Primary Atypical Parkinsonisms
PD plus syndromes

NEUROLOGY TRAINING FOR NON-NEUROLOGISTS IN WEST AFRICA

Focus on Parkinson’s Disease and other Neurodegenerative Disorders

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• Parkinson’s disease (PD) is a chronic and progressive neurodegenerative disease.

• Initially thought to be rare in Africa but..
Parkinson's disease

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Basal ganglia
What?

• Movement disorder(s)

• Ghana ? 12-15% of neurology clinic

• 3rd commonest cause of disability worldwide

• 1% over 60, 5% over 85 – now Ghana population ageing
Case study

• 60 year old woman
• Presented with poor balance and falls
• Also generalised ‘slowing down’

• O/E
• MMSE – 20/30
• Expressionless face, fixed stare
• Impaired eye movements (up and down gaze)
• Dysarthria
• Increased tone all 4 limbs and neck/axial rigidity
• Brisk reflexes

• What is the diagnosis?
  – Progressive supranuclear palsy

• What treatment would you recommend?
  – Levodopa, though response usually minimal
Progressive Supranuclear Palsy
PSP
The clinical definition of multiple system atrophy (MSA)

- is a progressive, idiopathic, degenerative process beginning in adulthood.
- Patients present with various degrees of parkinsonism, autonomic failure, cerebellar dysfunction, and pyramidal signs that are poorly responsive to levodopa or dopamine agonists.
- Glial cytoplasmic inclusions (GCIs) and a neuronal multisystem degeneration are the pathologic hallmarks of this clinically variable disorder.
When is a Parkinsonian syndrome not idiopathic Parkinson's disease? “Red flags”

- History of severe cerebral trauma, stroke, exposure to neurotoxins or anti-dopaminergic agents
- No rest tremor
- Symmetrical signs
- Early falls
- Associated ophthalmoplegia, pyramidal or cerebellar signs
- Associated autonomic dysfunction
- Rapid disease progression
- Poor response to levodopa
“Parkinson’s plus” syndromes

1. Multiple system atrophy
   - May look like PD (‘striatonigral degeneration type’) – but little/no L-dopa response
   - May be predominantly cerebellar syndrome (often with parkinsonism)
   - May be predominantly autonomic failure (Shy-Drager type)
     - prominent urinary symptoms
     - postural hypotension
   - Often more aggressive than IPD
   - Cognitive impairment rare
   - Minimal response to treatment
MSA What sign?
“Hot cross bun”
“Parkinson’s plus” syndromes

2. Progressive supranuclear palsy
   • Parkinsonian features but other key findings
     – Early postural instability (often presents with falls)
     – Early cognitive impairment
     – Typical examination findings
       • ‘staring eyes’
       • Trunkal rigidity
       • Impaired eye movements (esp downgaze)
   • Poor prognosis and poor levodopa response

3. Cortico-basal degeneration
   • Parkinsonism, cognitive decline, apraxia
Corticobasal ganglionic degeneration (CBGD)

• is characterized by frontoparietal cortical atrophy in addition to degeneration within the extrapyramidal system.
• The disease tends to occur in those aged 60-80 years, with a mean age of onset of 63 years.
• CBGD is a rare syndrome No familial or environmental factors appear to influence CBGD.
• Progressive supranuclear palsy (PSP) and multiple system atrophy (MSA) may initially be confused with CBGD. However, the true diagnosis becomes clear as the apraxia and dystonia develop.
• Again, the clinical and pathologic features of PSP and CBGD can overlap considerably.
• Symptoms on long-term follow-up include focal or asymmetric rigidity, bradykinesia, postural and action tremor, and marked dystonia.
• These problems usually arise predominantly in one upper extremity.
• Limb apraxia may become a serious problem, with independent movements occasionally as severe as an alien limb.
• The incidence of limb apraxia is far higher in CBGD than in PSP, with a similar level of cognitive impairment.
Diffuse Lewy Body Disease

• Diffuse Lewy body disease (DLBD) is a progressive neurodegenerative disorder characterized by the presence of parkinsonian symptoms and neuropsychiatric disturbances commonly accompanied by dementia.

• Progressive dementia is often the first and predominant symptom.

• Of note, longitudinal studies show that after a decade of motor symptoms, 78% of patients with Parkinson disease meet the criteria for dementia.
DLBD

• Approximately 20% of patients do not have any parkinsonian features.
• Neuropsychological deficits that have been described include aphasia, dyscalculia, and apraxia.
• A psychotic state develops in approximately 20% of patients.
• Depression, auditory and visual hallucinations, and paranoid ideation may occur.
• These patients are more likely to have cognitive adverse effects with levodopa therapy in early stages than patients with Parkinson disease.
• Not uncommon
• Look out for red flags and non-response to Levodopa

• PD/PSP/CBD/MSA