How can I be sure that I have Parkinson’s disease?

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Overview

• Background
• Diagnostic Criteria
• Supportive tests
• Atypical features
• Mimicking disorders
Parkinson’s Disease

• Originally described by James Parkinson in 1817

• "Essay on the Shaking Palsy"
  – Descriptions of 6 cases, three of whom he personally examined; three he observed on the streets of London

• Charcot later in the 19th century gave credit to Parkinson by referring to the disease as “maladie de Parkinson” or Parkinson’s disease
What is Parkinson’s Disease (PD)?

• Progressive neurologic disorder of largely unknown cause that results in the loss of specific cells in your brain that produce a chemical called dopamine

• Dopamine - a messenger responsible for transmitting signals within the brain that allow for coordination of movement

• Loss of dopamine leaves patients less able to control their movement, rendering them slow, stiff and/or shaky
How can I be sure I have PD?

• The road to diagnosis can be quite arduous and prolonged

• There is no blood test or Xray for PD

• Definitive diagnosis
  – Pathological confirmation at autopsy

• In clinical practice, diagnosis is based on
  – Thorough history
  – Physical examination
Questions you may be asked …

• Do you shake?
• Slower in your usual activities?
• Hand writing smaller?
• Trouble buttoning buttons, tying shoe laces etc?
• Difficulty standing up from a chair?
• Don’t swing your arms?
• Hunched over, walking “like an old man”?
• Voice quiet or soft?
What are the signs and symptoms of PD?

• Main Symptoms
  – Tremor (shaking)
    » 80% of people with PD
  – Bradykinesia (slowness of movement)
    » Facial expression, Finger taps
  – Rigidity (stiffness)
    » Passively move limbs
  – Postural instability (difficulty with balance)
    » Pull test

• If any 2 of these features are present, we can confidently say that you have *parkinsonism*
Diagnostic criteria - UK Parkinson’s Disease Society Brain Bank and the National Institute of Neurological Disorders and Stroke

• Bradykinesia + 1 other sign:
  – Shaking
  – Stiffness
  – Postural instability

• Possible
  – At least 2 features
  – Sustained response to levodopa

• Probable
  – At least 3 features
  – Sustained response to levodopa

• Definite
  – Pathologic confirmation

• Supportive features:
  – Symptoms start on one side of the body
  – Tremor at rest
  – Slow progression
  – Response to levodopa for at least five years
  – Dyskinesia
How sure are we that you truly have PD?

• The rate of misdiagnosis can be relatively high, especially when the diagnosis is made by a non-specialist.

• Using these criteria, specialist can make a diagnosis of PD with ~ 90% accuracy.
Are there any blood test or brain scans that can diagnose PD?

- No

- Dopamine Transporter Scan (DaTscan)
  - Approved in 2011
  - Shows level of dopamine activity in the brain, which is lower among people with PD

- Confirms clinical suspicion of parkinsonism

- The difficult part is figuring out what is causing the parkinsonism
What is the difference between parkinsonism and Parkinson’s disease?

- Parkinsonism
  - At least 2 symptoms (tremor, rigidity, slowness, balance problems)

- The vast majority of patients (~85%) with parkinsonism have Parkinson’s disease

- However, certain medications, vascular problems, and other neurodegenerative diseases can cause parkinsonism (Atypical parkinsonian disorders)
When should an Atypical Parkinsonian Syndrome be suspected?

- In patients with:
  - Rapid onset or progression
  - Poor response to medications (dopamine)
  - Early falls
  - Eye movement abnormalities
  - Early memory or behavioral changes (dementia)
  - Early autonomic dysfunction - postural hypotension, urinary and bowel incontinence
What are the Atypical Parkinsonian Disorders?

- Previously known as Parkinson’s plus syndromes
  - Progressive Supranuclear Palsy (PSP)
  - Corticobasal Degeneration (CBD)
  - Multiple System Atrophy (MSA)
  - Dementia with Lewy bodies (DLB)
  - Vascular Parkinsonism

- Differentiating between these disorders may be difficult as they share many clinical features of PD
- Early diagnosis is important because treatment and prognosis differ greatly from those with PD
What are the Atypical Parkinsonian Disorders?

- **Progressive Supranuclear Palsy (PSP)**
  - *Progressive* disease, causes weakness (palsy) by damaging certain parts of the brain above the nuclei that control eye movements (supranuclear)
  - 1:100,000 people over the age of 60
  - Symptoms include:
    - Loss of balance while walking
    - Unexplained falls
    - Personality and behavioral changes
    - Slurring of speech
    - Eye movement abnormalities
  - Rapid progression (5-8 years), minimal response levodopa
  - Supportive treatment: Speech and physical therapy; antidepressants
What are the Atypical Parkinsonian Disorders?

- **Corticobasal Degeneration (CBD)**
  - Progressive neurological disorder that results in loss of cells in the cerebral cortex and basal ganglia
  - Symptoms:
    - Slowness, stiffness that starts on one side
    - Dystonia - Twisting/pulling of one limb
    - Myoclonus - Muscle jerking/twitching
    - Apraxia - Difficulty performing familiar movements with one limb despite being physically capable
  - Rapid progression: 6-8 years; minimal response to levodopa

- Supportive treatment: botulinum toxin (Botox) for dystonia, antidepressants, speech and physical therapy may be helpful
What are the Atypical Parkinsonian Disorders?

• **Multiple System Atrophy (MSA)**
• Progressive neurological disease that affects mobility as well as the autonomic nervous system
• Symptoms include:
  – Slowness, stiffness on both sides of body
  – Poor balance
  – Difficulty with coordination, clumsiness
  – Autonomic dysfunction
    » Blood pressure problems (orthostatic hypotension)
    » Fainting spells
    » Bladder control problems
• Rapid progression: 5-8 years; minimal response levodopa
• Supportive treatment: speech and physical therapy; Lightheadedness may improve with certain medications (fludrocortisone, midodrine)
What are the Atypical Parkinsonian Disorders?

- **Dementia with Lewy bodies (DLB)**
- Second most common form of dementia after Alzheimer’s disease
- Symptoms:
  - Dementia + parkinsonism
  - Progressive intellectual and functional deterioration
  - Early hallucinations, confusion that fluctuates, problems with attention, problem solving, planning, the ability to produce and recognize figures)
- There are no known therapies to stop or slow the progression of DLB
- Certain medications (donepezil, rivastigmine patch) may be beneficial
What are the Atypical Parkinsonian Disorders?

- **Vascular Parkinsonism**
  - Parkinsonism caused by multiple small strokes
  - MRI is indicated
    - Cannot make a definitive diagnosis but may support clinical suspicion
- Symptoms:
  - Stiffness and slowness primarily affects legs
  - Walking difficulty
  - “Lower half or lower body parkinsonism”
- No specific treatment
  - 10-30% may respond to levodopa
  - Reduce risk factors:
    - High blood pressure, diabetes, heart disease, cholesterol problems, smoking etc
What are the conditions that mimic PD?

- Medication
- Essential Tremor
- Normal Pressure Hydrocephalus
What are the conditions that mimic PD?

- **Medications**
  - Any drug that blocks the action of dopamine is likely to cause parkinsonism
  - Drugs used to treat psychiatric disorders – neuroleptics (halodol, thorazine etc)
  - Anticonvulsants - valproic acid
  - Mood stabilizers - Lithium
  - Drugs to control nausea – Metoclopramide (Reglan), prochlorperazine (compazine)

- If parkinsonism is caused by one of these medications, symptoms should gradually disappear once the drug is stopped
- If symptoms persist, we may have “unmasked” underlying neurodegenerative PD
What are the conditions that mimic PD?

- **Essential Tremor (ET)**
  - Most common form of tremor
  - Different from the PD tremor
    - Affects both hands, occurs primarily with action (when performing a task like writing or eating)
    - Often involves head/neck and even voice
    - Does NOT affect legs
  - Progressive disorder but tremor remains the only problem
  - Treatment: various medications (propranolol, primidone)
What are the conditions that mimic PD?

• **Normal Pressure Hydrocephalus (NPH)**
  – Cerebrospinal fluid inside the brain does not drain properly
  – Results in a trio of problems:
    » Difficulty walking, slowed thinking, and loss of bladder control
  – Diagnosed by history, physical exam, MRI of brain, lumbar puncture (spinal tap)
  – The treatment involves surgery where a shunt is placed to help drain the excess fluid
To sum it up....

- PD is a clinical diagnosis
- There are many medications and other conditions that can mimic PD
- There are no blood tests or brain scans that can diagnose PD
- DATscan may be helpful in certain cases:
  - Parkinsonism from mimickers like ET, NPH, vascular or drug induced parkinsonism, or AD
- Specialists can make the correct diagnosis with ~90% accuracy
- Early diagnosis is important as there are many treatments available that can improve your quality of life
Learn more about it....