SMART PAPER – Movement Disorders Dementias

PROGRESSIVE SUPRANUCLEAR PALSY SYNDROME

Classic brain-stem variant – Diplopia / blurred vision. Photophobia. Brain stem atrophy. Vertical gaze palsy – going downstairs, tripping on objects, reading,. Slurred speech. Stiffness and motor slowing, though tremor usually absent.

Timed attention tests will be affected by slowness. Executive functions affected, esp phonemic fluency. VOSP number-location.

Reduced speech output. Nonfluent aphasia may occur. Impaired on recall rather than recognition memory.

Postural instability. High frequency of falls b'wards (early sign). Symmetrical Parkinsonian features. Later: swallowing difficulties

Early changes in personality may occur – apathy, emotional lability, slowed thinking. Cortical and other variants distinguished.

CORTICOBASAL SYNDROME

Limb apraxia, usually unilateral. Alien hand.

Copying unfamiliar hand postures impaired. Difficulty in tasks with one hand, e.g. toss coin

Executive dysfunction, memory impairment & parietal lobe deficits. VOSP spatial subtests.

Cortical sensory loss. Graphesthesia. Myoclonus (jerky movements). Parietal & frontal atrophy.

Language deficits may encompass both expression & syntactic comprehension. Memory less impaired than in AD. Cognitive symptoms may precede motor symptoms.

'PARKINSON'S PLUS' DEMENTIAS

'Parkinsons PLUS' conditions seldom have resting tremor, and usually respond poorly to dopamine. Progression is usually faster than in PD. CBD and PSP decline faster than AD. PD with dementia may differ from LBD only in major cognitive symptoms following motor symptoms in former, and vice versa in LBD. Asking patients to clap 3 times may be abnormal, esp in PSP.

LEWY BODY DEMENTIA

Acting out behaviour In vivid dreams

Onset usually after 65 yrs. Affects men more. Likely to have falls, & autonomic symptoms - fainting, urge incontinence. MR scan normal.

Rigidity, facial impassivity & symmetry of motor signs. Tremor less common. Behavioural precede motor changes.

Visual hallucinations – animals, people. Non-threatening. May be familiar. Presence of other ('extracampine'). Capgras may occur – thinks imposters exist.

Fluctuating cog. impairment. Stares into space for periods. Disorganized ideas, speech. Sleeps during day.

Impaired on block design, fig. copy, map search, fluency, trail making, frag letters, cube counting, backward span, clock drawing and copy. Executive, visuospatial, attention more impaired, & memory less impaired than AD. Attention & v-spatial more impaired than CBD, esp. early on.

PARKINSON'S DISEASE (PD)

Slowing down & motor symptoms usually precede cognitive. Sometimes weight loss, constipation, and olfactory loss reported.

Features include tremor, slowness, loss of facial expression, shuffling gait., small handwriting. Hypophonic speech. Depression common.

Visuospatial deficits usually mild. Executive deficits variable, often mild in early stages. Impaired on timed perceptual-motor tests. Small handwriting. Recall rather than recognition deficits.

Non-motor symptoms more common in older PD patients - loss of interest, visual or extra-campine hallucinations, cog symptoms, anxiety, change in libido / sexual activities. Marked olfactory loss in PD may predict dementia.

NORMAL PRESSURE HYDROCEPHALUS

Gait disturbance, incontinence and impaired cognitive function. Variable apathy.

Early or severe cognitive deficits suggests alternative additional pathology

Executive dysfunction & mild-mod. memory impairment. Slowing. No major language or visuoperceptual deficits.

Periventricular lucencies and minimal cortical atrophy.

SPORADIC CJD

Myoclonus (jerky movements) and gait disturbance. EEG, CSF and MR (caudate & putamen) changes.

Perseverations, intrusions from earlier tasks, backtracking, lapses of attention.

Rapidity of cognitive decline.

MULTIPLE SYSTEM ATROPHY

Onset in 50s/60s. Duration 5-7 years. Urinary incontinence. Orthostat. hypotension, constipation; erectile problems. Parkinsonian & cerebellar versions. REM sleep disorder. Slurred speech & pathol. laughter, crying occasionally seen. Relatively rapid decline. Parkinsonian features don't respond well to L-dopa.

In most cases, relatively mild cognitive impairment, primarily affecting executive functions, though may be more marked than in PD.

MELAS. Strokes, headache, hearing loss, early onset. **MS, ADEM**. VER, EEG, MRI white matter lesions (MS). In MS, usually attention and memory deficits.

OTHER CONDITIONS

Fragile X with tremor & ataxia – late-onset frontal-subcortical syndrome. PD signs. Cerebellum lesions.

Fahr's Disease. Basal ganglia calcification. Executive dysfunction.

Subacute diencephalic angioencephalopathy – features of patients with thalamic lesions.

Vasculitis. Sarcoidosis. Lymphomatous lesions: primary-sec.

Huntington's Disease. Early attent. & executive deficits.

Friedrich's Ataxia. Spino-cerebellar path. <20yrs onset

Adrenoleukodystrophy. White matter lesions. May have schizophrenia features.

Kuf's Disease. Myoclonic epilepsy, dementia & ataxia **or** Behavioural change, dementia, motor disorder & facial dyskinesia.

Frontotemporal dementia & Parkinsonian signs – abnormality on chromosome 17. PSNI Alzheimer mutation may include movement disorder.

Encephalitis Lethargica. Post-encephalitic. Sleep disturbance. Oculomotor signs. May make good recovery with steroids. Occasional occurrence of delayed-onset Parkinsonian signs.

Rare - Metachromatic leukodystrophy (demyelination on MRI), CADASIL, Nieman-Pick Type C, Syphillis, HIV, Mitochondrial Disease.

Autoimmune Disorders. Voltage-gated potassium channel antibodies. NMDA receptor antibodies. Anti-ma2 antibodies. Primary tumour may be present. Clinical features may include memory loss, psychiatric disturbance, movement disorder, seizures, impaired level of consciousness.

Systemic Lupus Erythematosus. May present with chorea & dementia, esp those with anticardiolipin antibodies.

Whipple's Disease. GI symptoms, oculomotor with chewing abnorm., hypothalamic symptoms.

Wilson's Disease. Copper metabolism tests. Distinct rings around iris. High signal in basal ganglia, esp putamen. Young age. PD signs, ataxia, or slurred speech.

Neuroacanthocytosis. Genetic disorder, with HD-like features. Frontal-subcortical dysfunction. Striatal atrophy

Motor Neurone Disease. Frontal dementia, esp in bulbar type. Nonfluent aphasia rather than semantic dementia. Verb comprehen. impaired.

Hashimoto's encephalopathy. Thyroid-related. Myoclonus. Occas. seizures. Steroid responsive. Female bias. Amnesia, attention deficits and aphasia may occur.

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