MOVEMENT DISORDERS



Parts of the motor circuits

- cortical centers: there are centers in all the cortical lobes
- subcortical centers:
- caudate nucleus
 putamen
 pallidum
 striatum
 nucleus lentiformis
- subthalamical nucleus (Luys)
- nucleus ruber
- substantia nigra



There are two main clinically different groups:

hypokinetic-hypertonic (rigid)

hyperkinetic-hypotonic



hypokinetic-hypertonic (rigid) syndrome

Parkinson's disease, Parkinsonian syndromes



The etiology of the Parkinsonism

- primer, idiopathic form: Parkinson's disease
- the second most frequent progressive neurodegenerative disease
- the prevalence is 1% in the 65 year old population and it increases to 4-5% in the 85 year old population
- it is most frequently sporadic
- approximately 10% of the cases are familiar (there are 16 indentified genes, wich cause autosomal dominant, or recessive forms of Parkinson's disease)

The etiology of the Parkinsonian syndromes

- symptomatic: Parkinson's syndrome
- trauma
- tumor
- inflammation (encephalitis)
- stroke (ischemic, hemorrhagic)
- toxic agents (CO, neuroleptic drugs)



The etiology of the Parkinsonian syndromes

Parkinson plus diseases

- Synucleopathies:

Multi System Atrophy (MSA)

- Taupathies (abnormaly fosforilated Tau protein):

Progresive Supranuclear Palsy (PSP)=

Steel-Richardson-Olszewski sy.

Corticobasal Degeneration

The neuropathologic basis of Parkinson's disease

- degeneration of the substantia nigra and the striatum (Lewy bodies- citoplasmatic protein aggregations)
- disintegration of the dopamin-acetilcholin balance in the central nervous system
- decrease of the dopamin level and cholinergic dominance

the main motor symptoms of Parkinsonism

- rigidity
- tremor
- hypo-, and bradykinesis
- postural instability



rigidity

- the tone of the agonist és antagonist (flexor and extensor) muscles increases paralelly
- the trunk and the extremities are in semiflected position
- during passive movement of the extremities we can feel permanent resistance (cogwheel phenomenon)
- very often assimetrical distribution



tremor

- the agonistic and antagonistic (flexor és extensor) muscles contract with rhytmic, 6-8/sec. frequency
- the tremor is most prominent in resting position, the stress could provoke it
- the intended movements decrease the tremor and it is stopped during sleep
- very often assimetrical distribution

hypo-, and bradykinesis

- movements and walking become slow, sluggish and difficult
- shuffling gait with decreased excursion of legs
- difficulties in turning
- freezing: sudden stop in walking and movements



other signs (motor)

- on-off phenomenon
- retropulsion
- hypomimia, or blank face
- monotonous speech
- micrographia



other signs (non motor)

- hyposmia, hypogeusia
- vegetative problems: orthostatic hypotension, incontinence, impotence, hyperperspiration
- gastrointestinal problems :swallowing difficulty, hypersalivation, gastroparesis, constipation
- pain and other sensory disturbances
- depression
- sleep disorders



Parkinson plus diseases Multi System Atrophy (MSA) - Papp-Lantos disease

- the stages of the disease:
 - striato-nigral degeneration
 - olivo-ponto-cerebellar atrophy
 - **Shy-Drager syndrome**
- no, or mild asimmetricity in the parkinsonic signs
- early falls down
- autonomic dysfunctions (orthostatic hypotension, impotence, incontinence, respiratory stridor)
- "dropped head"
- pyramidal signs, cerebellar signs
- no, or just partial respons to levodopa therapy



Parkinson plus diseases Progresive Supranuclear Palsy (PSP)Steel-Richardson-Olszewski sy.

- postural instability, early falls down, axial rigidity, freezing
- supranuclear gaze paresis (predominantly vertical) downwards gaze!
- frontal lobe signs (primitive reflexes, apathy, frontal demencia)



the Parkinsonian crisis could be life threatening!!

- complete immobility
- aphagia and anarthria
- extreme rigidity
- fever
- cardiovascular inssufficiency
- bedsores, pneumonia, deep vein thrombosis, lung embolism

treatment

DRUGS

- MAO B inhibitors (selegilin, rasagiline)
- Amantadin (PK Merz)
- Dopamin agonists (ergotamine type:bromocriptin, non ergotamin type: ropinirole, pramipexole, rotigotine, apomorphine)
- L-DOPA substitution (L-DOPA+DOPA decarboxilase inhibitor) per os and intrajejunal infusion DUODOPA
- COMT inhibitors (entacapone)
- L-DOPA+DOPA decarboxilase inhibitor+ COMT inhibitor
- anticholinergic drugs (metixene, procyclidine)

treatment

SURGICAL (just in severe cases, when the antiparkinsonian drugs are uneffective)

- deep brain stimulation DBS (uni-, or bilateral) target zones: STN, pallidum, thalamus VIM nucleus
- thalamotomy, pallidotomy (uni-, or bilateral)
- neurotransplantation (inplantation of fetal mesencephalic cells into the caudat nucleus and putamen region)

treatment

• physicotherapy (training, special exercises)

psychotherapy

