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Early-Onset Corticobasal Degeneration: A Case of Rapidly Progressive Dementia with primary Progressive Aphasia and Severe Behavioral Disturbances

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Abstract

Corticobasal degeneration (CBD) is a rare neurodegenerative disorder with diverse clinical manifestations, often overlapping with other conditions like Alzheimer's and frontotemporal dementia. We present the case of a 35-year-old male with early-onset CBD, characterized by rapidly progressive dementia, primary progressive aphasia, and severe behavioral disturbances. Initial imaging revealed left temporal lobe atrophy and right vertebral artery hypoplasia, progressing to significant cortical atrophy in the temporal and frontal lobes within a year. Despite extensive diagnostic evaluations and symptomatic treatments, including Donepezil and Memantine, the patient's condition deteriorated rapidly. This case underscores the challenges of diagnosing and managing CBD, particularly in young patients, and highlights the urgent need for improved diagnostic tools and effective therapies.

KeyWords: Corticobasal degeneration, early-onset dementia, primary progressive aphasia, neurodegenerative disorders, frontotemporal lobar degeneration, cognitive decline

Introduction

Corticobasal degeneration (CBD) is a rare, progressive neurodegenerative disorder characterized by cortical and basal ganglia dysfunction, often presenting with atypical parkinsonism, apraxia, and cognitive decline. Although commonly associated with motor and behavioral symptoms, its clinical manifestations are diverse and can overlap with other neurodegenerative diseases, such as Alzheimer's

disease (AD), frontotemporal dementia (FTD), and primary progressive aphasia (PPA) (1). The underlying pathology of CBD is primarily linked to the accumulation of 4R tau protein, classifying it as a specific tauopathy (2).

CBD is a challenge for clinicians due to its rarity, the heterogeneity of its clinical presentations, and the overlap of symptoms with other neurodegenerative disorders. Furthermore, its early diagnosis is complicated by the lack of specific biomarkers and reliance on clinical assessment, imaging, and exclusion of other conditions (1). Dementia, a core feature of advanced CBD, is a devastating syndrome that affects memory, cognition, and social behavior. While the prevalence of dementia is high in older populations, its onset at 35 years, as in this case, is extremely rare, making it significant for medical research and clinical understanding (3).

This case report highlights a unique presentation of CBD with early onset, rapidly progressive dementia, and atypical features such as primary progressive aphasia, aggressiveness, and severe behavioral disturbances. The absence of genetic or environmental risk factors further underscores its unusual nature. Despite extensive diagnostic evaluations, including advanced imaging and biochemical tests, the patient demonstrated rapid neurocognitive decline unresponsive to current treatment strategies. This case underlines the need for better diagnostic tools and therapeutic interventions for CBD and similar neurodegenerative syndromes.

Case Report

A 35-year-old male presented in 2021 with mild symptoms, including headaches, memory loss, dizziness, balance disturbances, numbness in the right arm and leg, mild speech difficulties, and occasional aggressiveness. The symptoms progressed over time, prompting further investigation. Initial neurological evaluations were inconclusive, but advanced imaging revealed gross atrophic changes in the cerebral cortex.

Imaging and Diagnostic Findings

Initial MRI conducted in 2021 (Figure 1) revealed significant atrophic changes in the ventral structures of the left temporal lobe, alongside right vertebral artery hypoplasia. Despite these findings, there was no failure in intermediate brain structures, and the sellar area, basicranium, retrobulbar tissues, and cerebellopontine angles were unremarkable. Internal carotid arteries appeared normal without any visible pathology. At this stage, the patient exhibited symptoms consistent with primary progressive aphasia, which were associated with severe neurocognitive impairment.

In 2022, follow-up imaging (Figure 2) demonstrated a marked progression of cortical atrophy. The gyri of the left temporal and frontal lobes appeared significantly thinner compared to the 2021 MRI, and

subarachnoid spaces were noticeably expanded. These progressive atrophic changes were more consistent with Frontotemporal Lobar Degeneration (FTLD).

By 2022, the patient's condition had deteriorated significantly, as evidenced by a decline in cognitive and behavioral function. At the first consultation, his Mini-Mental State Examination (MMSE) score was 23, indicating mild cognitive impairment. However, within a year, his cognitive and behavioral changes were so severe that he was uncooperative and unable to complete the MMSE.

A comprehensive diagnostic workup was performed to rule out other causes of dementia. Tests for syphilis (Anti-TP), HIV (Anti-HIV ½), and herpes simplex virus (HSV 1/2) were negative. Cerebrospinal fluid (CSF) analysis revealed normal concentrations of amyloid beta protein (420 pg/mL) and tau protein (260 pg/mL), which are often abnormal in Alzheimer's disease. MRI imaging confirmed progressive atrophy in the temporal and frontal lobes.

Management and Progression

The patient exhibited progressive neurocognitive and functional decline despite pharmacological interventions, including Donepezil, Memantine, and Citicoline. Psychiatric symptoms, including aggressiveness, were managed with sedatives like Sedarex and Grandaxin. However, the treatments only provided symptomatic relief without halting disease progression.

Discussion

This case underscores the complexity of diagnosing and managing rare neurodegenerative disorders such as CBD. The early onset of dementia at 35 years is atypical and poses diagnostic challenges (3). CBD's clinical presentation is heterogeneous, often overlapping with other neurodegenerative conditions, including Alzheimer's disease, Parkinson's disease, and frontotemporal dementia (1). In this case, the patient's symptoms, such as primary progressive aphasia and frontal lobe atrophy, further complicated the diagnosis.

The normal amyloid beta and tau protein levels in CSF argue against Alzheimer's pathology, while the absence of genetic predisposition or infectious causes suggests a sporadic form of CBD (4). Progressive cortical atrophy seen on MRI is consistent with CBD, but the absence of definitive biomarkers highlights the need for more precise diagnostic tools.

Pharmacological management in neurodegenerative diseases remains largely symptomatic (5). Donepezil and Memantine, commonly used for Alzheimer's disease, aim to improve cognitive function but are not curative (6). Similarly, Citicoline may enhance neuronal repair, but its efficacy in halting disease progression is unproven (7). The lack of response to these treatments in this case reflects the limitations of current therapeutic options for CBD and related disorders.

The rapid progression of the disease in this patient, despite the absence of known risk factors, highlights the aggressive nature of early-onset CBD. Behavioral changes, including aggression, further complicate management, emphasizing the need for multidisciplinary care. This case illustrates the importance of early recognition and reporting of such atypical presentations to enhance understanding of CBD's natural history and inform future diagnostic and therapeutic strategies.

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Conflict of Interest

No conflicts of interest.

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Consent

Informed consent was obtained from the patient's family for the publication of this case report.

References:

1. Constantinides VC, Paraskevas GP, Paraskevas PG, Stefanis L, Kapaki E. Corticobasal degeneration and corticobasal syndrome: A review. *Clin Park Relat Disord*. 2019;1:66-71. Published 2019 Aug 30. doi:10.1016/j.prdoa.2019.08.005
2. Kouri N, Whitwell JL, Josephs KA, Rademakers R, Dickson DW. Corticobasal degeneration: a pathologically distinct 4R tauopathy. *Nat Rev Neurol*. 2011;7(5):263-272. doi:10.1038/nrneurol.2011.43
3. Mendez MF. Early-onset Alzheimer Disease and Its Variants. *Continuum (Minneap Minn)*. 2019;25(1):34-51. doi:10.1212/CON.0000000000000687
4. Jack CR Jr, Bennett DA, Blennow K, et al. A/T/N: An unbiased descriptive classification scheme for Alzheimer disease biomarkers. *Neurology*. 2016;87(5):539-547. doi:10.1212/WNL.0000000000002923
5. Arvanitakis Z, Shah RC, Bennett DA. Diagnosis and Management of Dementia: Review. *JAMA*. 2019;322(16):1589-1599. doi:10.1001/jama.2019.4782

6. Chu LW. Alzheimer's disease: early diagnosis and treatment. *Hong Kong Med J*. 2012;18(3):228-237.
7. Bonvicini M, Travaglini S, Lelli D, Antonelli Incalzi R, Pedone C. Is Citicoline Effective in Preventing and Slowing Down Dementia?-A Systematic Review and a Meta-Analysis. *Nutrients*. 2023;15(2):386. Published 2023 Jan 12. doi:10.3390/nu15020386

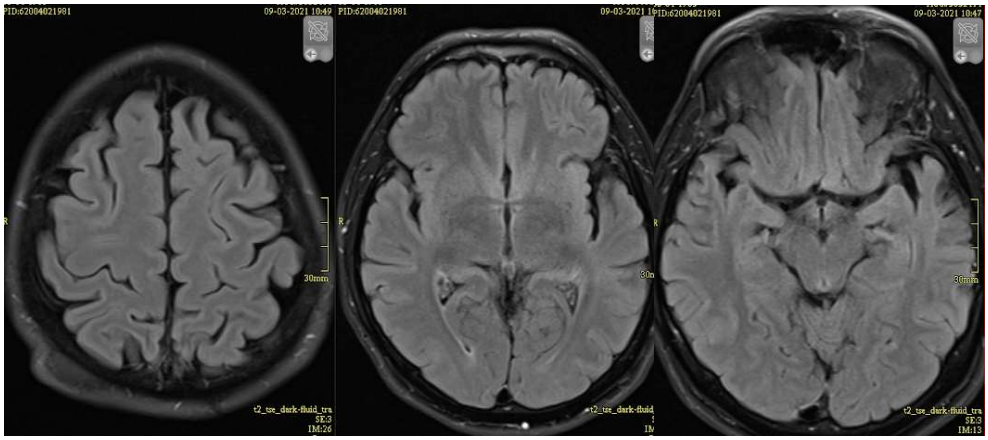

Figure 1: MRI Findings (2021): Initial Atrophic Changes in the Left Temporal Lobe

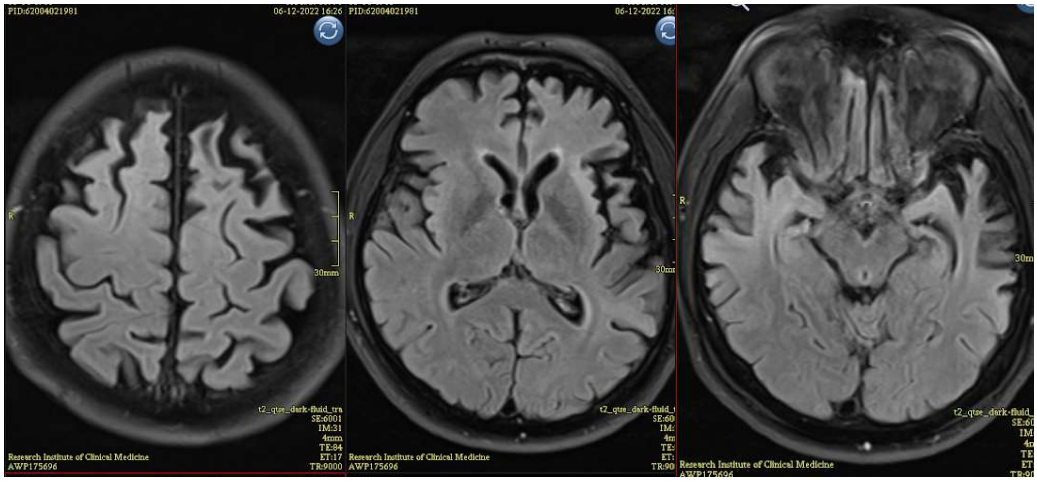

Figure 2: MRI Findings (2022): Progressive Atrophy in the Left Temporal and Frontal Lobe