ANTI-IgLON5 ENCEPHALITIS WITH COEXISTING CHOREA AND SLEEP APNEA: A CASE WITH DISTINCT FDG PET/CT FINDINGS

Elena Benevento[1], G. Palermo[1], F. Baldacci[1], C.Pizzanelli[1], E. Del Prete[1], M. Gastaldi[2], R. Ceravolo[1], D. Frosini[1]

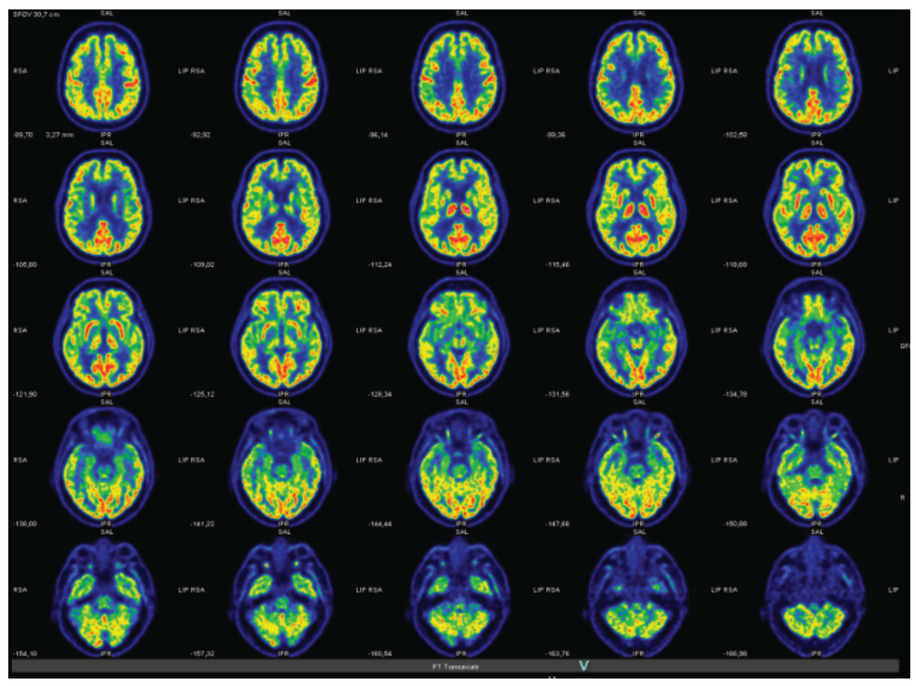
[1]Neurology Unit, Department of Clinical and Experimental Medicine, University of Pisa - Pisa, Italy
[2]Neuroimmunology Research Section, IRCCS Mondino Foundation - Pavia, Italy

Introduction

Anti-IgLON5 disease is a rare neurological disorder combining autoimmune and neurodegenerative features, first described in 2014. It manifests through diverse clinical syndromes, including sleep disturbances, bulbar dysfunction, progressive supranuclear palsy-like symptoms, cognitive decline, and movement disorders. The disease is associated with distinct, though incompletely characterized, brain metabolic and structural changes[1].
We present a 64-year-old man with anti-IgLON5 disease, highlighting an atypical FDG PET/CT metabolic pattern and the rare coexistence of chorea and obstructive sleep apnea (OSA).

Case Description

The patient presented a one-year history of progressive involuntary movements, predominantly affecting the right limbs, along with postural and gait instability, mood changes, dysphagia, weight loss, and speech disturbances. His medical history included smoking, previous alcohol abuse, type 2 diabetes mellitus, hypertension, and a prior polysomnographic diagnosis of OSAS. Neurological examination revealed dysarthria, choreoathetosis with right-sided predominance, dystonic neck posture, blepharospasm, and oral dyskinesias. Diagnostic workup, including brain MRI, CT, nerve conduction studies, rheumatological screening, genetic testing for Huntington's disease, and cerebrospinal fluid analysis, showed no abnormalities. However, FDG PET/CT demonstrated mild thalamic hypermetabolism with bilateral prefrontal cortex and right temporo-lateral hypometabolism. The detection of serum anti-IgLON5 antibodies directed the diagnosis toward IgLON5 encephalitis.



High-dose steroid therapy provided transient improvement, but plasmapheresis was poorly tolerated due to significant dysautonomia. Rituximab was initiated in response to disease progression. Despite treatment, symptoms worsened, influenced by comorbidities and poor therapy tolerance.

Conclusions

This case highlights the clinical heterogeneity of anti-IgLON5 disease and underscores the diagnostic value of FDG PET/CT in identifying unique metabolic patterns. The coexistence of chorea and OSA broadens the spectrum of motor and sleep disturbances linked to the disease[2].
Further research is needed to elucidate the pathophysiology and optimize therapeutic strategies.

Presenting author contact
Elena Benevento
University of Pisa Via Roma 67, Pisa, Italy
mail: elenabenevento@libero.it

Disclosure
The authors have no potential conflict of interest to disclose.

References

[1] Zhang YH, Ni Y, Gao YN, Shen DD, He L, Yin D, Meng HY, Zhou QM, Hu J, Chen S. Anti-IgLON5 disease: a novel topic beyond neuroimmunology. Neural Regen Res. 2023 May;18(5):1017-1022. doi: 10.4103/1673-5374.355742. PMID: 36254983; PMCID: PMC9827781.

[2] Qin S, Wang Y. Anti-IgLON5 disease with severe central sleep apnea-hypopnea syndrome: A case report. Heliyon. 2024 Aug 29;10(17): e36451. doi: 10.1016/j.heliyon.2024.e36451. PMID: 39296154; PMCID: PMC11408130.