Journal of the Neurological Sciences 405 (2019) 116417

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Letter to the Editor

MOG antibody-related isolated rhombencephalitis revealed by paroxysmal dysarthria

Dear Editor,

A 59-year-old female was admitted to the neurology ward with a two-month history of recurrent episodes characterized by an ascending feeling of constriction starting in the lower limbs, followed by speech impairment, dizziness and a lack of strength in both hands. The symptoms lasted 10 s and recurred 30–40 times a day. She did not report a prodromal infectious episode. She suffered from mitral regurgitation and from peptic ulcer and was treated by bisoprolol and pantoprazole. Her clinical neurological examination was unremarkable. Interictal electroencephalogram revealed no epileptic discharges. Carbamazepine (200 mg tid) was given with a dramatic decrease in the frequency of paroxysmal symptom occurrence. Brain magnetic resonances imaging (MRI) showed a bilateral tegmental midbrain lesion on T2-weighted and Fluid-Attenuated Inverted Recovery (FLAIR) sequences (Fig. 1a, b). Follow-up brain MRI at two months showed partial gadolinium enhancement on T1-weighted images (Fig. 1c). Laboratory tests were normal, including: thyroid hormones, angiotensin-converting enzyme, antinuclear antibodies, immunoglobulins, CRP, vitamin B12, folic acid, cupremia. The patient was seronegative for anti-HIV, anti-GQ1b, anti-LgI1 and anti-aquaporin 4 Abs. CSF examination showed a normal cell count and protein level, no CSF-specific oligoclonal bands and no anti-NMDA-R-Abs. The CSF-TPHA (Treponema pallidum hemagglutination) was negative. The patient was positive for anti-MOG antibodies in the serum (using a cell-based assay, Eurimmun). Her spinal cord MRI and ophthalmologic examination were normal. Having excluded alternative causes, the diagnosis of anti-MOG rhombencephalitis was retained, encountering the criteria proposed by Jarius et al. [1].

A 3-day course of intravenous methylprednisolone at 1000 mg/day was administered and oral prednisolone was started at 64 mg/day during 21 weeks with progressive tapering. Complete disappearance of her paroxysmal symptoms occurred in less than five days and no relapse occurred during the following six months but anti-MOG antibodies testing was still positive in the serum of the patient at the end of this follow-up. Another follow-up brain MRI at five months showed a small central tegmental lesion on T2-weighted and FLAIR sequences without contrast enhancement on T1-weighted images.

Our patient showed stereotypical paroxysmal symptoms, more commonly described in multiple sclerosis [2]. First described by Parker in 1946, paroxysmal dysarthria is a rare syndrome and manifests itself by sudden, recurrent, stereotyped, brief episodes of dysarthria [3]. Response to carbamazepine is often dramatic, but if left untreated, it can last for days up to a few months. It can be associated to ataxia or limb clumsiness (paroxysmal dysarthria and ataxia syndrome). The extent of the midbrain lesion would be related to the severity of clinical symptoms [4]. Rare other causes have been described as Behcet's disease, Bickerstaff encephalitis or midbrain infarct. Recently a patient with paroxysmal dysarthria and an isolated midbrain lesion depicted as solitary sclerosis was reported, but anti-MOG antibodies were not looked for [5]. Only a few patients having this symptomatology with an isolated brainstem lesion have been reported and our observation supports its causality in paroxysmal dysarthria occurrence. A lesion of the cerebello-thalamo-cortical pathway at the level of the decussation of the superior cerebellar peduncle fibres with ephaptic axonal activation within demyelinated fibre tracts was suggested as a possible explanation for paroxysmal dysarthria [6].

The spectrum of anti-Myelin oligodendrocyte glycoprotein (MOG) antibody (Ab)-associated encephalomyelitis includes optic neuritis, CRION (chronic relapsing inflammatory optic neuritis), neuromyelitis optica (NMO)-like disease, acute disseminated encephalomyelitis, transverse myelitis, multiple sclerosis (MS)-like disease and even pseudotumor cerebri-like or small vessels vasculitis-like presentations [7,8]. Anti-MOG Ab-related disease has a wide range of age of onset, with diverse clinical presentations, including in the paediatric population. Older children and adults essentially present with optic neuritis and/or transverse myelitis, while encephalitic presentations are more frequent the youngest patients [9]. Differences in the maturation of both the immune system and the CNS between children and adults are suggested to result in different disease phenotypes. In the case series by Jarius et al. up to 30% of patients had attack with brainstem involvement associated with either myelitis, optic neuritis or encephalitis but isolated brainstem lesions were uncommon (1.8%). Asymptomatic brainstem lesions were found in 5 cases [10].

This observation expands the clinical spectrum of MOG-related diseases in adults. Our case illustrates that paroxysmal symptoms may be the initial and only presentation of anti-MOG Ab-related disease. As the disease can induce significant disability, usually as a result of the initial attack, an accurate diagnosis is imperative to provide adequate early treatment. With the advent of specific cell-based assays to detect anti MOG antibodies, we suggest to screen adult patients presenting with an isolated brainstem demyelinating lesion.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical standards

Patient gave informed consent and details that might disclose the identity of the patient have been omitted.

Declaration of Competing Interest

None.

https://doi.org/10.1016/j.jns.2019.08.002
Received 18 May 2019; Received in revised form 30 June 2019; Accepted 1 August 2019
Available online 05 August 2019
0022-510X/

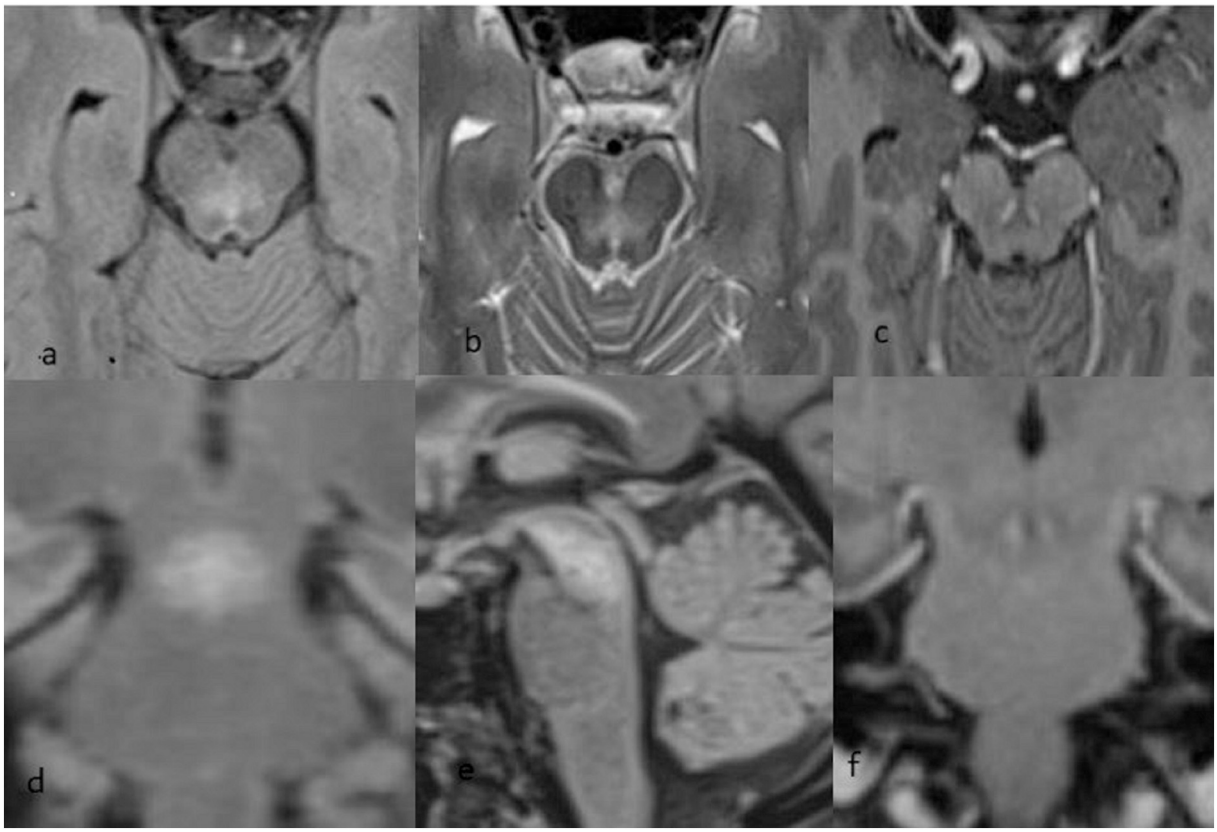

Fig. 1. (a) Axial FLAIR, (b) Axial T2 MR, (d) coronal FLAIR and (e) sagittal FLAIR studies showing bilateral tegmental midbrain hyperintensity, enhancing on axial (c) and coronal (f) T1 + gadolinium sequences upon a follow-up MRI examination at 2 months (c).

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