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Paroxysmal sympathetic hyperactivity in Juvenile neuronal ceroid lipofuscinosis (Batten disease)

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Abstract

Paroxysmal sympathetic hyperactivity (PSH) is a clinical syndrome of agitation and involuntary motor activity that particularly occurs in patients with severe acquired brain injury. The aim of the present study is to substantiate the assertion that paroxysmal non-epileptic attacks resembling PSH also occur in patients with Juvenile Neuronal Ceroid Lipofuscinosis (JNCL, Batten disease), which is the most common neurodegenerative disease in children.

The paper describes a case series of five patients with JNCL which during a period of fifteen years have been followed clinically and by consecutive investigations of the autonomic nervous system using heart rate variability (HRV) investigations. Following adolescence a significant autonomic imbalance with very low parasympathetic activity and an unchanged high sympathetic excitatory activity was documented. In addition, episodes of anxiety and agitation combined with involuntary movements were reported. Beyond the frightened facial expression and involuntary increased motor activity, excessive sweating, increased body temperature, high heart and respiratory rates were reported, and typically, the episodes occurred to stimuli that were either non-nociceptive or only minimally nociceptive. Thus, from a clinical point of view the non-epileptic paroxysmal condition with anxious behavior, agitation and motor hyperactivity seen in patients with JNCL fits to the clinical description of PSH which normally occurs following acutely acquired brain injury, and as the neuropathological basis in JNCL for development of PSH is similar to what is seen in patients with traumatic brain injuries, it seems reasonable to propose that PSH also occurs following adolescence in patients with JNCL.

Keywords

Batten disease, Non epileptic seizure, Neuronal ceroid lipofuscinosis, Paroxysmal sympathetic hyperactivity