

Type 1 Narcolepsy in anti-Hu antibodies mediated encephalitis: a case report M. Vitiello1, E. Antelmi1,2, F. Pizza1,2, E. Postiglione1, R. Poggi3, R. Liguori1,2, G. Plazzi1,2

1. Department of Biomedical and Neuromotor Sciences, Alma Mater Studiorum, University of Bologna, Bologna, Italy 2. IRCSS, Institute of Neurological Sciences, Bologna, Italy 3. Department of Medical Oncology, S. Maria della Scaletta Hospital, Imola, Italy

Introduction: Type 1 Narcolepsy (NT1) is a central hypersomnia due to an immune hypotalamic hypocretin neurons destruction. Rarely, NT1 has been reported in the context of paraneoplastic encephalitis.

Objective: The case at issue reports for the first time a patient with secondary narcolepsy associated with anti-Hu antibodies.

Patients: An 85-year-old man presented with an abrupt onset of transient psychosis that resoved spontaneously after two days. Soon after appeared a rapidly worsening generalized muscular weakness, brief and frequent episodes of facial grimaces, ptosis and slurred speech, with preserved consciousness, triggered by emotions, excessive daytime sleepiness and disrupted nocturnal sleep by awakenings, associated to vivid dream activity. Neurological examination revealed subcontinuous fluctuations in muscle tone with ptosis, facial grimaces and muscle sagging of upper limbs.

Methods and results: Neurophysiological tests (24-hour video-polysomnography and MSLT) and presence of cataplexy led to a diagnosis of Narcolepsy type 1 (NT1). (Fig. 1) Search for HLA DQB1*0602 antigen was negative and CSF hcrt-1 assay disclosed an intermediate level of 146,83 pg/mL.

Due to the atypical age of onset, to the peculiar clinical features, to the "HLA negativity" and to intermediate CSF hcrt-1 levels the patient was further investigated. Serological and biochemical analyses disclosed positivity for anti-neuronal nuclear antibody, type 1, ANNA 1 (anti Hu). Total body CT and fluorodeoxyglucose positron emission tomography scans showed a nodular hilarperhilar lung formation, with features characteristic of malignancy. *(Fig.2)*





Fig 2. (A) Chest TC shows a nodular formation, densitometrically inhomogeneous, in the hilar-perhilar region of right lung with hyperfixation in the same region on a whole-body fluorodeoxyglucose positron emission (B).

Conclusions: diagnosis of secondary NT1 related to anti-Hu antibodies and neoplasm of lung was made.

The list of anti-Hu antibodies associated paraneoplastic neurological syndromes, already encompassing opsoclonusencephalitis, brainstem encephalopathy, myoclonus, paraneoplastic cerebellar degeneration, myelopathy and peripheral neuropathy, should be enriched also by narcolepsy with cataplexy.

Fz-C C3-P T3-T5 E2-M1 E1-M1 Chin

Fig. 1 Cataplectic attach while under synchronized videopolygraphic recording: EEG disclosed a wake activity and the EMG tracings disclosed intermittent loss of tone over the EMG *leads, confirming the suspicion of cataplexy.*

References:

- Thomas J. Dye, Sejal V. Jain, Sanjeev V. Kothare. Central Hypersomnia. Semin Pediatr Neurol 2015.
- Joseph C. Landolfi, Mangala Nadkarni. Paraneoplastic limbic encephalitis and possible narcolepsy in a patient with testicular cancer: Case study. Neuro-Oncology 2003.
- Yaroslau Compta, Alex Iranzo, Joan Santamaría, et al. REM Sleep Behavior Disorder and Narcoleptic Features in Anti–Ma2-associated Encephalitis. Sleep 2007.
- Yves Dauvilliers, Jan Bauer, Valérie Rigau, et al. Hypothalamic Immunopathology in Anti-Ma–Associate Diencephalitis With Narcolepsy-Cataplexy. JAMA Neurol 2013









