

PERIPHERAL NERVE HYPEREXCITABILITY SYNDROME – LONGITUDINAL FOLLOW UP OF THREE PATIENTS

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Introduction: Peripheral Nerve Hyperexcitability (PNH) is characterized by continuous muscle fiber activity (CMFA). PNH is the chief manifestation of Isaacs' syndrome and cramp-fasciculations syndrome (CFS). Antibodies to voltage gated potassium channels (VGKC-Ab) can be present. We report longitudinal observation of three patients.

Case presentations: A 35-year-old female with mixed connective tissue disorder treated with hydroxychloroquine and prednisone, experienced muscle twitching, stiffness, cramps, delayed muscle relaxation, numbness in toes and difficulties walking. EDX testing showed generalized CMFA, (fasciculations, myokymic discharges, neuromyotonia) and axonal neuropathy. VGKC-Ab were negative. Anti-CASPR2 antibody was equivocal. Plasmapheresis and IVIG improved her walking. Long term treatment with carbamazepine was effective in alleviating the involuntary movements. Her function status remained improved on a three year follow-up.

Case 2: A 65-year-old male one year following resolved Guillain-Barré syndrome developed new symptoms of progressive generalized cramps, muscle twitching and difficulties walking on an incline. EDX revealed generalized CMFA compatible with CFS. VGKC-Ab were positive. Gabapentin, but not carbamazepine alleviated the symptoms. Three year follow-up showed continuous normal functional status.

Case 3: 67-year-old presented with four years history of cramps and involuntary muscle twitching in the calves. EDX revealed CMFA compatible with CFS including afterdischarges. There were no identifiable comorbidities. VGKC-Ab were negative. Brief trial of gabapentin and later of carbamazepine provided no satisfactory relief and the patient declined further treatment. His neurological status did not change over the course of ensuing two years.

Conclusions: PNH syndrome is a heterogeneous disorder. Longitudinal follow-up of our three patients showed good prognosis.