
CLINICIAN'S PERSPECTIVE

Clinical Diagnosis of Dementia: Core Clinical Features and Diagnostic Criteria

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ABSTRACT

In dementia of the elderly, the majority of patients with onset around the eighth decade have typical Alzheimer's disease; diagnostic workup is usually simple, and a plain computed tomography brain is sufficient to exclude pathology other than generalised brain atrophy. Advanced neuroimaging is often required when onset occurs at a younger age to confirm a major neurocognitive disorder and non-Alzheimer's disease. The prognosis and trajectory of the latter differs to that of Alzheimer's and, because of the younger age, has important psychosocial implications. Particular clinical features often suggest a non-Alzheimer's diagnosis, and although clinical criteria play a vital role, functional neuroimaging helps reach a definitive diagnosis. A simplified diagnostic chart at the end of this article aims to assist such diagnosis.

中文摘要

老年癡呆症的臨床診斷：主要臨床特徵和診斷標準

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大多數發病年齡在80歲左右的老年癡呆症患者會有典型病徵，其診斷檢查通常較為簡單，只需進行一項簡單的電腦斷層掃描，並排除是否罹患典型腦萎縮以外的病變便可。而年輕發病的患者往往須要較先進的腦神經影像學檢查，以確認主要病因是神經認知障礙或非阿爾茨海默氏病。非阿爾茨海默氏病的預後和病程發展與阿爾茨海默氏症不同，非阿爾茨海默氏病的年輕患者會承受很大的社會心理壓力。特殊的臨床表現通常傾向於非阿爾茨海默氏症的診斷。需承認臨床表現對診斷很重要，但腦功能影像學檢查卻能使診斷更為明確。本文結尾一個簡化圖表將有助對阿爾茨海默氏症進行診斷。

INTRODUCTION

Hong Kong has a rapidly ageing population. By 2041, 30% of the population will be aged 65 years or above,¹ and a significant proportion of these elderly will suffer from dementia. Previous local studies have estimated

the prevalence of dementia in the community to be about 10% for those aged 75 to 79 years; 19% for those aged 80 to 84 years; and 32% for those aged 85 years and above.^{2,3} Nonetheless, limited recognition of early symptoms and limited access to diagnostic services

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with long waiting queues means that only 11% of those affected in the community have a formal medical diagnosis.

A diagnostic consultation for suspected dementia should aim to establish a clinical diagnosis of the type of dementia and exclude obvious non-dementing illnesses. Timely diagnosis allows commencement of effective symptomatic treatment (drug and non-drug) for different types of dementia, such as the use of cholinesterase inhibitors to retard disease progression in Alzheimer's disease (AD). It also facilitates advance care planning while the patient has remaining mental capacity. From a health economic perspective, prompt diagnosis is cost saving as it minimises delayed institutionalisation and enhances quality of life for patients and their caregivers.⁴

Clinical differentiation between different types of dementia may sometimes be difficult due to overlapping clinical features. Consequently, increasing attention is placed on the role of early neuroimaging to identify characteristic patterns of brain involvement to narrow down the list of differential diagnoses. A deeper understanding of the more common types of dementia and a systematic and meticulous approach to identifying the dominant pattern of lobar atrophy / hypometabolism on neuroimaging will facilitate the clinician and radiologist alike in arriving at a clinical diagnosis earlier on in the course of disease. We summarise here the major clinical features of the more common forms of dementia.

COMMON FORMS OF DEMENTIA

Alzheimer's Disease

AD⁵ is the most common form of dementia with a prevalence that increases with age. In older age-onset dementia (aged ≥ 65 years), AD accounts for about 40% of cases. Early-onset AD (aged < 65 years) is familial in 60% of cases and constitutes approximately 1% to 6% of all AD patients. Patients present with impaired memory and executive function early in the disease course that progressively becomes incapacitating. Language is typically affected later on. Death usually results from general inanition, malnutrition, and pneumonia. The diagnosis of AD is largely clinical, but other neurodegenerative diseases such as frontotemporal dementia (FTD) and dementia of Lewy body (DLB) may be difficult to differentiate from AD based on clinical findings alone.

Frontotemporal Dementia

There are two variants of FTD⁶; the frontal variant

(fvFTD) that is characterised by behavioural disturbance and impaired executive function, and the temporal variant (tvFTD) that is characterised by speech impediments. Behavioural symptoms in FTD are related to frontal lobe dysfunction and may manifest as disinhibition, impulsivity, loss of personal and social awareness, and hyperorality. Recently, tvFTD has been further described as a new entity known as primary progressive aphasia (PPA).

Primary Progressive Aphasia

PPA⁷ has recently emerged as a separate entity to the tvFTD. PPA is characterised by relatively isolated language impairment with generally intact function in other cognitive abilities and memory. There are three subtypes of PPA — logopenic, agrammatic and semantic variants — and each corresponds to a distinctive pattern of brain atrophy. The logopenic variant is characterised by word-finding difficulty and decreased output but relatively preserved syntax, grammar, and comprehension ability. The agrammatic (also known as progressive non-fluent aphasia) variant is characterised by stuttering oral apraxia, impaired repetition and agrammatism, but relatively preserved word comprehension. The semantic variant is characterised by fluent, grammatically correct speech with loss of word and object meaning but relatively preserved syntactic comprehension skills.

Corticobasal Degeneration

Corticobasal degeneration⁸ (CBD) is a progressive asymmetrical movement disorder characterised by extrapyramidal features (such as rigidity, dystonia, and akinesia) and cortical motor dysfunctions (such as apraxia, alien limb phenomenon, myoclonus, cortical sensory loss). Cognitive decline is also common in CBD and can occasionally be the presenting feature of the disease.

Progressive Supranuclear Palsy

Progressive supranuclear palsy⁹ (PSP) is characterised by vertical gaze palsy, postural instability and falls, parkinsonian features, pseudobulbar palsy, and cognitive impairment. Clinical differentiation of PSP from Parkinson's disease and other atypical parkinsonian syndromes such as multiple system atrophy (MSA) and CBD may be difficult during the early course of disease.

Multiple System Atrophy

MSA¹⁰ is characterised by parkinsonian features, cerebellar dysfunction and autonomic disturbance, and

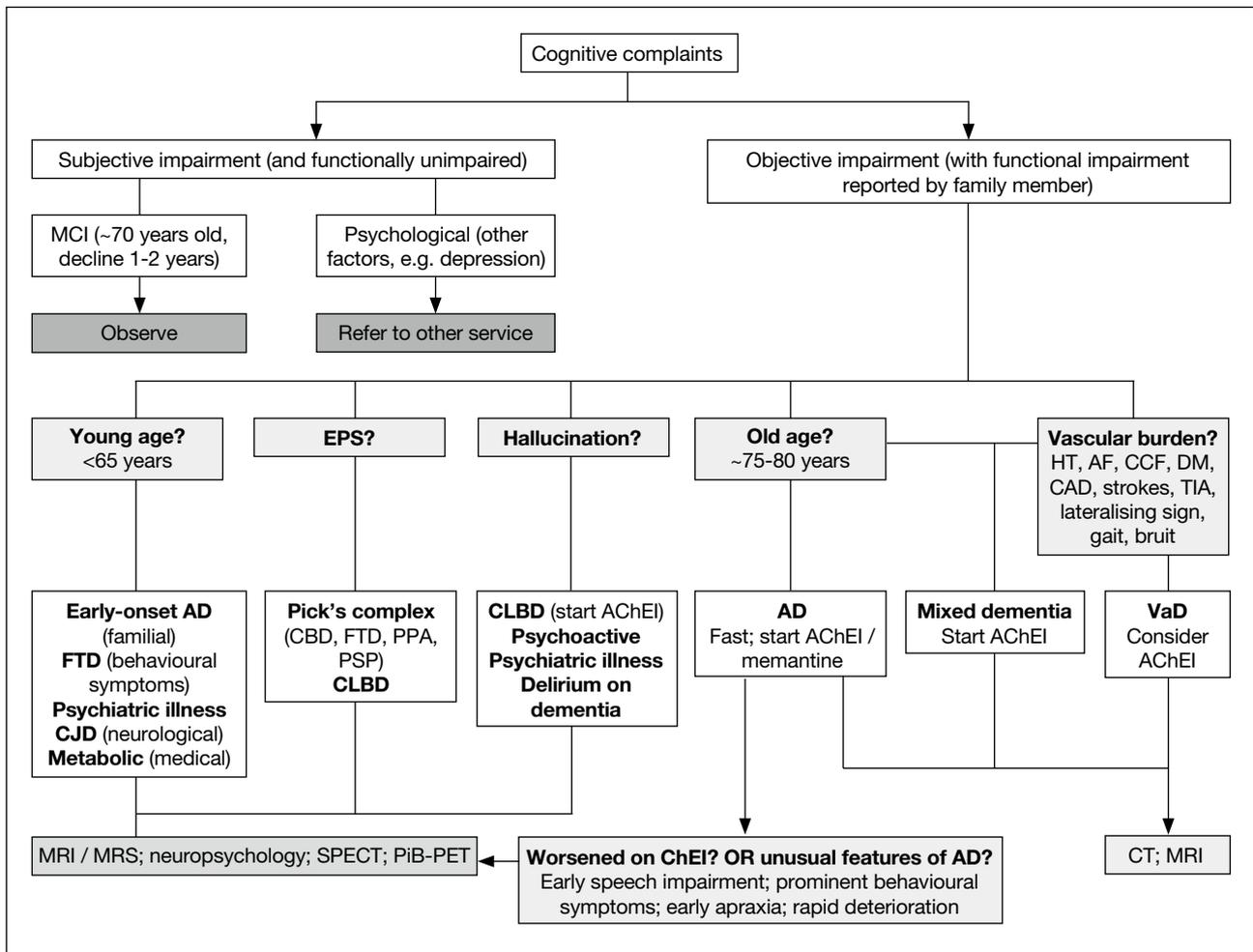


Figure. Diagnostic pathway for cognitive impairment.

Abbreviations: AChEI = acetylcholinesterase inhibitors; AD = Alzheimer's disease; AF = atrial fibrillation; CAD = coronary artery disease; CBD = corticobasal degeneration; CCF = chronic cardiac failure; ChEI = cholinesterase inhibitors; CJD = Creutzfeldt-Jakob disease; CLBD = cerebral Lewy body disease; CT = computed tomography; DM = diabetes mellitus; EPS = extrapyramidal symptoms; FTD = frontotemporal dementia; HT = hypertension; MCI = mild cognitive impairment; MRI = magnetic resonance imaging; MRS = magnetic resonance spectroscopy; PiB-PET = Pittsburgh compound B positron emission tomography; PPA = primary progressive aphasia; PSP = progressive supranuclear palsy; SPECT = single-photon emission computed tomography; TIA = transient ischaemic attack; VaD = vascular dementia.

is divided into two main clinical phenotypes depending on the initial and predominant symptom. MSA-P presents with predominant parkinsonian features and few, if any, cerebellar signs. MSA-C presents with predominant cerebellar dysfunction and fewer parkinsonian features.

Dementia of Lewy Body

DLB¹¹ is the second most common type of dementia following AD. It is characterised by parkinsonian features, complex visual hallucinations, neuroleptic sensitivity, and rapid-eye-movement sleep behavioural disorder. Attention deficits, daytime somnolence and apathy, disproportionate executive and visuospatial

impairment, relative preservation of episodic memory, dysautonomia (early urinary incontinence, dizziness, and falls) and good cholinesterase inhibitor response are also indicators for a diagnosis of cerebral Lewy body disease.¹² The diagnosis of DLB should be considered when patients present with parkinsonism within 1 year of the onset of dementia or if dementia is established within 1 year of the onset of parkinsonism, hence called the 1-year rule. This distinguishes the condition from dementia in idiopathic Parkinson's disease that usually occurs in later stages of Parkinson's disease.¹³

The diagnostic pathway outlined in the Figure may serve as a guide in diagnosing different types of dementia.

CONCLUSION

AD is the most common type of dementia, but there are many types of atypical dementia that present with non-cognitive features such as speech and language impediments. Clinical differentiation between different types of dementia may be difficult in the early course of disease due to significant clinical overlap. Nonetheless, neuroimaging can frequently aid the clinician in arriving at a specific diagnosis and restrict the list of differential diagnoses. An understanding of the key clinical features of these diseases, close collaboration with the radiologist, and detailed analysis of the pattern of cerebral atrophy and hypometabolism on imaging often help to resolve the clinical challenge of diagnosing dementia.

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