Corticobasal syndrome

Corticobasal syndrome (CBS) is a neurodegenerative disease diagnosed based on clinical manifestations such as asymmetrical parkinsonism, limb apraxia, and speech and language impairment. The background pathology of CBS is commonly a variety of proteinopathies, but association with cerebrovascular disease has also been reported. Foix-Chavany-Marie syndrome (FCMS) is a rare neurological disorder characterized by facio-pharyngo-glossal diplegia with automatic-voluntary movement dissociation presenting with bilateral paresis of the facial, lingual, pharyngeal and masticatory muscles. FCMS is commonly attributable to stroke. Transactive response DNA binding protein of 43 kD (TDP-43) proteinopathy is also known as the pathological background of FCMS, while the pathological background of the majority of CBS cases consists of diverse tauopathies instead of TDP-43 proteinopathy. In this report, we describe a case mimicking FCMS that was finally diagnosed as CBS with suggested 4-repeat tauopathy.

A 68-year-old female started experiencing difficulty speaking followed by difficulty writing, and especially texting, several years before her visit. Her impairment had been gradually worsening, and she came to our hospital. On neurological examination, she demonstrated the facial apraxia, frontal lobe dysfunction, and upper motor neuron signs. She presented some characteristics suggestive of FCMS. Her symptoms exhibited rapid progression and myoclonus, parkinsonism, and left-side dominant cortical sensory deficit occurred, resulting in the fulfillment of diagnostic criteria for CBS after 9 months. Tau PET imaging displayed notable ligand uptake in the brainstem, subthalamic nuclei, basal ganglia, and bilateral subcortical frontal lobe, suggesting that her pathological background was 4-repeat tauopathy. As a result of her progressive dysphagia, she became unable to eat and passed away after 12 months.

They hereby present an atypical case of CBS showing clinical features mimicking FCMS at first presentation. TDP-43 proteinopathy was suspected based on the clinical symptoms in the early stages of the disease; however, the clinical course and imaging findings including tau PET suggested that her pathological background was 4-repeat tauopathy [1].

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Nakamura K, Kuroha Y, Hatakeyama M, Kimura AM, Nakamura Y, Murakami Y, Watanabe M, Igarashi H, Takahashi T, Shimada H. Corticobasal syndrome mimicking Foix-Chavany-Marie syndrome with suggested 4-repeat tauopathy by tau PET. BMC Geriatr. 2023 Dec 12;23(1):838. doi: 10.1186/s12877-023-04564-z. PMID: 38087192.