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INTRODUCTION

IgLON5 antibodies define a heterogeneous disease manifesting with sleep disorder, chorea, extrapyramidal symptoms, bulbar dysfunction including dysphagia and cognitive decline. In addition, patients can show movement disorders including myoclonus and myorhythmia. Herein, we present a peculiar IgLON5 presentation with mylohyoid muscle myorhythmia leading to a repetitive movement of the throat.

RESULTS

A 61 year-old woman developed one month after Sars-Cov2 infection, lingual and facial paresthesias with unclear distribution, abdominal pain in the lower quadrants and progressive dysphagia.

Brain MRI was unremarkable, and polygraphy with electroencephalographic and electromyographic recordings revealed myorhythmia in the mylohyoid muscle (Figure 1). Lumbar puncture did not show inflammatory signs. A wide panel of neuronal antibodies revealed the presence of IgLON5 antibodies confirmed with both live cell-based assay and tissue-based assay (Figure 2).

The patient received intravenous steroids, immunoglobulins, and plasma exchange (PLEX), with almost no modification of the myorhythmia but improvement of the pain. Given the poor response to first-line treatments, the patient was treated with rituximab, and three months later she exhibited improvement in both pain and myorhythmia, and especially of the dysphagia. However, this improvement was moderate and only captured by a CASE, and not mRS, modification (Figure 3).

Figure 1. Polygraphy with electromyographic recordings revealing myorhythmia in the mylohyoid muscle.

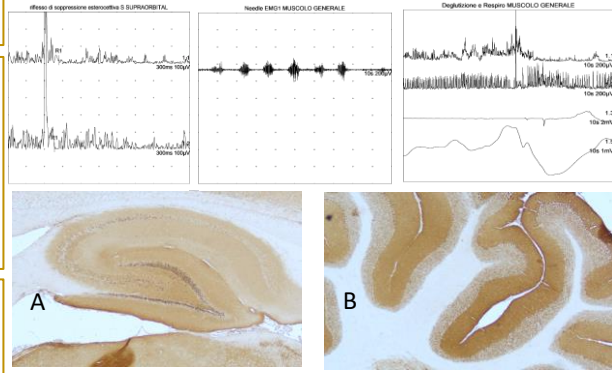


Figure 2. Brain rat IHC of the IgLON5-IgG+ patient. IgLON5-IgG lead to neuropilar staining on the hippocampus (A) and the cerebellum (B).



Figure 4: mylohyoid myorhythmia at initial presentation

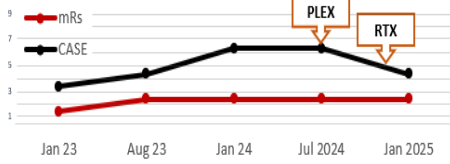


Figure 3. Timeline of the IgLON5-IgG+ patient's clinical progresses

DISCUSSION

Mylohyoid myorhythmia has previously been reported in association with pontine lesions adjacent to the trigeminal nerve nucleus, or as part of a complex laryngeal movement disorder referred to as "dancing larynx syndrome" or "frog like myorhythmia". The etiology is heterogeneous, spanning from Multiple Sclerosis to atypical parkinsonism. The presence of focal myoclonus, for example involving the tongue, has been reported in IgLON5 disease, but no cases of myorhythmia involving specifically the mylohyoid muscle have been described to our knowledge. By illustrating a rare but highly recognizable manifestation, our case provides clues for the early identification of a potentially treatable condition such as IgLON5 disease.

CONCLUSIONS

IgLON5 disease is a rare but potentially treatable differential diagnosis of the «dancing larynx syndrome» or «frog-like» myoclonus/myorhythmia. Screening for IgLON5 antibodies should always be considered when facing this rare neurological presentation.

