WHAT'S HAPPENING IN

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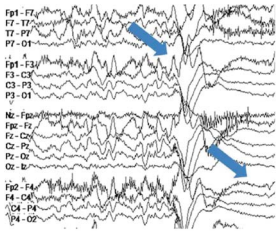
Abstracts

Articles appearing in the October 2018 issue

Late-onset Lennox-Gastaut syndrome: Diagnostic evaluation and outcome



Background We describe the clinical features and outcome in patients with late-onset Lennox-Gastaut syndrome (LGS).

Methods Adult patients evaluated between January 1, 2000, and March 1, 2017, who presented with onset of LGS ≥10 years, were identified. Data abstracted included age at seizure onset, seizure types, etiology, treatments, EEG and neuroimaging results, CSF findings, and autoimmune evaluation.

Results Ten patients (8 female) were identified. The mean age at onset of seizures consistent with LGS was 16.5 years (range 10–32 years). Seizure types included tonic, atonic, and tonic-clonic seizures (all), myoclonic seizures (n = 3), and atypical absence seizures (n = 7). Five patients had normal intellectual function at onset. Prolonged video-EEG monitoring recorded seizures and generalized interictal epileptiform discharges in all. All patients had drug-resistant epilepsy (range of antiseizure drugs tried, 7–16). Two patients had a history of intrathecal methotrexate to treat acute lymphoblastic leukemia. Two patients had malformations of cortical development. CSF analysis (n = 5) showed a mild elevation in the protein level without other abnormalities. Autoantibody determinations in the serum (n = 4) or the CSF (n = 5) and genetic testing (n = 5) were negative. At final follow-up, all but 1 patient was disabled and required a caregiver, and none was driving. One patient died of probable sudden unexpected death in epilepsy.

Conclusions Late-onset LGS represents a rare, treatment-resistant generalized epilepsy that is disabling and may be associated with progressive cognitive impairment. The elevated CSF protein level in our cohort could have been due to high seizure burden but increases the possibility of an inflammatory component to the pathophysiology of this disorder.

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Editorial

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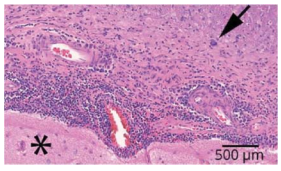
Rheumatoid meningitis: A rare cause of aseptic meningitis with frequently stroke-like episodes



Background Rheumatoid meningitis (RM) is a rare manifestation of rheumatoid arthritis (RA) and may present with stroke-like episodes. We describe diagnostic findings and the outcome in patients with RM.

Methods We identified 6 patients with RM in different stages of RA mostly admitted with stroke-like episodes or common features of meningitis. We used MRI, CSF, and histology for in-depth characterization.

Results We observed RM in 2 patients without history of RA, 1 patient with early seropositive RA, and 3 patients with late-stage RA. Recurrent stroke-like episodes occurred in 5 of 6 patients; headache and partial status epilepticus was in the foreground in 1 patient. Symptoms were accompanied by constitutional symptoms in all patients. MRI showed leptomeningeal or pachymeningeal fluid-attenuated inversion recovery hyperintensities with contrast enhancement. CSF mostly showed mild pleocytosis but can initially be normal. Anticitrullinated peptide antibodies (ACPA) and rheumatoid factor (RF) were positive in all patients. Histopathology revealed granulomatous inflammation in 2 patients. Response to steroids was prompt and further immunosuppressive treatment prevented recurrence.

Conclusions RM is a rare manifestation of RA and often presents with stroke-like episodes. It is currently not implemented in the workup of aseptic meningitis in national guidelines. Crucial clues for diagnosis included recurrent stroke-like episodes refractory to antiepileptic treatment, headache and constitutional symptoms, meningeal enhancement on MRI, CSF pleocytosis, and positive serology findings for ACPA and RF. Prognosis is favorable with early immunosuppressive treatment.

NPub.org/NCP/9306c

Practice Current

Neurology® Clinical Practice has launched its next Practice Current survey on a universally challenging topic: "How do you diagnose and treat post-concussive headache?" Please consider completing the survey to add your own perspective. In the June 2019 issue, readers will have access to opinions from David W. Dodick, MD (US), Mohammad Wasay, MBBS, MD, FRCP (Pakistan), and Karen M. Barlow, MSc, MBChB, MRCPCH, RACP (Australia).

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