Pathology

Dementia — Signs and Symptoms

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Increasing life expectancies have led to an increase in dementia. With the rise of postulated age-depending prevalence, a drastic increase of cases of sickness was witnessed. The definition of dementia and its diagnosis will be highlighted in this article.

Definition of Dementia

Dementia is defined as a decline in memory with the impairment of at least one other cognitive domain. Other cognitive functions that may be impaired include skilled movements, language, or executive functioning. To be classified as dementia, the impairment in memory and other cognitive functions must interfere with social and occupational functioning and cannot be accounted for by other psychiatric conditions or psychoses.

- Alzheimer’s disease is not synonymous with dementia but is the most common cause (75%).
- Dementia is not a feature of normal aging and is always indicative of pathology.
- Mild cognitive impairment (MCI) is a transition stage to Alzheimer’s disease; 10% of patients with MCI develop Alzheimer’s every year.
Dementia as a symptom complex

Dementia per se is not a diagnosis, but a symptom complex, which can occur as a consequence of different organic diseases. It is, therefore, a dementing syndrome, a state that is, according to ICD-10-criteria, characterized by:

- Impairment of memory
- Impairment of at least one other cognitive function such as thinking, orientation, perception, calculating ability, learning ability, language, and the ability to judge
- A change in emotional control, social behavior, and motivation, without impairment of consciousness, for a minimum of 6 months

Epidemiology of Dementia

Alzheimer’s disease as the main reason for dementia

The most common cause of dementia is Alzheimer’s disease (also known as Alzheimer’s dementia); this disease accounts for almost two-thirds of all dementia cases. This is followed by vascular dementia (15%) and combined forms of vascular and Alzheimer’s disease (15%). The remaining 10% of cases are caused by other neurological diseases or other disorders.

Dementia is age-related and has an estimated prevalence of 20% in those older than 85 years. Up to 11% of dementia cases are considered to be reversible. Early treatment in these cases can reverse the condition and restore normal functioning.

Symptoms of Dementia

Gradual begin of dementia

The first warning signs of dementia are often subtle and only apparent to close relatives,
not to the affected person themselves. They include repetition of the same questions, a narration of an always identical story, and the misplacing of objects (often, others are accused of having misplaced the item). Secure handling of financial issues is lost, as well as the ability to properly care for oneself.

An extraordinary psychological strain arises for dementia patients, especially during early phases of the disease. They suffer from apathy, aberrant motor behavior (wandering aimlessly), eating of non-edibles, irritability, aggression, sleep disturbances, and depression.

Insecure fine motor skills and decreased vocabulary accrue when the disease progresses. Inner self-reflection diminishes and unfounded outbursts of anger and violence occur. As the disease progresses, close relatives are no longer recognized and the ability to master everyday tasks is lost. Musculature shrinks and patients begin to suffer from urinary and fecal incontinence. Another characteristic is small, tripping steps.

The disease, in the end, leads to bedridden patients who are totally dependent on nursing care. Most patients only survive due to the implantation of a percutaneous endoscopic gastrostomy tube, as the ability to swallow is lost. Patients die due to pneumonia or heart failure.

Patients with Lewy body dementia suffer from hallucinations, which are mostly dominated by fear and feature topics such as kidnapping or similarly frightening scenarios.

Diagnostic Investigation of Dementia

Neuropsychological test devices for the diagnosis of dementia

The ICD-10 classification of mental and behavioral disorders criteria need to be fulfilled in order to diagnose a dementing disorder. Neuropsychological test devices such as the Mini-Mental State Examination (MMSE), the clock-drawing test, and the DemTect test are used for diagnosis and quantify cognitive deficiencies.

Mini-Mental-State-Examination (MMSE)

The MMSE is a test for the detection of cognitive impairments. It includes questions and tasks in 10 categories:

- **Orientation**: 10 questions concerning temporal and spatial orientation (maximum 10 points).
- **Retention**: Repeating 3 words, up to 3 repetitions (maximum 3 points).
- **Attention and calculation**: Progressive subtraction by 7, starting at 100 (maximum 5 points with 5 correct calculation steps).
- **Memory**: Remembering 3 words from the beginning (maximum 3 points).
- **Naming**: Correct naming of 2 shown items (e.g., a watch, pencil; maximum 2 points).
- **Repetition**: E.g., “without fuss or quibble” (maximum 1 point).
- **Active part and speech comprehension**: E.g., “Take this piece of paper in your hands, fold it in the middle, and put it on the ground” (1 point for every correctly performed action; maximum 3 points).
- **Reading**: Reading and performing an instruction (e.g., “close your eyes”; maximum 1 point).
- **Writing:** Writing any sentence (maximum 1 point).
- **Drawing:** Copying a given geometric figure (e.g., 2 interlocked pentagons; maximum 1 point).

The maximum obtainable score in the MMSE is **30 points**. A score lower than 26 may indicate mild dementia.

<table>
<thead>
<tr>
<th>Score</th>
<th>Grading</th>
</tr>
</thead>
<tbody>
<tr>
<td>27 – 30</td>
<td>No dementia</td>
</tr>
<tr>
<td>26 – 20</td>
<td>Mild dementia</td>
</tr>
<tr>
<td>10 – 19</td>
<td>Moderate dementia</td>
</tr>
<tr>
<td>&lt; 9</td>
<td>Severe dementia</td>
</tr>
</tbody>
</table>

**Clock Test**

The clock test examines **spatial imagination and visual thinking**. The patient is asked to draw the dial of a clock including a time set by the examiner (e.g., 10 to 12). The presentation of the dial (recognizability of the clock, correct filling in of the digits), the position of the clock’s hands (correct presentation of the set time), and the size of the hands in relation to each other are evaluated using a grading system.

**Differential Diagnosis of Dementia**

Understanding the possible reasons for cognitive decline is a key part of diagnosing either a potentially reversible or irreversible dementing disorder.

**Potentially reversible dementing syndromes**

Potentially reversible dementing disorders can arise within the scope of different diseases and disorders; this should be clarified as a possible reason before other investigations begin. They include the following:

- Psychiatric diseases (pseudodementia, or depression-related cognitive dysfunction)
- Anaemia
- Electrolyte imbalances
- Renal or liver dysfunction
- Endocrinological impairments (hyper- or hypothyreosis, **diabetes mellitus** [hypoglycemia])
- Malnutrition and hypovitaminosis (vitamin B12, B6, B3, or folic acid deficiency)
- Infectious diseases (syphilis, borreliosis, **HIV**)
- Overdose (e.g., from alcohol or digitalis glycosides)
- Metabolic diseases (e.g., **Wilson’s disease**)
- Autoimmune diseases (**SLE** or vascular inflammations, **multiple sclerosis**)
- Cerebral lesions (meningioma, metastasis)
- Traumatic brain injury

**Irreversible dementing syndromes**

Irreversible dementing disorders include different diseases that are characterized by structural brain damage. These include:

- **Alzheimer dementia** and **vascular dementia**, the main forms of dementia
Anamnesis of Dementia

Anamnesis, or a patient’s medical history, is also an essential part of diagnosing dementia. Subjective descriptions given by the patient (personal anamnesis), as well as descriptions from close relatives such as a spouse or children can give important clues to the underlying disease. A patient’s medical history should include the following:

- **Primary diseases** (exclusion of a reversible dementing syndrome)
- **Development** (subtle or acute beginning) and symptom expression (e.g., are there impairments in daily living?)
- **Family anamnesis** (Huntington’s disease, Parkinson’s disease)
- **Cardiovascular risk factors**/previous cardiovascular events (vascular dementia)
- **Prior trauma**
- **Attendant symptoms** such as a change in personality (frontotemporal dementia), gait disorder, or extrapyramidal movement disorders (Parkinson’s dementia, Lewy body dementia, subcortical dementia), visual hallucinations (Lewy body dementia), or urinary incontinence (normal pressure hydrocephalus)

**Regular medication** (e.g., drug-induced reversible dementing syndrome due to digitalis or benzodiazepine overdose)

Physical Examination of Dementia

Physical examination includes a thorough internal and neurological examination with a special focus on:

- Possible reasons for a reversible dementing syndrome
- Neurological disorders (reflex status, an examination of brain nerves and motor skills)
- Extrapyramidal movement disorders
- Gait disorders
- Signs of increased intracranial pressure (funduscoppy)

Laboratory diagnostics and further instrument examinations

The following examinations should be part of the standard repertoire of every dementia diagnosis:

- **Lab tests**: blood count, electrolytes, transaminases, bilirubin, gamma-GT, AP, creatinine clearance, urea, vitamin B12, folic acid, glucose, TSH
- **Imaging**: computed tomography or magnetic resonance imaging (MRI) (to uncover possible cerebral tumor, bleeding, atrophy or infraction areas)
Optional examinations should also be carried out depending on the suspected diagnosis. These include:

- Laboratory examinations (e.g., syphilis and borreliosis serology, HIV test, CDT [for suspected alcohol abuse], copper [for Wilson’s disease] and B vitamin test)
- Diagnosis of cerebrospinal fluid (especially in cases of suspected normal pressure hydrocephalus or Creutzfeldt-Jakob disease)
- EEG (repetitive triphasic waves with Creutzfeldt-Jakob disease)

**Approach for the Evaluation of Patients with Cognitive Dysfunction**

The assessment of cognitive function and dementia is often perceived as subjective. Accordingly, a stepwise approach that can help confirm the diagnosis of dementia and exclude the possible causes of potentially reversible dementia is recommended.

**Step 1:**
A detailed history should be taken. The main cognitive domain that is affected must be clearly defined from the history. It is important to document the onset and progression of cognitive decline since dementia is a progressive disease. The severity of functional impairment is also assessed in this step. A history of stroke, malignancy, metabolic derangements, or endocrinopathies needs to be explored. The patient’s medication list should also be noted.

**Step 2:**
A complete physical examination needs to be performed. A neurological examination is also a must for the evaluation of any patient with cognitive dysfunction.

**Step 3:**
Formal cognitive testing should be attempted. This can be done using an MMSE, the Blessed dementia scale, Alzheimer’s disease assessment, or a national adult reading test.

**Step 4:**
All patients presenting with new-onset dementia need to have a complete blood cell count, electrolytes, renal, liver, and thyroid function tests taken, as well as tests for vitamin B12 levels and routine relative peak ratio. These tests can help exclude some common potential causes of reversible dementia such as anemia.

Some patients, such as those with alcohol-use disorders, should also undergo thiamine testing. Patients who partake in high-risk sexual behavior should be given an HIV test. Rheumatologic screening is indicated in patients who have symptoms and signs suggestive of rheumatologic disease.

**Step 5:**
Routine brain imaging is indicated in all patients and should include a noncontrast computed tomography scan or MRI. Vascular dementia can be diagnosed with routine brain imaging.

**Step 6:**
The final step is to refer your patient to a dementia specialist, who will assess the patient again, order a neuropsychological evaluation, arrange an appointment and/or intervention by a social worker, and refer the patient to a geriatric psychiatrist if needed.

Review Questions

The correct answers can be found below the references.

1. What does not belong to the ICD-criteria of dementia?
   - A. Impairment of memory
   - B. Change of emotional control
   - C. Symptoms for at least 6 months
   - D. Affection of the consciousness
   - E. Impairment of thinking, language, and orientation

2. Which score refers to moderate dementia in the MMSE?
   - A. 10 – 19 points
   - B. 5 – 9 points
   - C. 27 – 30 points
   - D. 20 – 26 points
   - E. 14 – 21 points

3. A 63-year old patient with his wife visits a primary physician. The patient seems to be apathetic and is not able to answer questions appropriately. Sometimes he just repeats the asked question and sometimes he looks at his wife in search for help. She reports that her husband has changed over the past few months. At first, she noticed his small step gait just as if his feet were glued to the ground. Afterward, her husband became more and more oblivious, misplaced stuff and did no longer care for his exotic birds, and, for some days now, he was incontinent. What is your suspected diagnosis?
   - A. Parkinson’s disease
   - B. Lewy-Body dementia
   - C. Pick disease
   - D. Creutzfeld-Jakob disease
   - E. Normal pressure hydrocephalus

References

J.Heisel: Neurologische Differentialdiagnostik, Thieme, 2007


Correct answers: 1D, 2A, 3E

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