عنوان مقاله:

Symptomatic Hyperekplexia, Pseudoathetosis as Presenting Symptom of Multiple Sclerosis

محل انتشار:

شانزدهمین کنگره بین المللی ام اس (سال: 1398)

تعداد صفحات اصل مقاله: 2

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خلاصه مقاله:

Introduction: Hyperekplexia or startle disease is characterized by an exaggerated, abnormal startle response and a type of generalized myoclonus, consisting excessive motor response to unexpected auditory somesthetic or visual stimuli. Hyperekplexia are three types: familial as autosomal dominant, sporadic and secondary or symptomatic. Symptomatic hyperekplexia are due to local brainstem pathology as may be seen with anoxia, hemorrhage and multiple sclerosis (MS).In spinal multiple sclerosis (MS) patient may present with acute proprioceptive sensory impairment and pseudoathetosis in limbs. Pseudoathetosis or piano playing movements of hands, refer to a movement disorder that consisting of involuntary slow writing movements of fingers and indicates disruption of the proprioceptive pathway, from peripheral never to parietal cortex. Causes of pseudoathetosis are stroke, myelitis, vitamin B12 deficiency, spinal cord infarct, trauma and multiple sclerosis (MS). Pseudoathetosis clinically indistinguishable from true athetosis that caused by lesion in basal ganglia. In true athetosis there is no proprioceptive sensory loss and versus pseudoathetosis abnormal movements are no pronounced when the eyes closed.Case I: A 28 year- old woman presented with a 1- week history of excessive startle to sudden or closing door, she had previously never experienced such symptoms and there was no family history of exaggerated startle. One year ago she first had paresthesias in the lower legs and then developed a left optic neuritis two months later. By that time a diagnosis of MS had been made based on the clinical history, detection of oligoclonal bands in the CSF, and characteristic T2 and gadolinium- enhancing lesions on cranial MRI. Routin blood tests were unremarkable and for rule out secondary causes of hyperekplexia, angiotensin converting enzyme, borrelia burgdoferi, IgG and IgM antibodies in serum and CSF evaluated and were negative, on examination sudden acoustic, visual and tactile stimuli elicited an exaggerated and prolonged startle reaction, which consists of a brisk generalized myoclonic jerk with grimacing, shoulder elevation, arm flexion and extension of spine. The reaction rattern was not habitable and became worse with heightened expectation of the stimuli. The excessive startle was no response to clonazepam 3 mg for 5 days but sever hyperekplexia completely disappeared after treatment of high dose methylprednisolon and gabapentin.Conclusion: In patients with exaggerated startle response without no family history and no mutation in ∝1 ... subunit of the inhibito

کلمات کلیدی:

Hyperekplexia, Pseudoathetosis, Multiple sclerosis