information

ATYPICAL PARKINSONISM

Parkinson's is a progressive neurological condition, which is characterised by both motor (movement) and non-motor symptoms.

Idiopathic Parkinson's is typically a gradual progressive neurological condition which is primarily related to a lack of dopamine, due to degeneration of dopamine producing neurons, and aggregation of the protein alpha-synuclein. The aggregation of this protein in the form of Lewy Bodies is found in the brain at post mortem.

There are several other neurological conditions which resemble Parkinson's, however they have a poor response to Parkinson's medications and are more rapid in their progression. These are known as Atypical Parkinsonism. These are:

- Multiple System Atrophy
- Progressive Supranuclear Palsy
- Cortico Basal Degeneration
- Lewy Body Dementia

Multiple System Atrophy (MSA)

MSA has been called many things over many years including olivopontocerebellar atrophy, striatonigral degeneration and Shy-Drager syndrome. In 1969 there was a general consensus to use the term MSA.

MSA is a sporadic progressive neurological condition which has Parkinsonian symptoms of bradykinesia, rigidity, with early autonomic nervous system involvement.

MSA is relatively rare and it is currently estimated that in Australia there are approximately 1,200 people with this condition.

Clinical symptoms usually present between the ages 50-60 years however younger and older cases have occurred. A slightly higher incidence in males is reported. There is no evidence that MSA has a genetic factor.

The symptoms of MSA will vary from person to person with early onset autonomic nervous system changes being common. These include:

- Urinary problems
- Erectile dysfunction
- Postural hypotension
- Balance problems with falls

In addition the Parkinson's symptoms such as muscle rigidity and bradykinesia may be bilateral rather than the unilateral presentation of idiopathic Parkinson's.

Little or no response to levodopa therapy is an additional indicator although some people may experience a short-lived or partial response.

A diagnosis of MSA is made based on the clinical history and neurological examination. In some centres the treating specialist may suggest a MIBG scan to determine between Lewy Body Disease (Parkinson's and Lewy Body Dementia) and the other Atypical Parkinsonism conditions.

Abnormalities on scanning and imaging may not be seen early in the course of these conditions and therefore cannot usually be relied on to make the diagnosis. As in the case of Idiopathic Parkinson's, the Atypical Parkinsonism conditions are usually diagnosed on clinical assessment (the signs and symptoms on examination).

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Symptoms which may precede Parkinsonian symptoms include:

- Swallowing difficulties
- Inspiratory stridor
- Communication changes
- Slurred speech
- Urinary incontinence
- Increased muscle rigidity
- Impaired balance

Medical and pharmaceutical management may address symptoms of postural hypotension and constipation. Due to the lack of response to levodopa therapy allied health involvement is essential in order to maintain mobility, balance and safety (physiotherapy). The maintenance of functional independence will be assisted by occupational therapy. Speech and swallowing assessment and therapy will address these symptoms and the provision of an alternative communication aid may be required.

Parkinson's Nurse Specialists are familiar with this condition and can be a valuable resource and support for both the person and family.

Further information can be found in the MSA Information Kit available from Parkinson's Victoria - www.parkinsonsvic.org.au.

Progressive Supranuclear Palsy (PSP)

PSP was initially described by Steele, Richardson and Olszewski in 1964 and originally bore their names however Charles Dickens may have described the condition in his work *The Lazy Tour of Two Idle Apprentices* in 1867.

PSP is a sporadic condition with no association for toxic substances or genetics. The average age of onset of symptoms is 50-70 years and males are more commonly affected. It is thought to be associated with abnormal inclusions of tau which is a naturally occurring protein.

PSP is a rare condition with prevalence of 6:100,000 over the age of 55. It is estimated that there are 1,300 cases Australia wide.

A variety of early signs and symptoms include:

- Unsteady gait with falls
- Bradykinesia
- Rigidity
- Impaired eye movements
- Speech disturbances
- Cognitive impairment

The characteristic clinical sign of PSP is a supranuclear palsy which results in impaired voluntary eye movements. Downward gaze is initially affected followed by upward and subsequently horizontal gaze.

Blink rate is more reduced in PSP than Parkinson's and there may be an ocular stare with the upper eyelids resting above the irises.

In addition the Parkinson's symptoms such as muscle rigidity and bradykinesia may be bilateral rather than the unilateral presentation of idiopathic Parkinson's.

Little or no response to levodopa therapy is an additional indicator although some people may experience a short-lived or partial response.

The diagnosis of PSP is made using the same method and skills as MSA (see previous page).

Later symptoms include:

- Increasing falls usually backwards
- Rigid posture with neck extension rather than flexion
- Marked bradykinesia
- Loss of mobility
- Swallowing changes

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Levodopa therapy does not result in a positive response and symptoms are managed as they develop. As a result allied health involvement is essential in order to maintain mobility, balance and safety (physiotherapy). The maintenance of functional independence will be assisted by occupational therapy. Speech and swallowing assessment and therapy will address these symptoms and the provision of an alternative communication aid may be required.

Parkinson's Nurse Specialists are familiar with this condition and can be a valuable resource and support for both the person and family. Further information can be found in the PSP Information Kit available from Parkinson's Victoria - www.parkinsonsvic.org.au.

Cortico Basal Degeneration (CBD)

CBD was first described in 1968 with a study of three cases. There is no genetic link and it is less common than MSA or PSP. There have been no recorded cases diagnosed before the age of 40. There are no risk factors. Either gender may be affected.

It is thought to be associated with abnormal cellular tau related changes. Tau is a naturally occurring protein.

There are no classical signs seen on routine imaging. The diagnosis is based on the clinical symptoms and history. The unilateral presentation with a strong feature of 'alien limb' phenomenon will assist with the diagnosis.

Early symptoms are:

- Rigidity and bradykinesia
- An inability to perform purposeful tasks
- Postural and action tremor in some cases
- Clumsiness, jerking and sensory changes in one limb

These symptoms present unilaterally and CBD will progress steadily. There is poor response to levodopa therapy.

Later symptoms include:

- Visual apraxia
- Eating difficulties due to apraxia
- Self-care difficulties
- Progressive communication changes
- Dementia

Support and allied health input is recommended. Swallowing assessment and dietary modifications may be beneficial. Modification of the environment will assist with safety.

Parkinson's Nurse Specialists are familiar with this condition and can be a valuable resource and support for both the person and family.

Further information can be found in the CBD Information Kit available from Parkinson's Victoria - www.parkinsonsvic.org.au.

Lewy Body Dementia (LBD)

LBD is the second most common type of degenerative dementia and may also be referred to as Diffuse Lewy Body Dementia.

In the early 1900s Dr Friederich H Lewy identified abnormal deposits of the naturally occurring protein alpha-synuclein in the brain. These were later named Lewy Bodies. Lewy Bodies are found in both idiopathic Parkinson's and LBD but occur more diffusely in the latter. The term LBD was initially used in 1961.

There has been much debate on whether LBD is a separate identity or part of the progression of Idiopathic Parkinson's. Current thinking has defined the emergence of fluctuating cognitive changes within the first 12 months from diagnosis of Parkinson's (or commencement of symptoms) to be LBD. In addition early manifestation of hallucinations with little or no levodopa therapy is indicative of LBD.

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The age range of onset is 50-80 years. There are no identified environmental risk factors. Having a parent with LBD suggests the risk of developing dementia in the offspring is 20%. Either gender may be affected.

Early symptoms include:

- Bradykinesia
- Muscle rigidity
- Tremor
- Postural instability
- Dementia which fluctuates
- Visual hallucinations
- Spatial disorientation

Levodopa therapy may worsen the cognitive and hallucination symptoms.

There are no definitive laboratory tests for LBD and scanning and imaging will result in a similar outcome to idiopathic Parkinson's without determining which condition is present.

Later symptoms include:

- Inability to self care
- Excessive daytime sleepiness
- Complete dependence

As dementia is the major symptom referral to a state branch of Alzheimer's Association is recommended. Respite for the carer is essential and forward planning is recommended.

Summary

Atypical Parkinsonism may present initially as resembling idiopathic Parkinson's but the response to levodopa therapy is minimal.

The goal is to manage the symptoms as they arise. These conditions progress more rapidly and result in a shorter life expectancy than Parkinson's.

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