Athetosis

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Glossary

- Athetosis: Sinuous, slow, irregular, and involuntary movements affecting distal limbs.
- Proprioception: Sense of position of joints.
- Pseudoathetosis: Writhing finger and/or toe movements due to loss of proprioception.

Definition and History

The term athetosis (‘without fixed posture’) describes sinuous, slow, irregular and involuntary movements affecting distal limbs, particularly the arms. The term was coined by Hammond in the late nineteenth century, when describing a movement disorder in an alcoholic patient with onset following an episode of delirium tremens. Shortly afterward, Shaw also used the word athetosis to describe a similar sinuous movement disorder in a patient with cerebral palsy. The term has also been applied to movements seen in subjects with dysfunction of proprioception, but because the movements are a consequence of sensory abnormalities, this syndrome is called ‘pseudoathetosis.’ This latter term remains in the current medical literature, despite the sharp decline of the use of the term ‘athetosis’ in the last decades. The reason behind this decline is the realization that athetosis is better defined as dystonia occasionally associated with some degree of chorea. There are authors, however, who continue to advocate the usefulness of athetosis as a distinct type of hyperkinesia.

Etiology and Pathogenesis

Athetosis is usually associated with cerebral palsy caused by kernicterus, that is, lesions of the central nervous system due to severe jaundice in the new born. However, cerebral palsy of any etiology may cause this movement disorder. In the few patients who came to autopsy, there are reports of lesions in the putamen or caudate. (Please refer to the entry on Dystonia in this Encyclopedia for further details on the pathogenesis of athetosis).

In pseudoathetosis, the movements are thought to result from the inability of the fingers or toes to remain still because of the loss of proprioception. Traditionally, this movement disorder is associated with peripheral neuropathy. However, it may also result from central nervous system lesions causing impairment of sense of position. There are many reports of pseudoathetosis caused by lesions of spinal cord and, less commonly, thalamus. The causes listed in these reports are vascular lesions, B12 vitamin deficiency, and syringomyelia.

Epidemiology

There are no epidemiological studies of athetosis. It is possible to speculate, however, that it is becoming a much rarer condition nowadays. The main reason for the decline of its frequency is the improvement of obstetric care with the reduction of the number of cases of cerebral palsy. Traditionally, pseudoathetosis is considered as a rare condition. It is uncertain, however, if this is true since many patients may remain without diagnosis because of the lack of functional impairment caused by this movement disorders.

Clinical Features and Diagnostic Criteria

Athetosis is characterized by sinuous, slow, irregular, and involuntary movements affecting distal limbs, especially the arms. The phenomenology is usually characterized by a combination of dystonia and chorea, but myoclonus and spasticity are also common. Similarly, pseudoathetosis is characterized by slow, distal, writhing movements of the fingers or toes, which tend to worsen with the suppression of visual input. Invariably, these patients have proprioceptive sensory loss and often a Romberg sign. There are no formal diagnostic criteria for these conditions. However, a history of perinatal injury in athetosis and peripheral neuropathy or central loss of proprioception in pseudoathetosis are highly useful to make the diagnosis.

Differential Diagnosis

Athetosis and pseudoathetosis should be distinguished from other hyperkinetic movement disorders affecting distal limbs. Chorea is unpredictable; myoclonus has a shock-like nature, with a brief duration, usually less than 200 ms; and tremor is characterized by its rhythmic and oscillatory nature. The most important treatable condition with progressive athetosis is Wilson’s disease, due to abnormal copper metabolism.
Diagnostic Work-Up

The investigation of patients with athetosis should follow the guidelines to work up patients suspected to harbor a secondary dystonia. Of note, all patients should undergo tests for Wilson's disease (serum ceruloplasmin, serum copper, 24 h urine copper, liver function tests, and search for Kayser–Fleischer ring) and magnetic resonance imaging of the brain. Focused attention should be placed on the basal ganglia. In patients with pseudoathetosis, electromyography and, in case it is normal, magnetic resonance imaging of the spinal cord and even the brain are useful.

Management

No treatment is required for pseudoathetosis since it does not cause significant functional impairment. The situation is different, however, in athetosis where many patients are significantly disabled by the dyskinesia. Unfortunately, as a rule it is often resistant to all available therapeutic options. A few patients may improve with levodopa, clonazepan, baclofen, anticholinergics, or tetra-benzine. Most of them, however, are refractory to clinical treatment and even surgical treatment has not been successful. There are recent reports describing failure or poor results of baclofen pump or deep brain stimulation of the globus pallidus internal in alleviating dystikinesias in patients with cerebral palsy.

Prognosis

As a manifestation of static encephalopathy, athetosis usually remains stable along the time. However, as well described in the literature of cerebral palsy, some patients may develop worsening of the movement disorder at later age. The course of pseudoathetosis depends on the underlying cause: if there is improvement of the proprioception, the movement disorder may also decrease.

See also: Wilson's disease (430).

Further Reading


Relevant Websites

www.movementdisorders.org – Movement Disorder Society
Abstract:

Athetosis is a disorder with sinuous, slow, irregular, and involuntary movements affecting distal limbs. Despite the decline of its use, the term is still employed to describe the distal limb dystonia in cerebral palsy as well as writhing finger and/or toe movements due to loss of proprioception (pseudoathetosis). Occasionally, athetosis is seen as a long-term effect of a cerebrovascular stroke (posthemiplegic athetosis). In some cases, athetosis may respond to pharmacological therapy, but athetosis in cerebral palsy is usually refractory to therapeutic measures. Pseudoathetosis is not associated with functional impairment and does not require specific treatment. In all patients with progressive athetosis, the diagnosis of Wilson's disease should be considered.

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