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Bulbar Symptoms as an Unusual Presentation of Multiple Sclerosis: A Case Report

Martine Kai[1], Christian Matta[2], Georges Jalkh[2], Youmna Abdallah[1], Fady Haddad[1], Halim Abboud[2]

[1] Department of Internal Medicine and Clinical Immunology, Hôtel-Dieu de France, Faculty of Medicine, Saint Joseph University of Beirut, Beirut, Lebanon

[2] Department of Neurology, Hôtel-Dieu de France, Faculty of Medicine, Saint Joseph University of Beirut, Beirut, Lebanon

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ABSTRACT

Multiple sclerosis (MS) is a complex multifactorial disease with different clinical manifestations. Bulbar symptoms such as dysarthria and dysphagia are common in MS patients with advanced secondary progressive disease. However, they are not common at disease onset. We present the case of a 17-year-old male who initially presented with vomiting, dysarthria, and dysphagia. The investigations led to the diagnosis of MS, with an active lesion in the brainstem, more specifically in the area postrema region. Differential diagnoses were eliminated. The patient received intravenous methylprednisolone resulting in amelioration of symptoms. Treatment with fingolimod was started after discharge. The recognition of MS with atypical onsets is important to make an early accurate diagnosis and prescribe appropriate treatment for a disease known to be one of the most common causes of neurologic disability in young adults.

KEYWORDS

Multiple sclerosis, dysarthria, dysphagia, diagnosis, bulbar symptoms

LEARNING POINTS

- Multiple sclerosis can have atypical presentations.
- Bulbar symptoms such as dysarthria and dysphagia can be initial symptoms of multiple sclerosis, although uncommon.
- Clinicians should be able to recognize multiple sclerosis with atypical onsets in order to make an early accurate diagnosis.

INTRODUCTION

Multiple sclerosis (MS) is a complex multifactorial disease involving genetic and environmental factors[1, 2]. It is considered one of the most common causes of neurologic disability affecting young adults[2]. Although usually diagnosed in early adult life, it can also affect the pediatric population[3]. In fact, 3–10% of MS cases are diagnosed before the age of 18 years[4]. The most frequent clinical presentations are optic neuritis and brainstem and spinal cord syndromes[1]. However, the bulbar symptoms of dysarthria and dysphagia are not commonly described as presenting symptoms. We report the case of a 17-year-old male who presented with dysarthria and dysphagia and whose investigations led to the diagnosis of MS.

CASE DESCRIPTION

A 17-year-old male with a history of childhood asthma and iron deficiency anemia complained of epigastric pain with vomiting followed by dysarthria a few days prior to his presentation. He was treated symptomatically in another hospital. The vomiting ceased but the dysarthria

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persisted along with difficulty in swallowing mainly solids but also liquids. He then presented to the emergency department of our hospital. There were no paresthesia, hypoesthesia, muscle weakness, vertigo, headache, but there was slight bilateral blurring of vision without diplopia. He had no similar episodes in the past and his family history was non-contributory.

On physical examination, the patient was conscious, well-oriented, and hemodynamically stable with a blood pressure of 100/50 mmHg and heart rate of 60 bpm. He was afebrile (36.7 °C) and had good oxygen saturation (98%) breathing ambient air. His cardiac, pulmonary, and abdominal examination was normal. Neurologic examination revealed isolated paralysis of the 12th cranial nerve. The rest of the assessment was normal, including a normal sensory, motor and gait examination. Laboratory analysis revealed a normal hemogram, no electrolyte abnormalities, negative C-reactive protein, normal thyroid function, a slight deficit in vitamin B12 (213 pg/mL) and folic acid (1 ng/mL), and normal homocysteine levels (14.9 µmol/L).

Brain magnetic resonance imaging (MRI) revealed multiple supratentorial and juxtacortical white matter lesions as well as periventricular lesions, and to a lesser extent infratentorial lesions involving the cerebellar hemispheres and the area postrema region, the latter presenting contrast enhancement suggestive of active inflammatory demyelinating disease (*Fig. 1*). A cervical MRI scan showed no abnormalities involving the cervical spinal cord. A lumbar puncture was performed and showed the presence of 14 nucleated cells/mm^3, with 90% lymphocytes and 5% neutrophils, normal protein level (0.25 g/L), albumin 0.14 g/L, normal glucose level (4 mmol/L), and IgG index 1.05. Isoelectric focusing showed an oligoclonal type 2 profile in the cerebrospinal fluid (CSF). Serum anti-MOG (myelin oligodendrocyte glycoprotein) and anti-aquaporin 4 (anti-AQ4) were negative. All these elements suggested a diagnosis of MS.

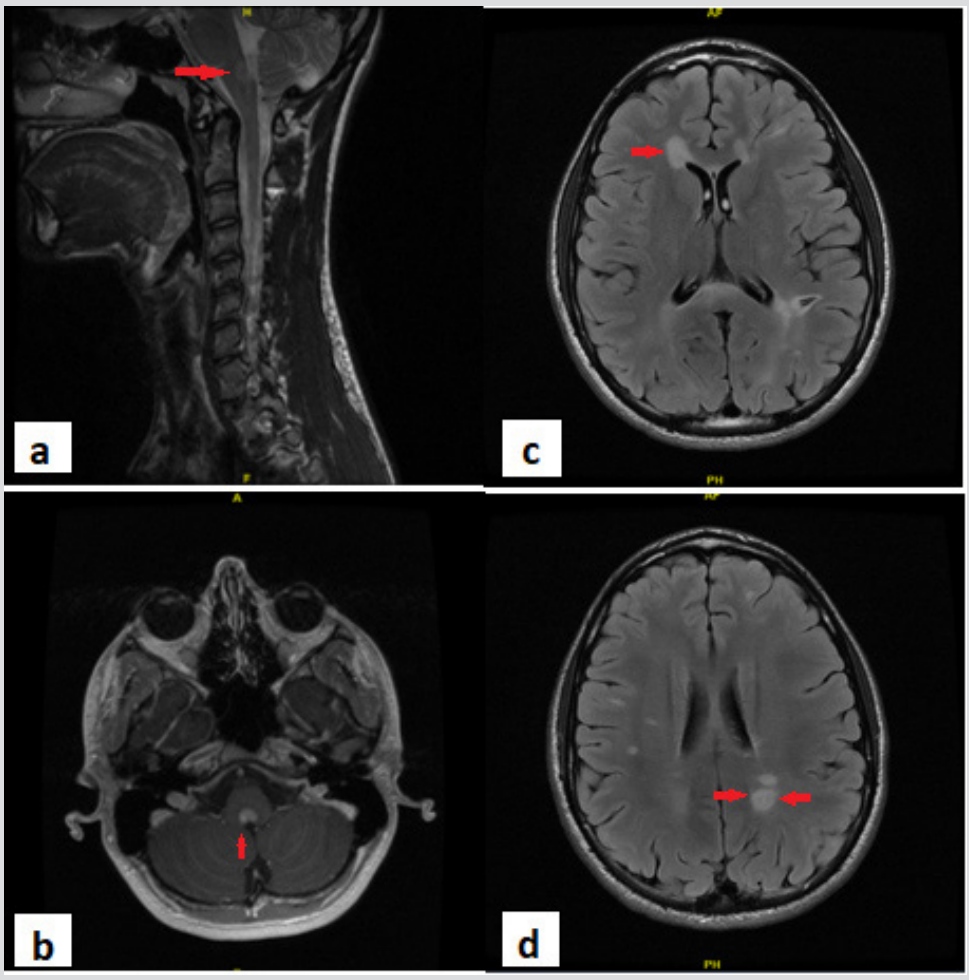

Figure 1. Brain MRI at time of presentation. (a) Sagittal T2 weighted image (T2WI) shows a hyperintense lesion in the area postrema, with enhancement after gadolinium injection in the axial T1 weighted image (T1WI C+). (b) (T1WI C+). (c, d) Axial fluid-attenuated inversion recovery (FLAIR) show hyperintense lesions of the juxtacortical and periventricular white matter

The patient received intravenous methylprednisolone 1 g daily for 5 days, with amelioration of the dysarthria and dysphagia. He was supplemented with vitamin B12 and folic acid and prescribed fingolimod on discharge.

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DISCUSSION

Based on the McDonald diagnostic criteria for multiple sclerosis[5], besides the presence of one clinical attack, the dissemination of lesions in the central nervous system in time and space are required to confirm the diagnosis. In this case, dissemination in time is fulfilled by the presence of CSF-specific oligoclonal bands. Dissemination in space is fulfilled by the presence of multiple asymptomatic lesions of the supratentorial, juxtacortical, and periventricular white matter, as well as symptomatic lesions of the same nature in the area postrema taking the contrast, and therefore reflecting the disease activity (*Fig. 1*). Alternatively, we excluded all other diagnoses such as neuromyelitis optica spectrum disorder (NMOSD) and anti-MOG syndromes, as anti-MOG and anti-AQ4 serum antibodies were negative on evaluation.

MS is known to occur more frequently in females, with a female-to-male ratio of 2–3:1[2]. Concerning pediatric-onset MS (POMS), sex ratios are equal in prepubertal children but the distribution shifts toward female predominance as children age, such as in adult-onset MS (AOMS) [4]. Our patient is a 17-year-old male, considered to be in the transition zone between the two groups; the cut-off age is between 16 and 18 years old, depending on the literature[3-4].

The most common presenting symptoms of MS are visual, sensory, motor, and balance related[6] due to the involvement of the optic nerve, brainstem, cerebellum, and spinal cord[1-2]. Less common are cerebral hemisphere symptoms[1]. Among brainstem and cerebellar symptoms, the most frequently described are diplopia, vertigo, gait ataxia, dysmetria, intentional/postural tremor, facial paresis, and/or hypoesthesia[2]. Cerebellar and brainstem involvement are more common in POMS patients than in AOMS patients[4].

Dysarthria is common in MS patients with advanced secondary progressive disease. However, it is not common at disease onset[7]. The particularity of our case is that the initial manifestation was dysarthria, which has been mainly linked to lesions in the corticobulbar pathway or the cerebellum[8]. Dysphagia is also recognized as a clinical finding in MS[9]. It has been described in more than one-third of MS patients and is more frequently seen in patients with a longer disease course[9]. The physiopathology has been linked to involvement of the corticobulbar tracts, cerebellar and brainstem dysfunction, lower cranial nerve paresis, and cognitive impairment[9].

A symptomatology of dysarthria and dysphagia was described in 2007 in a 34-year-old female patient, with already established relapsing-remitting MS,[10] and attributed to anterior opercular syndrome. Once again, the symptoms occurred during the evolution of the disease and not at onset; the patient had been diagnosed 7 years before with MS, with the first symptom being a left hemiparesis[10]. Her MRI had shown a 'large lesion involving the right perisylvian juxtacortical white matter, which enhanced with gadolinium … [and] a lesion on FLAIR imaging in the left perisylvian region. There were no lesions in the brainstem or cerebellum'[10]. This was not the case with our patient whose MRI showed an active lesion in the area postrema, which was localized in the medulla oblongata. The medulla oblongata also contains the origin of cranial nerves IX, X, XI, and XII[11], which can explain the symptomatology of dysarthria and dysphagia in our patient.

Another syndrome described in the context of MS is paroxysmal dysarthria and ataxia (PDA), in which the brain MRI shows a midbrain lesion involving the crossed fibers in the cerebello–thalamo–cortical pathway[12]. A non-ataxic variant of PDA was described in a 74-year-old man[13] who presented with sudden-onset spontaneous paroxysmal dysarthria also associated with a midbrain lesion, and who improved on high-dose steroids and carbamazepine. No midbrain lesion was found on the MRI of our patient, making this diagnosis unlikely.

Another interesting symptom in our patient was vomiting, which was present at the very onset of the disease. This is a similar finding to that of a case described in 2017 concerning a 10-year-old girl diagnosed with MS, who presented with acutely incoercible nausea and vomiting 1 week before she complained of double vision and instability[14]. The brain MRI of our patient showed an active lesion in the area postrema, which is the emetic reflex center on the floor of the fourth ventricle[15].

CONCLUSION

We presented the case of a 17-year-old male with initial presentation of MS as vomiting, dysarthria, and dysphagia due to a lesion in the brainstem, more specifically in the area postrema region. The recognition of MS with atypical onsets is important in order to make an early accurate diagnosis and prescribe appropriate treatment for a disease known as one of the most common causes of neurologic disability in young adults.

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