Klinične in psihometrične razlike med frontotemporalno demenco in Alzheimerjevo boleznijo

Clinical and Psychometric Differences between Frontotemporal Dementia and Alzheimer's disease

Rajka M. Liščić*, Martha Storandt**, Nigel J. Cairns***, John C. Morris****

Izvleček

Frontotemporalna demenca (FTD) je glavni vzrok ne-Alzheimerjeve demence pri mlajših od 65 let. Predvsem v zgodnjih fazah bolezni jo lahko klinično zamenjamo za AD. Delu bolnikov, ki dosegajo kriterije NINCDS - ADRDA za AD, obdukcija potrdi FTD. Retrospektivni pregled 48 nevropatološko potrjenih primerov FTD (27 jih je opravilo psihometrična testiranja) je omogočil primerjavo s 27-imi posamezniki z AD, izenačenimi po starosti, spolu, izobrazbi in stopnji težav. Ob prvem obisku se je FTD razlikoval od AD po vedenjskih spremembah, zlasti po impulzivnosti ($p<0.0001$) in dezinhibiciji ($p<0.0004$); po manjšem deležu socialnega umika ($p<0.01$) in po progresivni nefluentni afaziji. V obeh kliničnih fenotipih so bile motnje izvršilnih funkcij in spominske težave primerljive. Posamezniki s FTD so bolje kot tisti z AD opravili vidni test epizodičnega spomina ($p<0.05$), a slabše test besedne fluentnosti, ki je občutljiv znak frontalne disfunkcije ($p<0.05$) (uspešnost je povezana z afazičnimi značilnostmi). Histopatološko je bil AD prisoten pri 11/48 posameznikih s FTD. Klinične in kognitivne značilnosti FTD se lahko prekrivajo s tistimi pri AD, zlasti pri spominskih in izvršilnih funkcijah, vedenjske in govorne težave pa razlikujejo tiste s FTD. Delno

*Rajka M. Liščić, Washington University School of Medicine, St Louis, Missouri, USA; Institute for Medical Research and Occupational Health, Zagreb, Croatia

**Martha Storandt, PhD, Washington University School of Medicine, St Louis, Missouri, USA

***Nigel J. Cairns, PhD, MRCPath, Washington University School of Medicine, St Louis, Missouri, USA

****John C. Morris, MD, Washington University School of Medicine, St Louis, Missouri, USA

Acknowledgments: Supported by Fulbright Grant 68428174 (RML)

so lahko težave s spominom pri FTD posledica težav z iskanjem besed, ki so del govornih motenj.

Abstract

Frontotemporal dementia (FTD) is a major cause of non- Alzheimer's dementia (AD) in individuals younger than 65 years of age, and may be clinically mistaken for AD, especially in early stages of disease. A proportion of patients who meet the NINCDS-ADRDA criteria for AD have FTD confirmed at autopsy. A retrospective review of 48 neuropathologically confirmed cases of FTD (27 had completed psychometric testing) yielded clinical and psychological features for comparison with 27 age-, sex-, education-, and severity-matched individuals with AD. At first visit FTD was distinguished from AD by behavioral abnormalities, particularly impulsivity ($p< 0.0001$) and disinhibition ($p =0.0004$), less social withdrawal ($p=0 .01$), and progressive nonfluent aphasia. The two clinical phenotypes reported comparable executive dysfunction and memory problems. The FTD individuals performed better than AD individuals on a visual test of episodic memory ($p<$

0.05) but worse on word fluency, sensitive to frontal lobe dysfunction ($p<0.05$) (performance correlated with aphasic features). Histopathological AD was present in 11 of the 48 FTD individuals. Clinical and cognitive features of FTD may overlap with AD, particularly for memory and executive function, although behavioral and language difficulties distinguish those with FTD. Part of the complaints about memory in FTD may reflect word-finding difficulties stemming from language dysfunction.

INTRODUCTION

Frontotemporal dementia (FTD) is the second most common form of dementia in people under the age of 65 years after Alzheimer's disease (AD) (1). FTD refers to heterogeneous group of focal dementias affecting behavioral and personality changes, social functioning, language impairment, motor functioning, and later in the course of the disease memory. These disorders may be mistaken for AD, particularly in the early stage of disease. According to Neary et al. criteria (2) FTD encompasses three clinical syndromes: frontal variant FTD, progressive nonfluent aphasia, and semantic dementia with heterogeneous neuropathology (3). The frontal or behavioral variant FTD is associated with behavioral abnormalities, including disinhibition, impulsivity, apa-