

left ventricular hypertrophy with normal systolic function. Aortic prosthetic valve was functional. Neurological evaluation was normal. Electromyography was suggestive of ALS. The patient was discharged home under NIV.

RF as the initial manifestation of ALS is rare and, in such cases, diagnosis is extremely difficult.^{5,6}

The Portuguese group of De Carvalho et al.⁷ described 3 cases of ALS presenting with acute RF that required IMV.

In our cases, other diagnoses were firstly considered. However, inadequate response to treatment, difficult weaning, problems in eliminating secretions and swallowing dysfunction, raised the hypothesis of NMD.

Diaphragmatic weakness, confirmed in the first two patients, was the major cause of RF, as in the cases reported by De Carvalho et al.⁷ Supine evaluation of vital capacity is a highly informative parameter¹ however, this was not assessed in any of the cases reported here which constitutes a limitation of this study. Long term NIV was the solution for one patient. IMV with tracheostomy was the option for the other patient that survived. It is indicated when there is severe bulbar dysfunction and clearing secretions techniques failure.^{8,9} When there is total dependence of NIV, tracheostomy is also an option, depending on patient's wishes.⁸

Diagnosis of NMD in RICUs is uncommon. However, despite no previous neurological history, NMD should be investigated in patients difficult to wean from ventilatory support. A multidisciplinary approach to the clinical, psychological and social factors should be available for these patients.

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Conflict of interest

The authors declare that they have no conflict of interest.

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Central alveolar hypoventilation due to progressive multifocal leukoencephalopathy



Dear Editor,

Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease caused by John Cunningham (JC) virus and occurs in immunosuppressed patients. PML affects mainly the cortical and subcortical white matter, includes neurological symptoms such as ataxia, hemiparesis, visual anomalies and can also be accompanied by behavioural

alterations. PML can also cause lesions in other central nervous system (CNS) areas, such as the brainstem or the spinal cord.^{1,2}

PML may occur in patients with severe brainstem injuries, such as tumours, strokes or infections which might affect the central chemoreceptor zone, causing central alveolar hypoventilation syndrome (CHS).^{5–7}

We report the case of a 27-year-old woman who had undergone a double intestinal-kidney transplantation in 2013. Induction immunosuppression consisted of corticoids, tacrolimus and eculizumab. In 2014 she gradually developed diplopia, nystagmus, ataxia and dysphagia. A magnetic resonance imaging (MRI) showed a bilateral asymmetric

were resolved by switching to a spontaneous-assisted mode so we decided to maintain the NIV in that mode and to discharge the patient. The rest of the study was normal, with an IAH of 15.6 due to those mode changes. The patient was set on long-term home NIV, and hyperventilation symptoms resolved.

PML is a CNS disease that affects particularly immunosuppressed patients, e.g. HIV seropositive patients or following an organ transplant. It is produced by the JC virus which leads to a lytic infection of oligodendrocytes and astrocytes whose neurologic deficits correspond to areas of demyelination in the brain, frequently localized in the subcortical hemispheric white matter or the cerebellar peduncles. The disease can occur in other regions of the central nervous system including the brainstem, the optic nerves or the spinal cord. The PML presentation begins with focal neurologic deficits that depend on the location of the lesions. The diagnosis of the disease requires MRI, performing PCR for DNA JC virus or a brain biopsy.

Currently, there is no specific treatment for JC virus infection, although a few antiviral medications have been studied for treatment of PML. The only treatment available is the restoration of immune function through immunomodulatory therapies. In this case report, eculizumab was replaced by everolimus when PML was diagnosed. Viral load (measured by PCR in CSF) and immune response are significant factors for the prognosis, although more than half of the cases treated have some significant neurological consequences.^{1,2}

According to The International Classification of Sleep Disorders (ICSD), alveolar hypoventilation secondary to medical condition is relatively uncommon.³ CHS can result from neoplasms, strokes, encephalitis and neurodegenerative conditions which can be developed as a result of severe injury or trauma to the brain or brainstem, but there are no reported cases of bulb lesions to PML disease.⁵⁻⁷

Our patient presented persistent brainstem injuries, symptomatic hypercapnia and demonstrated hypoventilation, showing the association between these alterations. Moreover, after management and outcome of negative JC virus PC, repeated episodes of central apnea by RP and PSG were observed. The medulla damage was considered irreversible, so long term NIV is prescribed.^{4,8,9}

This case report reveals the importance of studies for the diagnoses of sleep disorders in patients with medulla infectious lesions. It shows how central apneas may cause central alveolar hypoventilation. Close monitoring of respiratory function and non-invasive ventilation support when it is needed are important in the treatment of these patients in order to improve survival and quality of life.

Conflicts of interest

The authors have no conflicts of interest to declare.

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