CLIPPERS MIMICKING ATYPICAL PARKINSONISM

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Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLLIPERS) is a chronic inflammatory disease of the CNS with complex etiopathogenic mechanisms not fully elucidated. Although CLIPPERS can virtually affect any part of the brain, the pons is most frequently involved, followed by cerebellum, basal ganglia, corpus callosum and spinal cord. Therefore, there is a wide variety of clinical manifestations that entangle diagnosis.

A 67-year-old woman recently diagnosed with PD was admitted to our clinic for subacute dizziness, balance/gait impairment, and episodic binocular horizontal diplopia in primary gaze. She had previously been investigated with non-contrast brain MRI which showed hyperintense T2/FLAIR millimetric lesions in bilateral globus pallidus. Neurological examination revealed mild asymmetrical parkinsonism, global ataxia, bidirectional nystagmus and dysarthria. We performed brain MRI with contrast that identified punctate perivascular gadolinium enhancing lesions supra and infratentorial, with "salt and pepper" appearance in the pons. CSF analysis revealed minimal pleocytosis, hyperproteinorachia and intrathecal IgM synthesis.

The workup for neuroinfections and autoimmune disorders (angiotensin converting enzyme, ANA panel, MOG, GAD and onconeuronal antibodies) was negative. We administered pulse therapy with methylprednisolone, with marked clinical improvement. One year afterwards, she returned for recurrence of symptoms. Brain MRI identified an increase in the number of contrast-enhanced punctate lesions. She received long-term corticotherapy, with sustained clinical response.

We retrospectively established the diagnosis of probable CLIPPERS. Considering the lack of specific biomarkers and the difficulty in performing brain biopsy, we should rely on a high clinical and radiological index of suspicion.