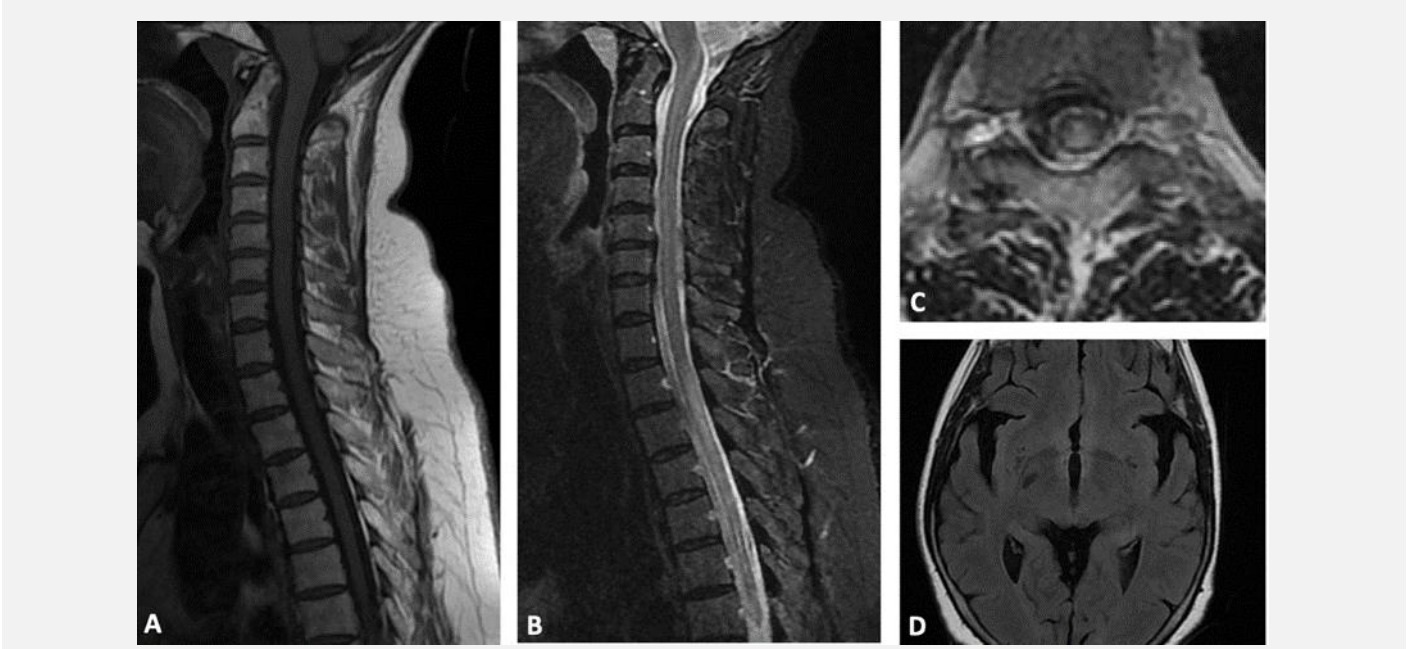

Fig 1: Spinal cord and brain MRI of the patient with seropositive NMOSD. (A) sagittal T1-weighted section of the spinal cord shows an isointense lesion of thoracic spinal cord. (B) sagittal STIR-weighted section of the spinal cord shows an extensive hyperintense lesion from D2 to D4. (C) axial T2-weighted section of the spinal cord shows a hyperintense lesion occupying more than 50% of the surface area with a bright spotty lesion sign. (D) axial FLAIR-weighted section of brain shows a diencephalic hyperintensity around the third ventricle.

Discussion

The sudden onset of NMOSD is unusual, it is a red flag which requires a prompt investigation of differential diagnoses, particularly vascular diseases. This clinical presentation could be explained by the pathophysiology of NMOSD, in which cytotoxic edema is caused by complement activation after anti-aquaporin 4 antibodies bind to their astrocytic receptors (complement-dependent cytotoxicity), and activation of effector cells such as natural killer cells (antibody-dependent cell-mediated cytotoxicity), inducing cell death and thus extensive demyelination and necrosis [2]. The second atypical feature in our patient's case was the occurrence of the second relapse after a long period of time (24 years after the first one), whereas relapses in NMOSD seropositive patients are frequent on short term in a majority of series of the literature. A large cohort reported that 60% of patients untreated with immunosuppressants have a second relapse in the first year and up to 90% have it in the third year [3]. Despite these clinical and evolutionary variations, we redirected our etiological focus towards NMOSD, in accordance with the encephalic and spinal cord MRI findings, specifically the presence of the bright spotty lesion and the diencephalic hyperintense lesion which are radiological arguments suggestive of NMOSD. The absence of vertebral body hypersignal was also an argument against spinal cord infarction [3]. To our knowledge, few case reports have been published on this subject, testifying to the rarity of this "pseudo-vascular" mode of revelation of NMOSD, which may be a source of diagnostic error or delay [4,5].

Conclusion

A sudden onset of symptoms is rarely reported in the NMOSD requiring the search for radiological and biological biomarkers allowing it to be distinguished from vascular etiologies. Other reports are necessary to understand the mechanism of this atypia.

Statements and Declarations

Conflict of Interest

The authors declare no conflict of interest regarding this case report.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed Consent

Informed consent was obtained from the patient included in the study.

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References

[1] Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology. 14 juill 2015;85(2):177-89. doi: 10.1212/WNL.0000000000001729.

[2] Papadopoulos MC, Verkman AS. Aquaporin 4 and neuromyelitis optica. Lancet Neurol. juin 2012;11(6):535-44. doi: 10.1016/S1474-4422(12)70133-3

[3] Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol. sept 2007;6(9):805-15. doi: 10.1016/S1474-4422(07)70216-8

[4] Mansour M, Ghariani A, Baradai N, Meselmani M, Kacem A, Mrissa R, et al. Neuromyelitis optica spectrum disorder (NMOSD): pseudo vascular presentation. Clin Case Rep J. 2021;2(7):1-3. ISSN: 2767-0007

[5] Elkhoyaali A, Omari A, Hanafi Y, Bouabid Y, Chammout F, Bargach T and al. Neuromyelitis optica (Devic's disease) presenting as reversible sudden bilateral blindness in a child. J Fr Ophtalmol. 2017 May;40(5):e167-e168. French. doi: 10.1016/j.jfo.2017.01.008.