PICKS DISEASE

Background
1. Definition
   o Dementia secondary to focal degeneration of the frontal and/or temporal lobes
   o Progressive personality and behavioral changes
2. General Information
   o First described by Arnold Pick in 1892
   o Also known as: Frontotemporal dementia or degeneration, Frontal lobe dementia or degeneration, Pick Complex
   o Behavioral variant most common; may also present with language and motor impairments

Pathophysiology
1. Pathology of Disease
   o Degeneration and atrophy of frontal and/or temporal lobes with fibrous scarring due to astrocyte proliferation and neuronal loss
   o Tau protein depositions (Pick bodies) found in some variants; more commonly in familial cases. Up to 50 Tau mutations have been found
   o Various other protein mutations have been found on autopsy
   o Serotonin binding decreased in frontal lobes
2. Incidence, Prevalence
   o Exact prevalence unknown; estimates range from 10-20% of dementias
   o Relative frequency of 3-10% according to autopsies
   o Mean age at onset - 57 years
   o Rare before age 40 or after age 75
   o Possible male predominance
   o Most cases have no family history of dementia; some variants are noted to have autosomal dominant inheritance
3. Morbidity / Mortality
   o Gradually progressive
   o Advanced disease predisposes patients to serious complications such as injuries and pneumonia

Diagnostics
1. History
   o Variable symptoms at presentation
   o Cognitive and visual-spatial abilities initially preserved
   o Behavioral variant
     - Personality changes: apathy, impulsivity, disinhibition, etc.
     - Lack of insight and loss of concern for self or others
     - Loss of social decorum
     - Ritualistic behaviors: hoarding, pacing, etc.
     - Emotional blunting
     - Difficulty adapting to change
     - Changes in attention ranging from distractible to perseverative
   o Language variants
• Progressive non-fluent aphasia characterized by difficulty naming objects and dysfluent speech
• Semantic dementia characterized by fluent empty speech with loss of word meaning
  o Motor variants
   • Motor neuron disease resulting in atrophy, flaccidity, muscle fasciculations
   • Corticobasal degeneration characterized by rigidity and uncontrolled spontaneous movements
   • Progressive supranuclear palsy characterized by vertical gaze palsy, rigidity, bradykinesia, dystonia

2. Physical Examination
  o Primitive reflexes such as grasp, rooting, sucking noted as frontal lobe disease progresses
  o Incontinence
  o Rigidity, tremor, akinesia
  o Low blood pressure

3. Diagnosis
  o Primarily clinical
  o Laboratory evaluation may be helpful to rule out other causes of delirium and dementia; no diagnostic labs specific to Pick's disease
  o Diagnostic imaging
   • MRI required to rule out illness due to other structural cause
   • Atrophy of affected frontal and temporal lobes noted in later disease
  o Other studies
   • Neuropsychologic testing can help quantify impairment but not diagnostic
   • MMSE may be normal in early cases
   • EEG remains normal despite dementia
   • Definitive diagnosis by postmortem autopsy

4. Diagnostic Criteria
  o Development of behavioral or cognitive deficits manifested by either
    1a: Early and progressive changes in personality; emotional blunting and/or loss of empathy; difficulty modulating behavior; often resulting in inappropriate responses or activities, or
    1b: Early and progressive change in language; problems with expression of language or severe naming difficulty; problems with word meaning
  o 1a or 1b deficits cause significant impairment in social or occupational functioning; represent significant decline from previous level of functioning
  o Course characterized by gradual onset and continuing functional decline
  o 1a or 1b deficits not due to other nervous system conditions (e.g., cerebrovascular disease), systemic conditions (e.g., hypothyroidism), or substance-induced conditions.
  o Deficits do not occur exclusively during delirium.
  o Disturbance not better accounted for by psychiatric diagnosis (e.g., depression).
Differential Diagnoses
1. **Alzheimer disease**
   - Often later age of onset; less behavioral feature; more prominent memory loss than frontotemporal dementia
   - Has more cholinergic deficits
   - Definitive diagnosis postmortem
2. **Lewy body dementia**
   - Visual hallucinations common, but not commonly seen in Picks disease
3. **Psychiatric disorders (depression, OCD, Bipolar disorder, etc.)**
   - Earlier age of onset
   - May have delusions or hallucinations; uncommon with Picks disease
   - Respond more favorably to pharmacologic treatment
4. **Structural disease (tumor, infarct, trauma, abscess, etc.)**
   - Seen on CT or MRI

Therapeutics
1. **Supportive treatment**
   - No pharmacologic treatment slows progression
   - Established routine and patterns may help avoid agitation
   - SSRIs may help with obsessive compulsive behaviors, disinhibition, depression, and carbohydrate craving
   - Trazodone and antipsychotic medications tried with varying success
2. **Long-Term Care**
   - In-home nursing care or skilled nursing facility may be needed to provide cares and supervision as disease progresses

Prognosis
1. 50% mortality at 5 years from diagnosis, 80% at 8 years
2. Semantic deficits, word-finding problems, and language difficulties at the time of diagnosis give a worse prognosis
3. Positive family history and age >64 years at onset also have a worse prognosis

Patient Education
1. Association for Frontotemporal Disorders, [www.ftd-picks.org](http://www.ftd-picks.org)

References


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