

Clinical Characteristics, Management, and Outcomes of CLIPPERS: A Comprehensive Systematic Review of 140 patients from 100 Studies

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Introduction: Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) is a rare inflammatory disorder of the central nervous system, characterized by symptoms referable to the brainstem and cerebellum such as, diplopia, gait ataxia and cerebellar dysarthria. The features and outcomes of CLIPPERS remains uncertain. Wwe conducted this comprehensive systematic review to summarize the existing studies that described CLIPPERS in the literature and to provide a quantitative assessment on the clinical characteristics, management, and outcomes of this rare syndrome.

Methods: A search of PubMed and Web of Science databases was conducted from inception until January 15, 2022, was conducted. We included the cases that reported probable or definite diagnosis of CLIPPERS based on Taieb et al.'s criteria. The quality of the included studies was assessed using the JBI Critical Appraisal Tool. Descriptive statistics were performed to analyze the studies.

Results: We identified 140 patients with CLIPPERS (mean age: 46±18 years and males were 60%). The average follow-up duration was 32.27±57.8 months. Ataxia was the most common presenting symptom. Sixteen percent of the cases were associated with malignancy, mostly hematologic malignancies. The overall relapse rate was 59.2%, and the duration of steroid therapy was considerably shorter in the relapsed cases than in the non-relapsed (mean 6.19±7.9 vs. 10.14±12.1 days, respectively, P=0.04). The overall mortality rate was 10%, but mortality in patients with malignancy was 30% and it was 12% in patients with relapses.