of strategies designed to decrease the burden associated with the care of patients with AD.

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"Alien face" in corticobasal degeneration syndrome: extending clinical features

Corticobasal degeneration syndrome (CBDS) is a neurodegenerative disorder characterized by higher cortical dysfunctions associated with progressive asymmetric akinetic-rigid syndrome, and limb dystonia or focal myoclonus (Lang *et al.*, 1994). One of the most typical and specific clinical features in CBDS is considered alien limb phenomenon, usually present in advanced disease stage (Doody and Jankovic, 1992).

Here we describe the case of a patient with CBDS complaining of a newly reported symptom, which, on account of its intriguing features, we termed "alien face" phenomenon. The patient gave her written consent to publication.

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A right-handed, 71-year-old woman with 13 years of education was admitted to the Movement Disorder Center, Neurology Unit, University of Brescia, Italy for clumsiness, stiffness, and inability to use utensils with her left hand. Her past medical and family history for neurodegenerative conditions were unremarkable.

She has progressively developed asymmetric extrapyramidal syndrome since the age of 69, characterized by moderate bradykinesia, increased tone and bilateral cogwheel sign, mainly affecting the left side, as well as a reduced arm swing on the left (Unified Parkinson Disease Rating scale, UPDRS-III = 35/108). No substantial improvement after levo-dopa challenge was observed. Clinically defined left-limb apraxia was detected (De Renzi test = 48/72, cut-off = 62/72) as well as agraphestesia in the right face and extinction to double sensory stimulus. Standardized neuropsychological assessment showed global cognitive functions within the normal range (Mini-mental State Examination = 28/30), with visuo-spatial deficits (Rey Complex Figure, copy = 26/36, cut-off = 27/36). The patient's mother-tongue was French and she was bilingual in Italian with high proficiency. She reported subtle language deficits exclusively in her use of Italian, characterized by anomia and phonemic paraphasia. The immediate and delete verbal and non-verbal memory were intact. No behavioral disturbances apart from apathy were reported by the proxy carer.

Routine laboratory examinations were within normal range. EEG recordings demonstrated bilateral slowing over the frontal and temporal derivations without irrigative features. Brain MRI showed the presence of asymmetric cerebral cortical-subcortical atrophy (right>left) without vascular lesions (see Figure S1, available as supplementary material attached to the electronic version of this letter at www.journals.cambridge.org/jid_IPG), and Single Photon Emission Tomography DATSCAN was positive for severe asymmetric (right>left) degeneration of pre-synaptic nigro-striatal dopaminergic pathway. The diagnosis of clinically defined CBDS was made according to current criteria (Lang *et al.*, 1994).

Additionally, over the past year the patient had complained of an abnormality in the perception of her own face. She explained: "I feel as if the right part of my face is no longer my own, as if it doesn't belong to me as if it was dead" and "I sometimes cover my head with a bandage, to strap the right side of my face to my body, because it seems to be separate, and in this way I find comfort in feeling my face again This is an unpleasant feeling, but it is not painful."

This symptom has progressively worsened in frequency and intensity over time, occurring daily in the last three months. No headache or other associated symptoms were reported. We termed the above phenomenon "alien face" in CBDS, paralleling the other core feature of this syndrome, i.e. alien limb phenomenon.

The pathogenesis of alien limb is still a matter for debate. The common feature of alien limb syndrome is the involuntary autonomous activity of the affected extremity, which is perceived as being controlled by an external force. Different mechanisms have been put forward to explain alien limb, such as

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(1) damage to the medial frontal or corpus callosal regions with disinhibition of inappropriate motor routines, or (2) lesions of the parietal lobe with disturbances of the perception of self-initiated movement (Spence, 2002). Recently, alien leg phenomena have also been described in two patients with CBDS. In both cases, the structural MRI scan showed focal atrophy in the corresponding leg area of the homunculus, suggesting a substrate for the alien features (Hu *et al.*, 2005). An atypical form of CBDS has also been described, as characterized by progressive spasticity (Hasselblatt *et al.*, 2007). This case report represents a typical onset of CBDS, but it extends the clinical features of alien limb to alien face. The sensory representation described in this patient appears to be the basis of an alien face type of phenomenon. In fact, even if the involuntary movement disorders were not observed in the face muscles, it is evident that the sensory perception of the left face is abnormal. These changes are also underlined by the presence of agraphestesia in the same area.

As the cerebral substrate of alien limb is not yet known, the correlation of focal atrophy and alien face is only a matter of speculation. Further studies are needed to investigate the clinical aspects of CBDS, clearly defining the brain's structural underpinnings of the disease.

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