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Atypical Parkinsonism: Classification and Clinical Features

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Study Description

There are a number of degenerative causes of atypical Parkinsonism, including progressive supranuclear palsy, multiple system atrophy and corticobasal degeneration. The main difficulty with these conditions is that they can easily be confused with Parkinson's disease and there are almost no reliable diagnostic tests to tell them apart. Often the diagnosis will become clearer with time as most of these atypical Parkinsonism conditions respond poorly, if at all, to levodopa are more rapidly progressive than Parkinson's disease and have additional symptoms or signs not usually associated with Parkinson's disease. However, differentiation of the atypical conditions themselves is also very challenging [1-3].

Progressive supranuclear palsy

Progressive Supranuclear Palsy is a degenerative disease characterized by symmetrical Parkinsonism, cognitive changes, a characteristic supranuclear palsy of vertical gaze, early falls, dysarthria and dysphagia. The prevalence of Progressive Supranuclear Palsy is approximately 5 per 100,000. Men and women are equally affected. Average age at onset is 63 and mean time from symptom onset to death is 7 years. No proven cases have begun before 40 and only one before age 45. Progressive Supranuclear Palsy is almost always sporadic and only a handful of familial cases have been reported. Progressive Supranuclear Palsy is a 'tauopathy'-a disorder associated with abnormal aggregates of tau protein. Progressive Supranuclear Palsy is characterized pathologically by degeneration of several subcortical structures including the substantia nigra, the subthalamic nucleus and the midbrain; neurofibrillary tangles are present in these areas. The clinical features include symmetrical Parkinsonism, usually without tremor with prominent neck dystonia leading to stiffness and retrocollis. Slowed vertical saccades progressing to vertical supranuclear gaze palsy. The doll's eye manoeuvre in Progressive Supranuclear Palsy is normal. Note that many older people have a limitation of up-gaze. This is not usually pathological, but a limitation of down-gaze is always abnormal. In last-stage Progressive Supranuclear Palsy eyes may become relatively fixed and there may be little or no correction with doll's eye manoeuvre. Postural instability leading to early falls. Current clinical research criteria for the diagnosis of probable Progressive Supranuclear Palsy insist that falls occur within the first year of symptoms, but many cases start to fall later than this.

There are no diagnostic tests for Progressive supranuclear palsy, and it remains a clinical diagnosis. There is no treatment that alters the course of the disease wither. Supportive features on investigation include MRI findings of atrophy of superior cerebellar peduncles, midbrain atrophy, high signal in midbrain, atrophy or signal increase in the red nucleus, globus pallidus and the absence of cardiovascular autonomic dysfunction. Supportive treatment, particularly regarding swallowing and preventation of falls, prolongs survival and improves quality of life. A trail of levodopa and amantadine is worthwhile [4,5].

Multiple system atrophy

Multiple system atrophy is a degenerative disorder characterized by Parkinsonism, and/or cerebellar signs and autonomic failure. The name multiple system atrophy was coined in 1969, unifying less than one name Shy-Drager syndrome, olivopontocerebellar atrophy and some causes of sporadic striatonigral degeneration. Multiple system atrophy was then subdivided into Multiple System Atrophy-P (Parkinsonism predominant) and Multiple System Atrophy-C (cerebellar signs predominant). The prevalence of the disease is about 4 per 100,000. The typical age onset is 55-60. Onset prior to age 30 has never been reported. All cases are sporadic; familial Multiple System Atrophy has never been documented. Males and females are equally affected. Two-thirds of patients have MSA-P; one-third has MSA-C. Survival times for both types are similar; mean of 6-9 years from symptom onset. Functional disability generally occurs earlier in MSA-P. The pathological hallmark of Multiple System Atrophy is glial cytoplasmic inclusions containing α-synuclein, mainly found in the basal ganglia, cerebellar structures and motor cortex.

In MSA-P, characteristic features are Parkinsonism with signs of autonomic failure: postural drop in blood pressure of more than 20 mmHg systolic/10 mmHg diastolic, urinary incontinence or incomplete bladder emptying, erectile dysfunction. In MSA-C, cerebellar signs occur with autonomic failure. During disease progression, cerebellar signs often develop in patients with MSA-P and Parkinsonism develops in MSA-C. A number of other clinical features occur in Multiple System Atrophy that is helpful in distinguishing it from other parkinsonian conditions. These may be present some years prior to the onset of Parkinsonism or cerebellar signs. These 'red flags' include Early falls and postural instability, dystonia affecting the orofacial muscles occurring spontaneously or when levodopa treatment is initiated, Dystonia affecting the trunk and neck leading to anterocollis and flexion of the trunk and emotional incontinence: inappropriate crying/laughing. Clinical features that would tend to exclude the diagnosis of MSA include dementia, hallucinations, positive family history, gaze paresis.

Cortico-basal degeneration

Cortico-basal degeneration was first described by Rebeiz and colleagues as a syndrome of progressive slowness and stiffness in the limbs, dystonia, numbness or 'deadness' of the affected limbs and gait disturbance. The condition has gone by a number of names in the past including cortico-nigral degeneration with neuronal achromasia, cortical degeneration with swollen chromatolytic neurons and cortico-basal ganglionic degeneration. Cortico-basal degeneration is not just a movement disorder; Clinical criteria for the diagnosis of Cortico-basal degeneration have mainly resulted from analysis of clinical features of

patients referred to movement disorder clinics. However, pathologically proven cases of Cortico-basal degeneration have been reported with an almost entirely 'non-movement' disorder presentation, usually including prominent dementia. Therefore the current understanding of the clinical features of Cortico-basal degeneration is changing, although at the present time the diagnostic criteria for Cortico-basal degeneration remain focused on the movement disorder symptoms and signs.

Prevalence of the disease is unknown, but considerably less than that of PSP or MSA. Males and females are equally affected. Average age at onset is 63, with youngest age at onset reported as 45. Average time from symptom onset to death is 8 years. Cortico-basal degeneration is almost always a sporadic condition, although a few familial cases with pathological confirmation have been reported. Cortico-basal degeneration is characterized by widespread deposition of hyperphosphorylated tau protein in the brain. There is marked neuronal degeneration in the substantia nigra and the fronto-parietal cortex. The clinical features include motor symptoms of patients typically present with asymmetrical rigidity and brady kinesia affecting one limb. Progressive dystonia often also affects the limb. Patients with Cortico-basal degeneration may have difficulty in initiating saccades, but once initiated, they are of normal velocity. The main abnormality is usually in horizontal sacces, whereas vertical saccades are often normal. These points are useful in differentiating Cortico-basal degeneration from PSP. Apraxia is a cardinal clinical feature of Cortico-basal degeneration. It can be difficult to demonstrate in the more affected arm because of sever bradykinesia, rigidity and dystonia. Dementia was previously thought to be rare in Cortico-basal degeneration, but is increasingly recognized as an important part of the phenotype. It may be the presenting or predominant clinical feature, neuropsychometry typically reveals frontal executive defects together with parietal lobe dysfunction. In contrast with Alzheimer's disease, episodic memory is usually well preserved. Treatment options for motor symptoms are very limited. A trial of levodopa is worthwhile, but rarely affective. Amantadine may be helpful for Parkinsonism and gait disturbance. Valproate or levetiracetam may help myoclonus.

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