LETTER TO THE EDITOR

To The Editor

Alien Leg Syndrome in Two Cases of Corticobasal Syndrome

Keywords: Alien hand, Alien limb, Alien leg, Corticobasal degeneration, Corticobasal syndrome

Corticobasal degeneration (CBD) is a rare neurodegenerative tauopathy with asymmetrical cortical and basal ganglia involvement. Apraxia, parietal lobe sensory dysfunction, cortical myoclonus, and alien limb phenomenon are common manifestations of cortical dysfunction in corticobasal syndrome (CBS) which is one of the five clinical phenotypes of CBD.[1] As the first presenting feature of CBS, alien limb phenomenon is rare but it eventually occurs in about 50–60% of patients and when presents early, it may cause diagnostic challenge.[2]

The alien limb syndrome (ALS) refers to denial of one's limb possession with unwilled, purposeless, and complex motor activities. Patients are aware of these movements but describe them as estranged and alienated sensation in the affected part of the body with loss of its identity.[3] This syndrome is commonly unilateral in the nondominant hand. Three main subtypes of ALS based on the involved anatomical regions are frontal, callosal, and posterior variants.[4]

Neurodegenerative disorders are the most common etiologies of ALS. In addition to CBS, progressive supranuclear palsy (PSP), posterior cortical atrophy, primary progressive aphasia, and dementia with Lewy bodies have been reported as the causes of ALS. Less common causes are stroke, prion disease, tumors, demeyelination and iatrogenic.[1]

There are limited case reports of alien leg syndrome caused by CBS or other neurodegenerative disorders. Almost all of them have been reported with concomitant upper limb involvement.[3] Herein, we describe two patients with clinically diagnosed CBS, manifesting alien leg phenomenon without concomitant alien hand syndrome (AHS).

Case 1. A 65-year-old man visited our clinic with a 3-year history of levodopa unresponsive slowness of movements, abnormal posture, and jerky movements in the left hand. Neurologic examination revealed slow saccadic eye movements in all directions, hypophonia, and asymmetric limb hypokinesia with rigidity (more severe on the right side). He was suffering from dystonic posture in the left hand with superimposed myoclonic jerks. He also had ideomotor apraxia with agraphesthesia on the right upper extremity (movie 1).

Considering this constellation, we made the diagnosis of probable CBS.[5] In follow-up visit, 6 months later, the patient had developed spontaneous, involuntary, and uncontrollable movements in his right leg, which had been present since 3 months earlier. Examination revealed stereotyped repetitive slow involuntary flexion extension movements of the right knee and elevation of the right leg. These movements resembled the apraxia when performing heel to shin testing (but in our patient the movements were spontaneous) (movie 1). Magnetic resonance (MR) scan of brain revealed atrophy of the left temporal and parietal lobes (Figure 1A).

Case 2. A 74-year-old right-handed man presented with progressive slowing of movements and gait disturbance, which had started 3 years earlier, along with abnormal posture of the left arm. Rapid progression of these symptoms made him wheelchair bound. Neurologic examination revealed asymmetric hypokinesia and rigidity more severe on the left side. He had stimulus sensitive myoclonus in the right fingers and severe dystonic posture of the left hand, which had made this limb useless.

There were involuntary movements in the patient's right leg, which were out of his control. These movements comprised repetitive flexion-extension of the right foot and levitation of the right leg from the hip joint. The patient had difficulty on pushing forward the foot and putting it on the floor voluntarily (movie 2). MR scan of brain showed severe diffuse atrophy with focal atrophy in the parietal lobes more on the left side (Figure 1B).

Herein, we reported two CBS cases with alien leg phenomenon without AHS. Alien limb phenomenon is a common feature in CBS, occurring in 50–60% of patients, and usually involves the upper limb. Practically, many clinicians consider the alien limb phenomenon equivalent to AHS, which brings to mind CBS. Considering its prevalence, the alien limb phenomenon is one of the proposed clinical phenotypes in the diagnostic criteria of CBS alongside apraxia and cortical sensory deficit, asymmetric limb rigidity, hypokinesia, dystonia, and myoclonus as the major clinical features.[2] In contrast to AHS, the alien leg phenomenon without hand involvement is rare and less recognized in CBS and may be misdiagnosed by physicians as lower limb joint problems, painful leg moving toes, or other disorders involving primarily the lower limbs. A possible explanation for the infrequency of alien leg syndrome as compared to AHS is the smaller representation site of the lower limbs compared to the upper limbs on the cortical motor and sensory homunculus.[2] Some may argue that these abnormal leg movements may be a mixture of motor disorders such as dystonia, apraxia, and even clonic perseveration or stereotypy, but the description of these movements from the patients' perspective as being involuntary and uncontrollable as well as having a fixed pattern and not being distractible or changeable in response to a new cue convinced us to consider them as ALS.[6]

Up to now, only six patients are reported with alien leg syndrome. The underling diagnosis of these cases have been stroke, sporadic Creutzfeldt–Jakob disease, or CBS.[1,2,7,8] Our cases had the alien leg phenomenon without alien hand, which has been reported in few CBS cases.[2,7] One of our patients had also no evidence of limb apraxia. As the differential diagnosis for the first case, PSP-CBS (a variant of PSP) can be considered due to slow saccades, but slow saccadic movements are present in many parkinsonian syndromes and the existence of marked asymmetric motor symptoms as well as apraxia, myoclonic jerks, agraphesthesia, and MR scan findings are more suggestive for CBS.

Another interesting notion regarding case 1 was the scattered signs with dystonia and myoclonus over left side, apraxia and alien leg phenomenon over right side, and cortical atrophy over left side. It is well known that in cases of CBS, the signs are concentrated on one side of the body (especially during evolution of disease) and that side is usually contralateral to the side of

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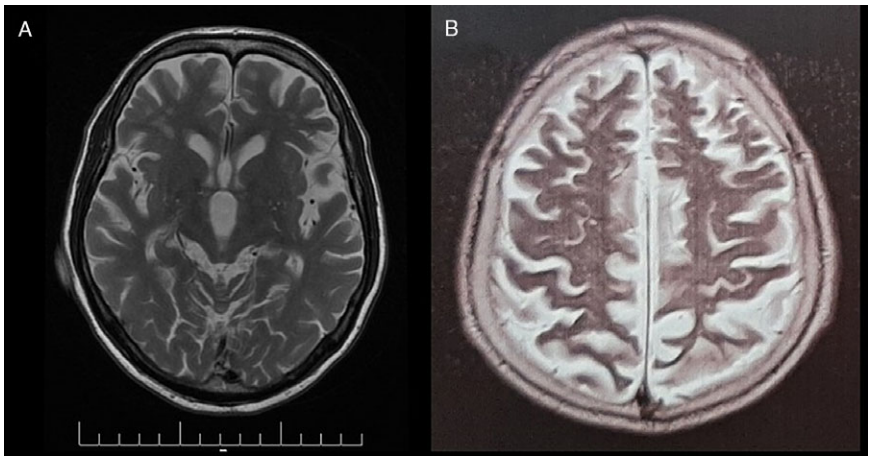

Figure 1: *(A) Axial T2-weighted MRI of the case 1 showing left temporal atrophy. (B) Axial T2 sequence of second case revealing severe atrophy of left parietal lobe.*

atrophy on magnetic resonance imaging (MRI). These findings can be explained by the fact that although the cortical atrophy in left side was more prominent, there was bilateral, especially biparietal atrophy which may explain this dissociation of signs over both sides. Additionally, the "posterior" variants of alien limb tend to occur more often with right hemispheric (parietal) lesions, yet in both our cases, the right legs were involved. This finding does not seem to be related to handedness, as both patients were right-handed. Biparietal atrophy might be an explanation for this, but the exact pathophysiologic mechanism still remains obscure to us.

To conclude, alien leg syndrome, although rare, should be considered as an important sign for diagnosis of CBS and clinicians must be aware of this easily negligible sign to avoid missing this diagnosis.

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Conflict of Interest

The authors have no conflicts of interest to declare.

Statement of Authorship

The conception of this report was set off by MR and organized by ME. GH, YS, and SA contributed in executing the project and writing the primary draft. FH and AS reviewed and reorganized the cases and the manuscript.

Supplementary Material

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