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COGNITIVE IMPAIRMENT OF DEMENTIAS

ALTERACIONES COGNITIVAS EN LAS DEMENCIAS

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Abstract

Dementia is a clinical syndrome characterized by a loss of cognitive and emotional abilities of sufficient severity to interfere with social or occupational functioning, or both. Although the causes of dementia and characteristics are not always fully understood, it is understood that it is not a natural part of aging. Definitive diagnosis of dementia is made only through the autopsy and although the diagnosis of probable or possible dementia is complex is achieved by the intervention of several specialists in the area, such as neurologists, geriatricians, neuropsychologists etc. Currently it is clear the role of the neuropsychologist and the utility of the assessment of cognitive functions and emotional needs of patients to determine the precise diagnosis. We know that early identification of cognitive disorders and early diagnosis of dementia are important to the development of any potential therapy to delay or prevent altering the progress of the disease. A clinical assessment will confirm whether cognitive impairment stabilizes or progresses to dementia. The aim of this paper is to describe some of the more common dementias and provide information on the differential diagnosis and distinction from other possible causes of cognitive deficits in dementia.

Key words: Dementia, cognitive impairment, neuropsychology.

Resumen

La demencia es un síndrome clínico caracterizado por una pérdida de capacidades cognitivas y emocionales que pueden interferir con la actividad laboral o social o con ambas. Aunque las causas y las características de la demencia no se comprenden en su totalidad, se sabe que esta no es un proceso natural del envejecimiento. El diagnóstico definitivo de la demencia solo se hace a través de
la autopsia, y aunque el diagnóstico de probable o posible demencia es complejo se obtiene gracias a la intervención de varios especialistas en el área, como neurólogos, geriatras, neuropsicólogos etc. Actualmente es evidente el papel del neuropsicólogo y la utilidad que tiene la evaluación de las funciones cognoscitivas y emocionales de los pacientes para determinar el diagnóstico preciso. Se sabe que la identificación temprana de las alteraciones cognoscitivas y el diagnóstico precoz de un cuadro demencial son de gran importancia para el desarrollo de cualquier terapia potencial ya sea prevenir, retrasar o alterar el progreso de la enfermedad. Una correcta valoración clínica es determinante para confirmar si el trastorno cognitivo se estabiliza o, por el contrario, progresa a demencia. El objetivo de este trabajo es describir algunas de las demencias más comunes y proporcionar información sobre el diagnóstico diferencial y la distinción de otras posibles causas de los déficits cognitivos en la demencia. 

Palabras clave: Demencia, deterioro cognitivo, neuropsicología.

Introduction

Dementia, a word that perhaps originated in the 19th century from a Latin word meaning “madness” or being “out of one’s mind,” has long been misunderstood. Originally confused with symptoms now understood to be related to schizophrenia and later incorrectly thought of as being a “normal” occurrence in aging, dementia as a clinical entity is now conceptualized as a syndrome of multiple possible causes that affects one’s memory, thinking, behavior, and overall daily function (Alzheimer’s Disease International, 2010).

Although the causes and characteristics of dementias may not always be fully understood, we understand that it is not a natural part of aging. The American Psychiatric Association describes an official diagnosis of a dementia as having several requirements. First, the individual must experience memory impairment as well as at least one other cognitive disturbance in the form of aphasia, apraxia, agnosia, or disturbance in executive dysfunction. Second, these must be severe enough to cause impairment in daily life and must represent a decline from previous functioning. Lastly, these cognitive deficits must not occur exclusively during the course of a delirium, which we cover later in this chapter (DSM-IV-TR, 2000).

Dementia prevalence rates worldwide are high and a recent report estimates that over 35.5 million people are currently living with dementia and account for a total estimated global cost of US$604 billion (Alzheimer’s Disease International, 2010). Currently, dementia is the fourth leading cause of death in high-income countries (WHO, 2011). The condition in general mostly affects those older than 65, and the likelihood of developing dementia is estimated to nearly double every five years. Estimates vary, but some have reported that dementia affects up a third of the elderly population over the age of 80 (Ritchie & Lovestone, 2002).

The diagnosis of dementia can be difficult to make and most diagnoses of definite dementia are only given at autopsy. However, the need for early diagnosis has been emphasized by research demonstrating how early detection is related to better outcomes as current treatments continue to show most effective when used in the earliest stages of these disorders (Thal, 1999). Moreover, early interventions can help decrease caregiver burden. The increasing utility of cognitive assessment in dementias is evident and most diagnoses are given with the help of a neuropsychological evaluation of the individual’s mental abilities in conjunction with a neurological examination and brain imaging (e.g., Magnetic Resonance Imaging).

The aim of this paper is describe some of the most common dementias and provide information on differential diagnosis and distinction from other possible causes of cognitive deficits.

Dementia or Delirium?
An important question to ask oneself before considering a dementia diagnosis is whether the presenting pattern of symptoms is categorized
better as part of a dementia or as part of a delirium. However, this question is often difficult to answer seeing as both conditions often present very similarly and both can exist simultaneously. However, it is important to note that delirium is considered to be more acute in onset and the severity of daily symptoms fluctuates with deteriorating course. Additionally, the primary cognitive deficit in delirium is attention, which is relatively preserved in most dementias (Meagher et al., 2010).

Alzheimer’s Disease
Of all the dementias, Alzheimer’s disease, or AD, is the most common, accounting for nearly 60-80 percent of all cases of dementia. Most cases are diagnosed after age 65, although about 4-6 percent of cases are diagnosed at a younger age. The disease seems to also affect more women than men (Thies & Bleiler, 2011).

Originally identified by Dr. Alois Alzheimer in 1901, this age-related disease is a degenerative brain disorder that is characterized by the abnormal accumulation of protein fragment plaques of beta-amyloid and tangles of the protein tau that affect association cortices of the frontal, temporal, and parietal lobes as well as in limbic structures (e.g., hippocampus) in the medial temporal lobe (H. Braak & E. Braak, 1991). The deposition of these proteins subsequently destroys synapses between neurons, which seemingly impacts the interaction between various regions of the cortex. For this reason, some consider AD as a “disconnection syndrome” (Salmon & Bondi, 2009). The plaques and tangles lead to necrosis, or cell death, that ultimately results in brain atrophy.

Cognitively, the disease’s pathology relates to the domains that are impacted. Often, the first characteristic trait of AD is a deficit in semantic memory (memory for general impersonal facts, like knowing the direction the earth rotates) and episodic memory (memory for personal information, like one’s birthday) that occurs in the mild stages of the disease. As the disease progresses, abstract reasoning, executive functions, attention, and visuospatial abilities are often at least moderately impaired (Butters, Delis, & Lucas, 1995; Salmon & Bondi, 2009).

Learning and memory deficits seem to be the most significant traits of mild AD and can help differentiate from normal aging. Encoding of new information is often poor, resulting in poor retention and impaired delayed recall. The primacy effect, the recall of words from the beginning of list, is attenuated and new material or information tends to displace other recently learned information (Salmon & Bondi, 2009). There is evidence, though, that at least in the earlier stages of the disease remote events (e.g., what one did several decades ago) are remembered better than recent events (Butters et al., 1995). Lastly, AD patients seem to be more likely than healthy patients to produce intrusion errors on memory tests when attempting to recall new material (Salmon & Bondi, 2009).

AD is related to behavioral changes in patients as well. High rates of lack of concentration, tremors, depression, and lack of cooperation have been seen in this population (Fernández, Gobartt, & Balañá, 2010), which have been related to further disability in demented patients and to increased caregiver stress (Swearer, Drachman, O’Donnell, & Mitchell, 1988). Additionally, paranoid delusions are common, usually related to thoughts of theft resulting from forgetful misplacing of items and to the reduced recognition of family members, and anxiety related to fears of separation or abandonment (Levy & Chelune, 2007).

As with most dementias, early detection is of high clinical interest in Alzheimer’s disease. A large area of study has focused on the “preclinical” phase of the disorder since mild deficits can sometimes be identified three to four years prior to diagnosis (Salmon & Bondi, 2009). A separate clinical entity named Mild Cognitive Impairment (MCI) has been used to classify the intermediate phase between normal aging and dementia before objective cognitive deficits have impact on daily life (Petersen et al., 2001, 1999). Progression to dementia within a few years of an MCI diagnosis seems to occur in about half of cases with up to 25% of MCI cases progressing to dementia within one year (Eschweiler, Leyhe, Klöppel, & Hüll, 2010).

Vascular Dementia
Sometimes referred to as multi-infarct dementia, vascular dementia (VaD) is arguably the second-most common type of dementia, accounting for roughly 15-29% of dementia cases (Mendez et al., 1996; Ott et al., 1995) and rates rise steeply with
Much like MCI in AD, VaD is often preceded by a milder subclinical form, often referred to as Vascular Mild Cognitive Impairment, or VaMCI (Hatcher, 1999).

Dementia with Lewy Bodies
Comparable to VaD prevalence rates is Dementia with Lewy Bodies, or DLB, which accounts for somewhere between 10 and 15% of dementias cases (McKeith et al., 1996). Diagnoses tend to occur in late middle age and old age and the disease seems to be slightly more common in men than in women (Nelson et al., 2010).

Lewy bodies were first described in 1914 by Dr. Frederick Lewy in patients with Parkinson's disease. In DLB, these cytoplasmic inclusion bodies, which consist of \( \alpha \)-synuclein and ubiquitin, are typically found in the brainstem, limbic structures, and neocortex (Levy & Chelune, 2007; Andersson, Zetterberg, Minthon, Blennow, & Londos, 2011). These bodies generally accumulate in parietal and occipital regions, causing catastrophic loss of cholinergic and dopaminergic pathways to these regions (Levy & Chelune, 2007). Although Lewy bodies were originally seen in Parkinson's disease, DLB is now identified as a separate disease that is characterized by fluctuating cognition, visual hallucinations, and parkinsonism (McKeith et al., 1996). The time of onset of cognitive symptoms in relation to motor symptoms is used to differentiate between Parkinson's disease and DLB (Andersson et al., 2011), where cognitive deficits must either present in advance or concomitantly with motor symptoms for a DLB diagnosis (Levy & Chelune, 2007). It is important to note, however, that Lewy bodies can be found in other disorders, including AD.

DLB can often be confused with AD, making a careful cognitive assessment crucial for differential diagnosis. The cognitive profile of DLB is associated with impairment in both spatial and perceptual networks of visual information processing (Collerton, Burn, McKeith, & O'Brien, 2003), probably due to the accumulation of Lewy bodies in brain regions that are associated with these processes. Therefore, DLB patients consistently demonstrate greater impairment in attention than AD patients as well as greater impairment on visuospatial and constructional tasks. Patient with DLB generally
perform better on tests of episodic memory than AD patients (Oda, Yamamoto, & Maeda, 2009), but show a pattern of poor initial learning of new material (Levy & Chelune, 2007).

Behavioral changes are common in DLB and psychotic symptoms are more common than in AD, with visual, auditory, and olfactory hallucinations as well as delusions being the most frequent (Levy & Chelune, 2007). Unlike AD, delusions in DLB are less associated with memory loss and more associated with visual hallucinations and perceptual problems. However, the rate of psychotic symptoms becomes equivalent in both conditions with increasing dementia severity (Simard, van Reekum, & Myran, 2003).

**Frontotemporal Dementia**

Less common, but accounting for up to 10% of dementia cases, is Frontotemporal Dementia, or FTD. Once called Pick’s disease, FTD is now understood to encompass several distinct conditions, of which Pick’s disease is one. Onset of the disease occurs on average between 52 and 56 (Boxer & Miller, 2005) and seems to be more prevalent in men than in women (Westbury & Bub, 1997).

The first clinical case of FTD was identified in 1892 by Dr. Arnold Pick, after whom the cell bodies of pathological tau protein seen in Pick’s disease were named. Since then, FTD has been described as a progressive dementia that is due to specific atrophy of the frontal and anterior temporal lobes. The causes and pathology vary depending on the kind of FTD, but the disease is generally categorized as either impacting mostly behavior (e.g., frontal variant) or language (e.g., fluent variant, non-fluent variant) (Dickson, 1998).

The clinical presentation of each of the variants of FTD is unique, but the most common variant typically begins with the insidious onset of personality and behavioral changes such as inappropriate social conduct, apathy, disinhibition, perseverative behavior, loss of insight, hyperorality (with an increased craving for carbohydrates and sweets in particular), and decreased speech output (Levy & Chelune, 2007; Salmon & Bondi, 2009). These changes occur in the presence of or just before cognitive changes in executive function, attention, and or language. Memory and visuospatial abilities appear to be mostly spared (Boxer & Miller, 2005). Due to the location of the disease pathology in the frontal lobe, executive dysfunction disproportionate to deficits in other domains as seen in FTD helps differentiate it from other dementias (Levy & Chelune, 2007; Salmon & Bondi, 2009). Diagnosis of FTD is usually given with the help of both a cognitive assessment and neuroimaging that support this focalized impact on the frontal lobe.

FTD is largely diagnosed based on behavioral criteria. Rates of apathy, disinhibition, euphoria, and aberrant motor behavior are higher in FTD than in AD (Levy & Chelune, 2007). Rates of other psychiatric features like delusions, hallucinations, depression, anxiety, agitation, and irritability are similar in both disorders, but the depression observed in FTD is associated with irritability/agitation without dysphoria or anhedonia (Levy & Chelune, 2007).

**Other Causes of Cognitive Decline**

Besides the dementing disorders described in this paper as well as the occurrence of “mixed dementia,” in which deficits are attributable to a combination of various pathological origins, cognitive decline can be associated with various other causes. Therefore, it is important to consider these other causes when evaluating a patient for dementia. There are numerous alternate causes ranging from infections to traumatic brain injury, but for the purposes of this article we consider a few common causes.

**Huntington’s Disease**

Huntington’s disease (HD), a rare genetic disorder that causes deterioration of subcortical structures in the neostriatum (caudate nucleus and putamen), is typified by movement disorder, behavioral changes, and dementia. Characteristically, HD dementia is associated with slowness of thought, impaired attention, executive dysfunction, poor learning, as well as visuoperceptual and constructional deficits. Due to systematic retrieval deficits, HD patients can demonstrate mild to moderate memory impairment. Working memory, planning, and strategy use are also affected early in the disorder due to the disease’s pattern of deterioration (Salmon & Bondi, 2009).
Alcohol Abuse
Extended use and abuse of alcohol has been related to multi-systemic changes that can also affect cognition. Alcoholism is related to greater dysfunction in older persons than in younger individuals and there is evidence of possible improvement in cognitive functioning after at least five years of sobriety. Mild to moderate deficits are often seen on more challenging tests of new learning and memory, conceptualization, and problem-solving, but greater dysfunction in visuospatial skills relative to verbal abilities appears to be one of the more notable traits in these patients (Butters et al., 1995).

Depression
A common occurrence in aging populations is geriatric depression. Although often comorbid with dementia, depressive symptoms can worsen cognitive symptoms as well as account for some cognitive complaints. Depression most commonly accounts for deficits in attention, concentration, and memory that are correlated to the severity of the depressive symptoms (Braaten, Parsons, McCue, Sellers, & Burns, 2006). However, while increased subjective memory complaints are related to depressive symptoms, research demonstrates variable incidence and severity of objective cognitive symptoms (Fischer et al., 2008).

Summary
The diagnosis of dementia requires a decrease in memory or intellectual skills to alter the activities of daily living. The clinical uses the mental status assessment to distinguish the changes of dementia from those associated with delirium, “normal” aging, and other conditions. The mental status assessment further characterizes the specific deficits of the dementia and may disclose patterns of neurobehavioral deficits that suggest specific dementing disease. Clinical and experimental neuropsychology has made considerable progress and advances in the differentiation cognitive changes related to the onset of dementia. Although not always an easy task, comprehensive neuropsychological assessment has proven to be crucial in both the diagnosis and early intervention of dementing disorders. The diagnosis and treatment of dementias can often be complicated and can involve multiple sources of support and involvement. Therefore, a collaborative team that integrates the perspective of an effective neuropsychologist with that of the patient’s medical provider (e.g., neurologist) and information from the patient’s neuroimaging is crucial.

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