

Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids syndrome: a case report

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BACKGROUND: Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (**CLIPPERS**) is a recently described inflammatory disease of central nervous system with distinct clinical and radiological features. All reported patients present with diplopia and gait ataxia and have similar typical MRI findings with punctuate gadolinium enhancement of the pons and midbrains. Extrapontine involvement seems to be frequent. Although the exact pathology has not been fully elucidated, neuro-pathological findings and clinical response to immunosuppressant therapy suggest an autoimmune or other inflammation mediated pathogenesis.

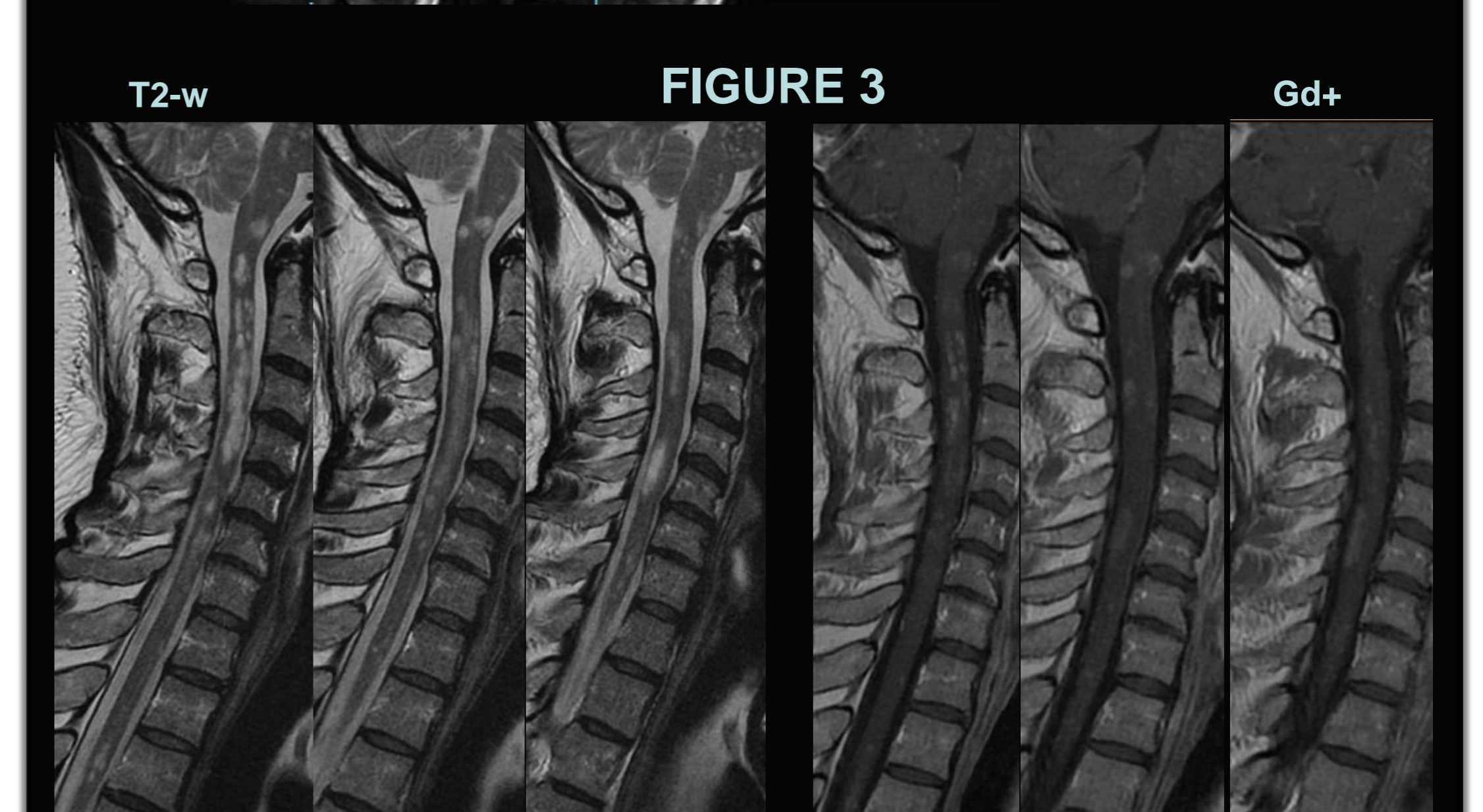
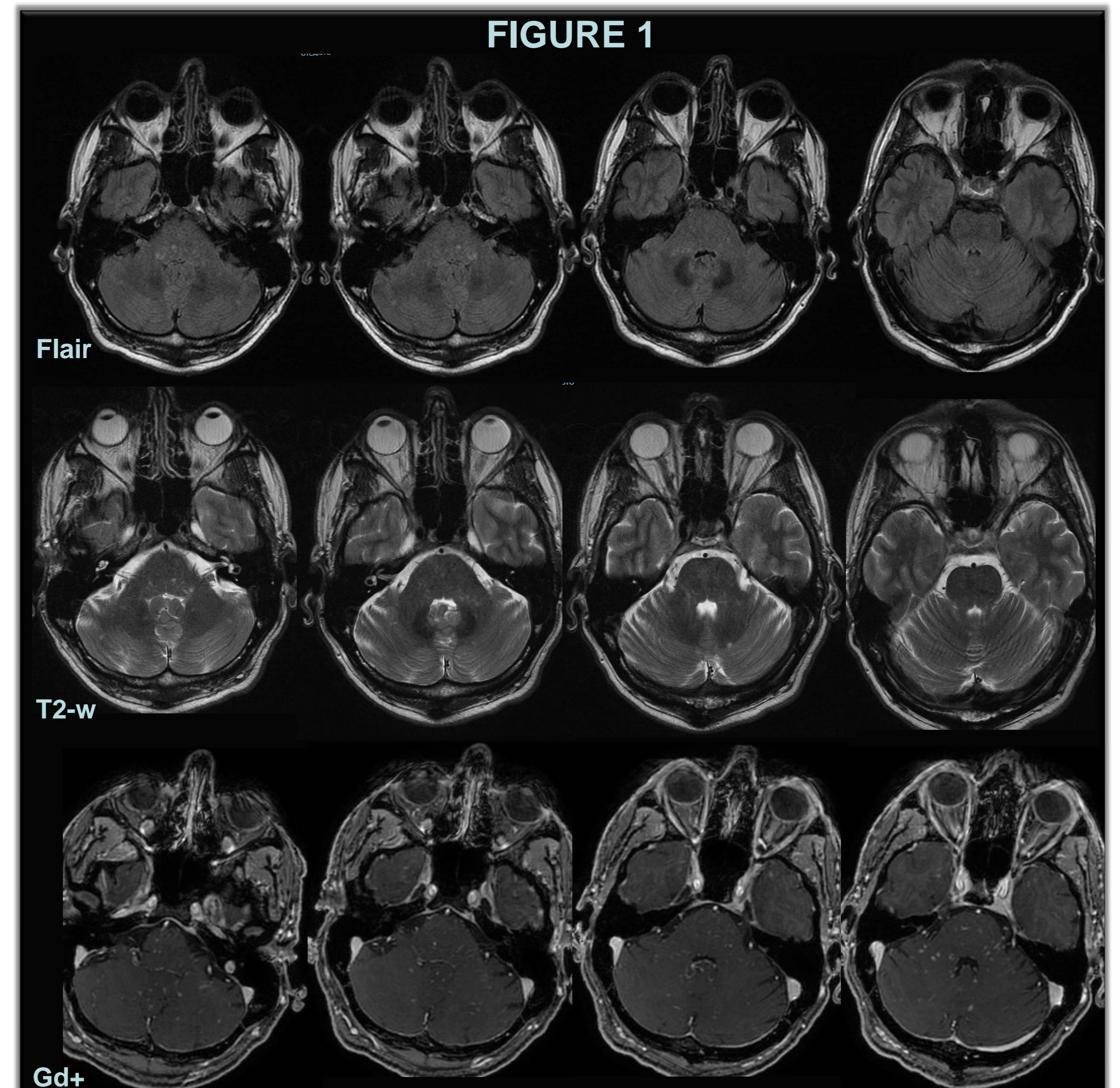
OBJECTIVES: to describe a case of CLIPPERS with severe spinal cord involvement in which steroids led to initial clinical and radiological recovery, but relevant psychic side effects and clinical and radiological relapses forced steroid tapering with initiation of immunosuppressive treatment.

MATERIALS and METHODS: case report.

RESULTS: A 44-year-old male, presented to our emergency room with progressive pancerebellar syndrome followed by gait ataxia and paraparesis.

Magnetic resonance imaging (MRI) disclosed patchy spot like gadolinium enhancement in a "salt and pepper like appearance" in the pons, midbrain and cerebellum [FIGURE 1]. Moreover, severe cervical spinal cord involvement was observed [FIGURE 2]. Cerebrospinal fluid analysis only showed elevated protein levels and positive oligoclonal bands; all other additional investigations were normal. Alternative diagnoses were excluded by means of laboratory and radiological tests. The probable diagnosis of CLIPPERS was made and high dose intravenous corticosteroids were administered, followed by oral steroid maintenance. This led to rapid clinical recovery and decrease of enhancement of lesions on the MRI-scan. Nevertheless, only two months later, we observed a spinal cord relapse [FIGURE 3], which was successfully treated with plasmapheresis. After four months of oral steroid treatment, he started complaining about insomnia and dysphoria, and slowly developed psychotic behavior. A follow up MRI detected again patchy inflammation in the same areas of the spinal cord. Due to the number of relapses and the concomitant side effects of steroids, intravenous pulse cyclophosphamide therapy was started with steroid tapering and subsequent disease stabilization.

DISCUSSION AND CONCLUSIONS: Clippers is still an exclusion diagnosis and its pathogenesis and long-term management are still an enigma. If CLIPPERS is a disease entity or just an immunological response to a disease elsewhere has to be determined. Steroids lead to a considerable initial response but the well-known side effects as well as insufficient response to lower doses might limit its use in the long-term; therefore, other immunosuppressant may be needed to prolong clinical and MRI recovery.



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