Anti-Caspr2 Encephalitis Initially Misdiagnosed as Frontotemporal Dementia: A Case Report

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INTRODUCTION

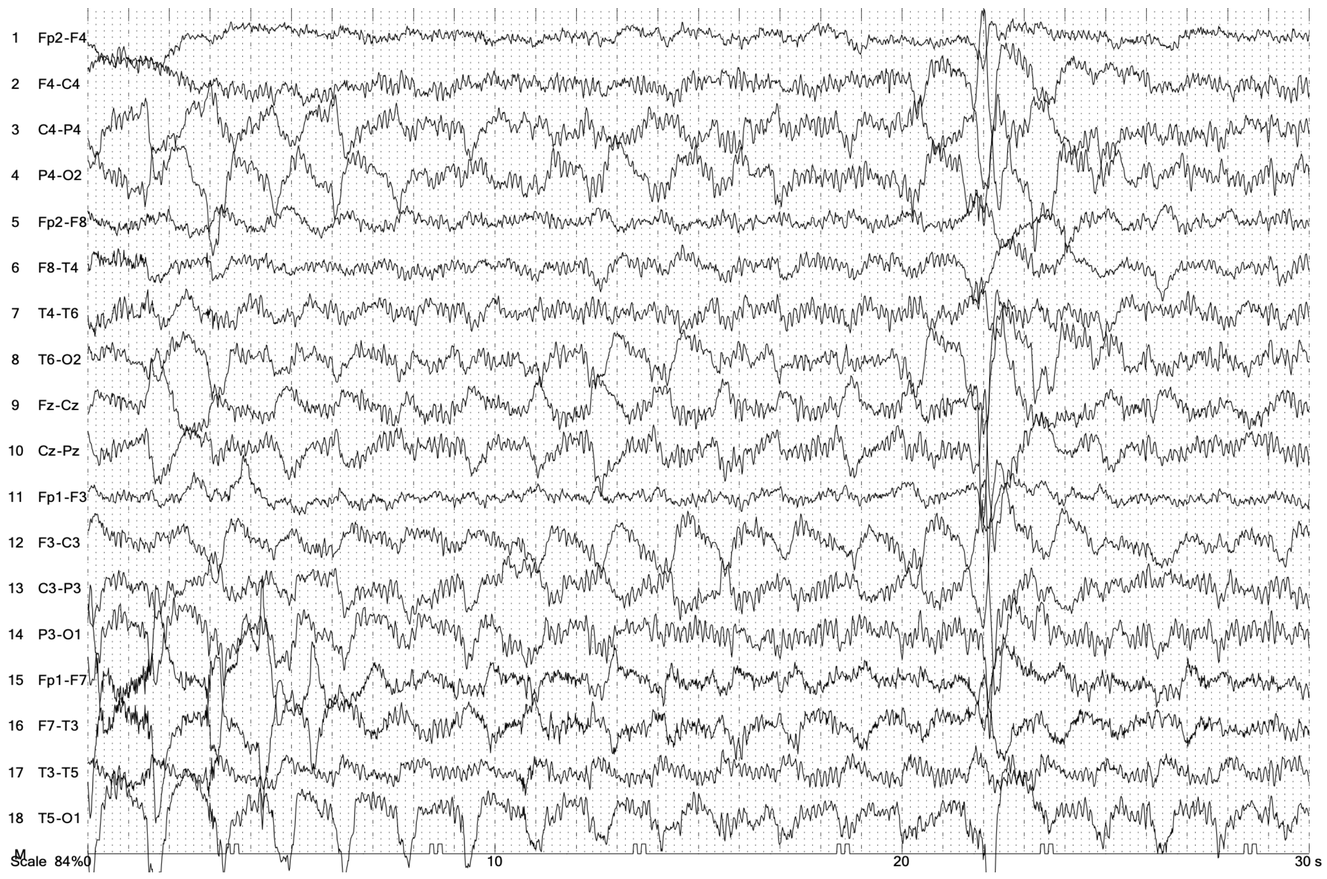
Autoimmune encephalitis (AIE) associated with anti-contactin-associated protein-like 2 (Caspr2) antibodies can mimic neurodegenerative dementia syndromes, leading to diagnostic challenges and treatment delays [1]. We report a case of anti-Caspr2 encephalitis initially misdiagnosed as frontotemporal dementia (FTD).

METHODS

A 67-year-old male with history of alcohol abuse presented with progressive behavioral changes over 5-6 years, including agitation, hallucinations, delusions, gambling addiction, verbal aggression, and disinhibition. Initial assessment suggested FTD based on behavioral symptoms and cognitive decline. Neurological examination also revealed anterocollis, oro lingual dyskinesias, diaphragmatic spasms, myoclonic movement of the limbs, which are atypical for FTD. Further investigations included cerebrospinal fluid analysis, brain MRI, EEG, and screening for autoimmune encephalitis antibodies.

RESULTS

Cerebrospinal fluid analysis revealed anti-Caspr2 antibodies, leading to diagnosis revision to autoimmune encephalitis. Brain MRI showed leucoaraiosis and atrophy, while EEG demonstrated global slowing. Following intravenous immunoglobulin therapy, significant clinical improvement of behavioral symptoms and myoclonus was observed. This aligns with literature reporting that 84% of AIE patients improve with immunotherapy.



DISCUSSION

This case highlights the importance of considering autoimmune etiologies in patients with apparent dementia syndromes, especially when red flags are present: rapidly progressive cognitive decline, subtle seizures (myoclonus in our patient), and atypical findings for neurodegeneration. Recent studies indicate that 38% of AIE patients aged ≥45 years fulfill dementia criteria without prominent seizures early in the disease course.

CONCLUSION

Anti-Caspr2 encephalitis can present with behavioral and cognitive symptoms mimicking FTD. Early recognition and antibody testing are crucial, as timely immunotherapy can lead to substantial clinical improvement, contrasting with the irreversible progression of neurodegenerative dementias.

References

1.Bastiaansen AEM, et al. Autoimmune Encephalitis Resembling Dementia Syndromes. Neurol Neuroimmunol Neuroinflamm. 2021;8(5):e1039