PROGRESSIVE SEMANTIC DEMENTIA

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April 8, 2026

Progressive Semantic Dementia: A Review

Abstract

Progressive semantic dementia (PSD) is a rare form of frontotemporal dementia, which is characterized by progressive changes in language, semantics, and conceptual thinking. This review summarizes current literature on PSD, including epidemiology, clinical presentation, diagnosis, and treatment options. In addition, the review discusses the pathology of PSD, its genetic basis, and its neuroimaging findings. Finally, this review highlights the importance of early diagnosis of PSD and suggests future research directions.

Introduction

Frontotemporal dementia (FTD) is a group of neurodegenerative diseases characterized by changes in behavior, personality, language, and executive functions. Progressive semantic dementia (PSD) is one of the subtypes of FTD, and it is characterized by progressive changes in language, semantics, and conceptual thinking. It is a rare form of FTD, with an estimated prevalence of 0.2-1.9% (Mendez et al., 2008). The diagnosis of PSD is challenging due to its atypical presentation and the lack of specific biomarkers. Early diagnosis is important to ensure access to appropriate medical care and management of the disease. This review provides an overview of PSD, including epidemiology, clinical presentation, diagnosis, and treatment options. In addition, the review discusses the pathology of PSD, its genetic basis, and its neuroimaging findings.

Epidemiology

The estimated prevalence of PSD ranges from 0.2-1.9%, and it is more common in males than females. The mean age of onset is 59 years, and the mean duration of the illness is 7 years (Mendez et al., 2008).

Clinical Presentation

The clinical presentation of PSD is variable and can include deficits in language, semantics, conceptual thinking, and executive functions. Language deficits include anomia, impaired comprehension of spoken and written language, and impaired verbal fluency. Semantic deficits include difficulty understanding language, impaired semantic memory, and impaired word meaning. Conceptual deficits include impaired ability to make abstract connections, difficulty understanding metaphors, and impaired judgment. Executive deficits include impaired problem solving, impaired planning and organization, and impaired abstract thinking.

Diagnosis

The diagnosis of PSD is based on clinical criteria and is made by a neurologist or psychiatrist. Clinical criteria include a progressive deterioration in language, semantics, and conceptual thinking, with relative sparing of other cognitive functions. Neuropsychological testing may be used to assess semantic deficits and executive functions. Neuroimaging studies, such as structural MRI or FDG-PET, may be used to confirm the diagnosis.

Treatment

There is no cure for PSD, and treatment is focused on symptomatic management. Antipsychotics, antidepressants, and cholinesterase inhibitors may be used to manage behavioral and cognitive symptoms. Speech-language therapy may be used to improve language and communication. Physiotherapy and occupational therapy may be used to improve motor function.

Pathology

The pathology of PSD is characterized by atrophy in the frontal and temporal lobes, and the amygdala and hippocampus. In addition, there is a loss of neurons and synapses in the frontal and temporal lobes. There is also an accumulation of tau protein and TDP-43 protein in the brain.

Genetic Basis

The genetic basis of PSD is unknown, but mutations in the PGRN and GRN genes have been associated with the disease. In addition, mutations in the C9ORF72 gene have been associated with the disease.

Neuroimaging Findings

Neuroimaging studies may be used to confirm the diagnosis of PSD. Structural MRI studies typically show frontal and temporal lobe atrophy. FDG-PET studies may show hypometabolism in the frontal and temporal lobes.

Conclusion

PSD is a rare form of FTD characterized by progressive changes in language, semantics, and conceptual thinking. Early diagnosis of PSD is important to ensure access to appropriate medical care and management of the disease. This review provides an overview of PSD, including epidemiology, clinical presentation, diagnosis, and treatment options. In addition, the review discusses the pathology of PSD, its genetic basis, and its neuroimaging findings. Future research should focus on identifying biomarkers for early diagnosis, as well as on developing effective treatments for PSD.

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