PARKINSON’S DISEASE OR PARKINSONISM

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Parkinson’s disease is the third most common neurological disorder in elderly after stroke and dementia. It is name after James Parkinson, a general practitioner from Hoxton, London, who described in his famous essay “The Shaking Palsy” a description of his patients that he observed. The classical features in his original essay were of “Rhythmic tremors, flexed posture, monotonous speech, tendency to fall, where senses and intellect were uninjured.” Nothing was much changed until the mid-20th century when neuro-pathogenesis, Dopamine deficiency, and treatment in terms of L-dopa replacement therapy was discovered. However, over the last 50+ years much advancement has occurred in understanding of the different symptomatology, therapeutic advancement and different variants of Parkinson’s disease from the original description of James Parkinson’s which has now been established.

The diagnosis of Parkinson’s disease remains clinical, what we observe, what we see and how we interpret, although there are technical advancements in terms of DAT scanning, SPECT and neuro-imaging by MRI and PET scanners, which is not widely available and at present remains a research tool. There are difficulties in distinguishing the disease from non-Dopa responsive Parkinson’s plus syndrome to Dopa responsive Parkinson’s disease. My paper briefly describes the simple way of distinguishing the classical Parkinson’s disease or its variants to Parkinson Plus syndrome in a broad manner.

As we all know that the classical Parkinson’s disease has three main core features, that is, bradykinesia, tremor and rigidity, with symptoms which are usually asymmetrical, involving primarily upper half of the body initially, progressing to involve the whole body at a later stage.

**Bradykinesia** – Slowness of movement remains a core feature and can be judged by noticing delay in initiation, slowness in exhibition of movements with or without loss of emotional expression, may or may not be associated with posture instability.

**Tremors** – Presenting features in 70% of patients with Parkinson’s disease. Normally they are unilateral, resting, aggravated by emotional stress/anxiety and decreased by action which disappears during sleep. To contrast essential tremors which are more common are primarily action tremors aggravated by performing fine tasks such as the use of a screwdriver etc.

**Rigidity** – A plastic resistance to movements and stiff muscles, stiffness by patient. It could be a continuous lead type of rigidity or more common “cogwheel” type that is broken up by rhythmic catches, may or may not be related to excessive tremors.

Nocturnal problems of Parkinson’s disease are often ignored such as difficulty in turning in bed, increased stiffness during the night, bladder instability and restless legs/cramps. Restless leg syndrome is now a new emerging entity which may or may not be related to underlying Parkinsonism. Similarly, non-motor symptoms of Parkinson’s disease are often overlooked which includes apathy, fatigue and neuro-psychiatric problems. Among the neuro-psychiatric problems depression/insomnia and obsessive compulsive trait are commonly observed symptoms. Slowness of thought, speech and swallowing difficulties, sphincter problems, nausea and excessive salivation are other features seen in Parkinson’s patients belonging to non-motor symptoms/problems of Parkinson’s disease.
It is also interesting to observe and note pre-morbid personality of Parkinson’s patients which was described in the early 1900’s, some of these features still hold true in many of the Parkinson’s patients. Parkinsonian patients are normally, successful, claim, reliable, shy, introverted, loyal and law abiding people. They are over controlled have restrictive emotions for love and do suppress aggressive tendencies. They may have some olfactory dysfunction with oily, greasy skin and usually are non-smokers. Mild obsessiveness, claustrophobic nature, compulsive neurosis and manic depression is also associated.

In a classical idiopathic Parkinson’s disease all the symptoms are normally unilateral, axial with core features of bradykinesia, tremors and rigidity associated with or without non-motor symptoms.

Now, I would like to mention variants of Parkinson’s disease:

1. **Tremors Dominant Parkinson’s disease (TDPD)** – These patient presented with typical Parkinsonian tremors, slow rhythmic unilateral, may be pill-rolling tremor without any or perhaps minimal rigidity and again without any bradykinesia. These peoples are normally very agile and may or may not have associated hypertension and postural instability. They have good responses to L-dopa replacement therapy and normally runs the benign course of the disease. At a later stage, these patients may develop signs of classical Parkinson’s disease.

TDPD is sometimes difficult to distinguish from essential tremors which are normally bilateral, generally have positive family history and predominately action tremors. They are ten times more common than PD and in fact 25% of PD patients diagnosed actually have essential tremors. In a difficult scenario, a DAT scan (Dopamine transporter agonist scan) is a very useful tool. It is highly specific that no essential tremors cases were mistaken as PD. It helps to diagnostic confidence and therefore improves patient’s management.

2. **Diffuse Lewy Body Disease.** There is a Dopa responsive Parkinsonian condition normally present with Parkinsonian symptoms with fluctuating mental state. Visual hallucinations are common, response to L-dopa replacement therapy is good and in selected cases use of acetyl cholinesterase inhibitors are indicated.

3. **Drug induced Parkinsonism.** Patient on long-term psychotropic medications, neuroleptics as well as other drugs which blocks synaoptic dopamine transmission develop Parkinson’s disease. Facial amimia, lack of arm swing movement, motor restlessness and stereotyping are essential features. Withdrawal of drugs, +/- replacement therapy improves in patient’s symptoms.

4. **Atherosclerotic Parkinsonism.** Predominately present with lower half akinesia where patient had difficulty in initiation of movements, turning corners, where tend to glue to the ground with freezing festination gait and have tendency to fall backwards. “Marche Petits Pas” is a classical gait for these patients. They have exaggerated deep tendon reflexes, manifestation of pyramidal tract involvement. A good history of recurrent TIA’s/previous stroke with or without CT scan evidence are useful tools. Some of these patients may be partially responsive to low dose L-dopa therapy which may confuse the diagnosis of classical Parkinson’s disease. On increment of L-dopa doses these patients normally develop more complications in relation to frequent falls etc.

5. **Parkinson’s disease with Vasculopathy.** This is a difficult entity where people may have physical signs of underlying Parkinson’s disease and later on develop stroke, cerebrovascular disease or vice versa. As people are living longer, this group of patients are becoming more and more evident and it is important to relate underlying different disorders needing a different type of treatment and management. A good history, CT head scan, DAT scan and clinical examination distinguishes this group of patients.

Now I would like to briefly discuss Parkinson’s Plus syndrome which primarily do not respond to L-dopa replacement therapy or if at all have partial response again in low dosage. Normally these people have features of Parkinson’s disease with some other neurological overlapping problems.

**Multi-systemic atrophy** normally presents in early 50’s associated classically with impotence and incontinence and usually have speech problems in terms of aphonia, anarthria and dysphasia. Slow and poor speech is a common feature, taken an underlying depression. Dementia is not a feature. Median survival from the date of diagnosis is around 9 years. When most of the features are present, the patients are normally non-demented Parkinsonian patients who are impotent and incontinent and have associated pyramidal tract signs of autonomic or cerebellar tracts. In other words there is a basal ganglia involvement plus pyramidal tract involvement which is manifested by exaggerated reflexes, gait apraxia or basal ganglia plus autonomic nervous system involvement or Parkinsonian features with cerebellar signs.

**Progressive Supra-nuclear Palsy** (Steel Richardson Olsezewski syndrome). It usually begins in the 7th decade manifested by Parkinson syndrome and vertical rigidity. A tendency to fall backwards along with postural instability associated with growling and gruff dysarthria again are the presenting features. Involuntary groaning/dysphasia is also seen. Most importantly tremors are never seen in this entity. They have 6th nerve palsy, levators inhibitions giving rise to difficulty in feeding or reading have given rise to “sloppy tie”
sign. These patients do not respond to L-dopa replacement therapy.

**Corticol Basal Degeneration** – Normally occurs sporadically in middle or late life. The onset is insidious and the course is progressive. Common features are clumsiness of one limb usually the arm due to a mixture of rigidity, akinesia and apraxia. The intellect and language remains intact and the average course of the disease is around 7 – 10 years. The “alien limb” may be presented as uncontrolled myoclonic jerks.

As the diagnosis of Parkinson’s disease/syndrome predominately remains clinical, it is vital to distinguish these symptoms correctly as their therapeutic response, prognosis and development of disabilities differs in different subgroups. Unnecessary long-term treatment and morbidity related to drugs, their side effects and patient expenses, drug costs, all could be save by accurate diagnosis.

**REFERENCES**
