

The Evaluation and Treatment of Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS)

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Clinical Presentation - Visit 1

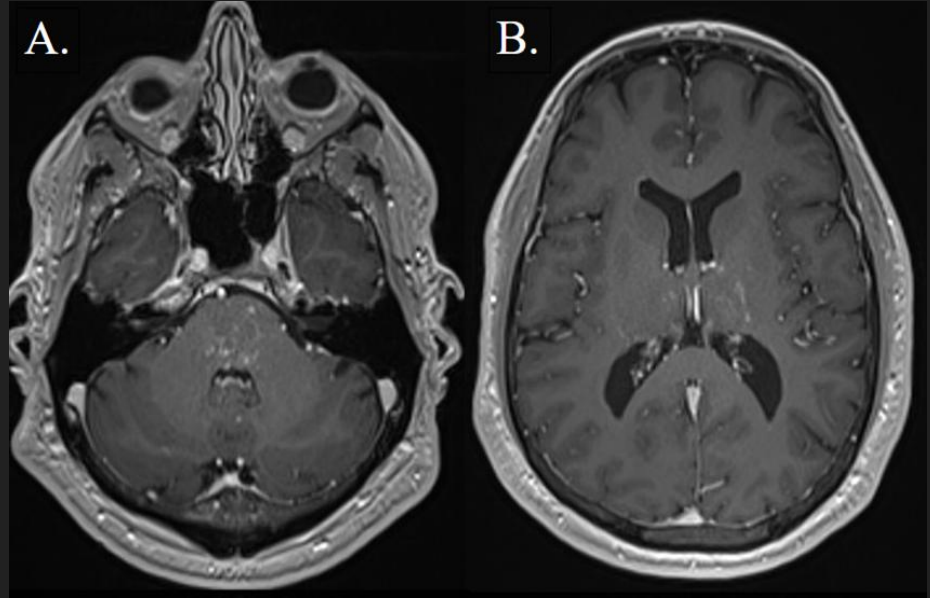
- 53 year old male with no significant PMH presents to the outpatient neurology clinic for one month of facial and bilateral upper extremity paresthesias.
- Neurology physical examination unremarkable aside from the above chief concern.
- Initial MR Brain w/o IV contrast demonstrated abnormal T2 prolongation within the periventricular white matter and brainstem
 - Nonspecific, may represent sequelae of chronic microvascular ischemic changes, infectious etiologies, inflammatory etiologies, or demyelinating disease

Clinical Presentation - Visit 2

- A few weeks later patient returned to the neurology clinic with new diplopia.
- Neurology physical examination now demonstrated new decreased left eye adduction
- Outpatient lumbar puncture demonstrated elevated T cell markers on flow cytometry but otherwise was within normal limits. All other laboratory testing was also within normal limits.
- Subsequently, contrast enhanced MR Brain and Orbits was performed

Imaging Discussion

- T1-weighted contrast-enhanced axial images through the pons (Figure A) and the basal ganglia (Figure B) demonstrate irregular nodular foci of gadolinium enhancement through the pons, bilateral basal ganglia, and thalami

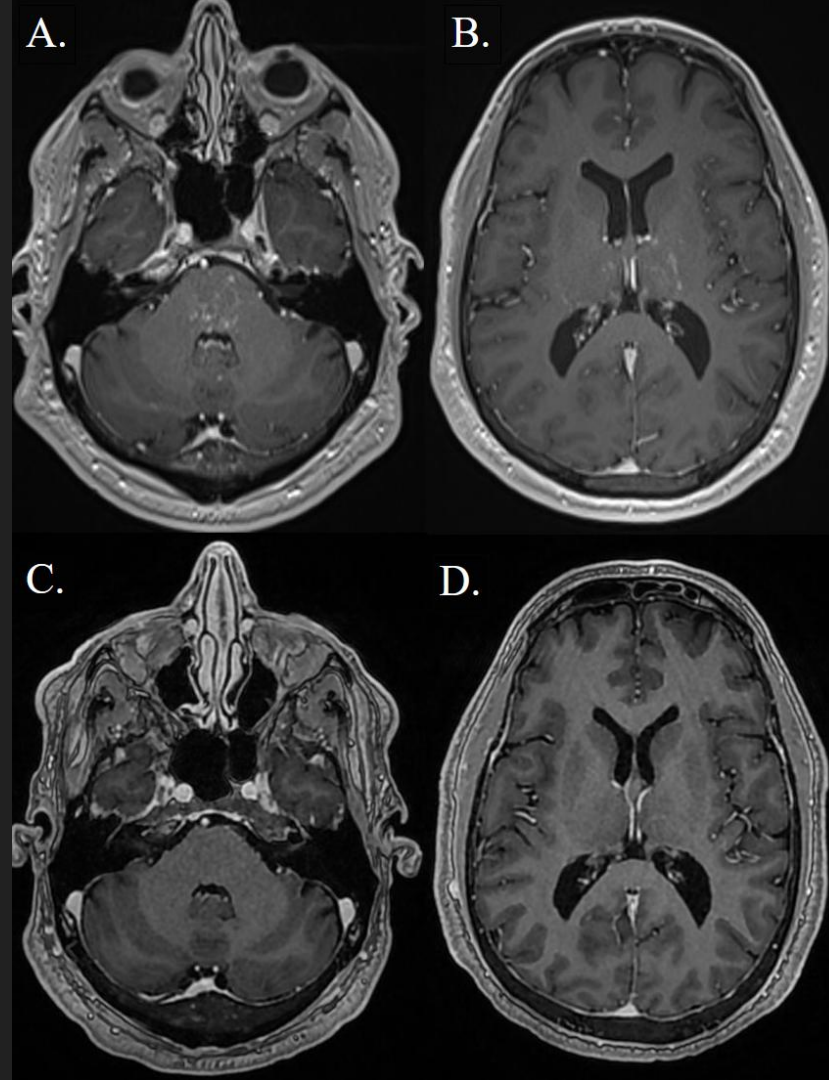


Diagnosis and Management - CLIPPERS

- Given the extent of irregular nodular enhancement throughout the pons, clinical symptomatology, and CSF findings, the primary differential diagnosis was chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS)
 - T-cell mediated encephalitic condition which typically involves the pons and has been described to spread via the corticospinal tracts to supratentorial structures including the basal ganglia, thalami, and capsula interna.
 - Symptomatology includes ataxia, diplopia, and facial paraesthesia.
- Treatment was initiated with intravenous methylprednisolone with gradual seven week taper.

Post-Treatment Imaging Discussion

- Post-treatment T1-weighted contrast-enhanced axial images through the pons (Figure C) and the basal ganglia (Figure D) which demonstrate interval near resolution of irregular nodular foci of gadolinium enhancement through the pons, bilateral basal ganglia, and thalami.
- Pre-treatment imaging (Figures A and B) provided for comparison



Clinical Outcome

- Over the course of the patient's steroid taper, their symptomatology slowly improved.
- Subsequent MR Brain with IV Contrast status post treatment demonstrated near complete resolution of the previously described regions of enhancement.
- Follow-up MR Brain approximately one year later demonstrated complete resolution of abnormal enhancement.

Conclusions and Take Home Message

- CLIPPERS is an encephalitic condition which typically involves the pons and has been described to spread to supratentorial structures including the basal ganglia, thalami, and capsula interna.
- Symptomatology of CLIPPERS includes ataxia, diplopia, and facial paraesthesia.
- In the appropriate clinical and radiologic setting, it is crucial to provide CLIPPERS within the differential diagnosis as treatment with intravenous steroids is generally curative.

References

- Dudesek A, Rimmele F, Tesar S, et al. CLIPPERS: chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids. Review of an increasingly recognized entity within the spectrum of inflammatory central nervous system disorders. Clin Exp Immunol. 2014;175(3):385-396. doi:10.1111/cei.12204
- Axelerad AD, Stroe AZ, Mihai C, et al. CLIPPERS, chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids: A challenge in neurological practice, clinical landmarks (Review). Exp Ther Med. 2021;22(4):1191. doi:10.3892/etm.2021.10625