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Corticobasal syndrome in a 23-year-old female

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Abstract

Background: The Corticobasal Syndrome (CBS) is characterized by unilateral parkinsonism and higher cortical dysfunction including apraxia, cortical sensory loss, agnosia, and alien limb phenomena. The underlying neurodegenerative causes of CBS include tauopathies (Corticobasal degeneration [CBD], progressive supranuclear palsy), Alzheimer disease, Lewy body disease, and prion disease, with CBD being most common (CBS-CBD). Disease onset is typically in the fifth to sixth decade of life. Here we present a case of a woman with sporadic CBS due to underlying tauopathy with first signs and symptoms of disease noted at the age 23 years

Method: The patient is a participant of the University of California, San Francisco (UCSF) Memory and Aging Center (MAC) observational, longitudinal research program. The patient underwent structured neurological history and examination, neuropsychological testing, brain MRI, and neurodegenerative disease biomarker studies

Result: The patient is 28-year-old, right-handed, bilingual (Spanish and English) woman with 12 years education. She presented with a 5 year history of slowly progressive left upper and lower extremity sensorimotor deficits, involuntary movements of the left upper extremity, and imbalance. There was no family history of progressive neurological or psychiatric illness. Her neurological examination was notable for left upper and lower extremity rigidity, myoclonic jerks, and dystonic posturing of the left hand. She demonstrated ideomotor apraxia, agraphesthesia, astereognosis, and finger agnosia of the left hand. Neuropsychological examination revealed mild deficits in frontal-executive functions. Serological and cerebrospinal fluid analyses were unrevealing. Brain MRI with and without gadolinium enhancement showed severe right-sided perirolandic atrophy. Tau PET imaging showed asymmetric moderate tracer uptake over the dorsal fronto-parietal cortices. Genetic testing was negative for MAPT, C9orf72, GRN, TARDBP, FUS, PSEN1, PSEN2, and APP mutations/expansions.

Conclusion: CBS is rare before the age of 50 years. We present a case of a 28-year-old woman who meets research criteria for CBS, with brain MRI and tau PET imaging findings suggestive of underlying tauopathy, most likely CBD (CBS – CBD). To our knowledge, this is the youngest case of CBS – CBD ever reported.

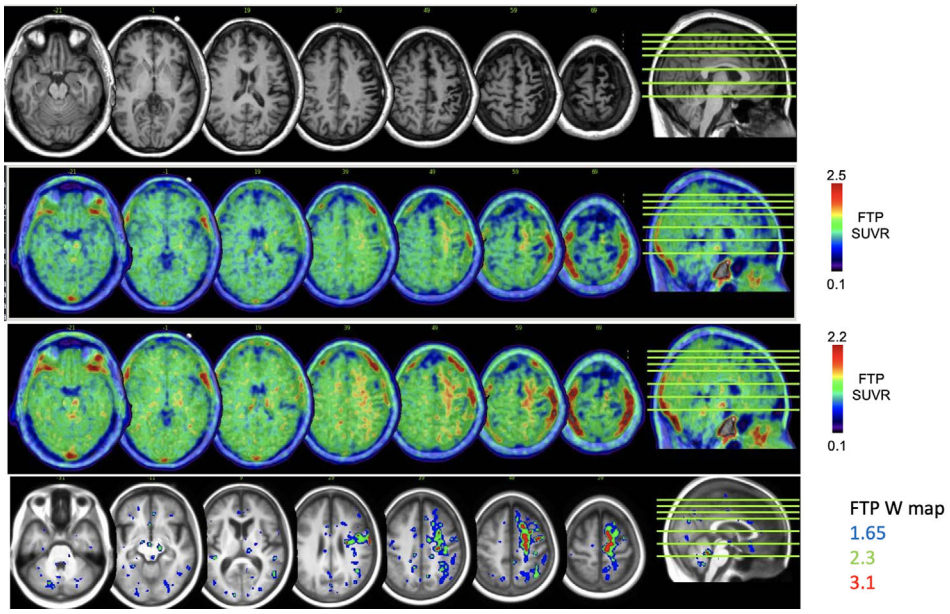

FIGURE 1