Case Report

Alien hand syndrome: Contradictive movement and disorder of color discrimination

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A senile Chinese female patient with alien hand syndrome of vascular etiology is reported. This case exhibited contradictive movement, left-lateral paresis and disorder of color discrimination, which might be a new subtype of the alien limb syndrome.

Key Words: Alien hand syndrome, Involuntary movement, Cerebrovascular diseases, Disorder of color discrimination.

Introduction

Alien limb syndrome or alien limb phenomenon is summarized as: the patient complains that an arm or leg has a 'life of its own' and is almost always asymmetrical; on examination, the limb may move and even grasp objects involuntarily; best assessed by distracting the patient with some other task and observing the limb; often seen in corticobasal degeneration (CBD).1,2 It is related with a cluster of symptoms characterized by the involuntary movement of a single upper limb in conjunction with the experience of estrangement from or personification of the movements of the limb itself. However, a reliable, anatomically derived definition of the alien hand syndrome has been elusive. A reason for this broadened acceptable application has been the identification of theoretically dissociable subtypes.3 We present a case of a specific variety or subtype of the alien hand syndrome, and discuss its etiology and possible anatomical reasoning.

Case Report

A 74-year-old right-handed woman suddenly developed left-sided slight weakness, feeling as if the “left hand was controlled by a ghost” and sighted a gray wall as having five-six colors on its surface. Nine weeks after this attack, she became dysphagic and dysarthric. Left hemi-inattention was also noted. Initial brain CT without contrast showed probable infarct in the right temporal-parietal-occipital region involving the cortex and adjacent white matter. Follow-up MRI in a week post-onset showed chronic ischemic lesion in the right temporal-parietal-occipital gray and white matter, but callosal involvement was not seen. She complained often that her left arm “doesn’t do what its supposed to do. It always is controlled by a ghost or god.” The nature of the involuntary movement in the left arm was primarily uncontrolled levitation with intermittent writhing of her fingers. No exploratory or self-stimulating behavior (grasping skin, clothing) was ever noted to occur, and she often restrained her left arm with her right hand. She could not perform bimanual tasks. The patient had no formal education, had no positive personal or family history or any mental disorder. There was no history of alcohol or drug abuse.

Neurological examination revealed that her tongue was turned towards the left. There were no tremors. The tendon reflexes in all the four limbs were exaggerated. The patient showed right-left disorientation and did not exhibit grasp reflex or apraxic symptoms in either upper limb. The revised Hasegawa’s dementia scale showed her score as 9, indicating that her intelligence was below normal. The routine serum biochemical tests were normal. Her spontaneous verbal production was nearly normal in rate and frequency of utterances, but she had mild dysarthria. She exhibited pronounced left visual neglect on line bisection and target cancellation tasks. Proprioception and kinaesthesia were both severely impaired bilaterally; worse in the left than the right upper limb. Tactile sensory and proprioceptive impairment prevented meaningful testing of the patient’s ability to differentiate between her affected hand and the examiner’s hand when held out of sight. Stereognosis and tactile object naming were not possible with the left hand, and were impaired with the right.

After 12 weeks of rehabilitation and the preventive management of cerebral vascular risks and a small dose of tiapride, the levitation and complaints of avolitional movement had reduced considerably in frequency, though visual neglect, slight lower limb weakness, and hemianesthesia remained. Throughout, the patient seemed perplexed rather than angered or depressed by her contradictive movement.

Discussion

The alien hand syndrome was originally used to describe cases involving the anterior part of corpus callosal lesions (split-brain person) producing involuntary or contradictive move-
ments and a concomitant inability to distinguish the affected hand from an examiner’s hand when these were placed in the patient’s unaffected hand. Some varieties or subtypes of the alien hand syndrome have been reported, involving lesions of the corpus callosum alone, the corpus callosum and the dominant medial frontal cortex, and the temporal or/and parietal cortical/sub-cortical areas, and rarely, non-dominant hemisphere thalamic infarcts. The pathogenic chart of the alien hand syndrome has often been found to contain CBD mainly. It has also been reported in the acute cerebral vascular diseases, herpes viral encephalitis (Avrahami-Heller), Alzheimer’s disease, progressive supranuclear palsy, epilepsy and Creutzfeldt–Jakob’s disease, which have been seen in Europe, North and South America, and Asia.

Given this patient’s involuntary movements and her verbal expressions of perplexity and estrangement from the movements, she clearly manifested the alien hand syndrome as defined in recent studies. She did not, however, exhibit the mutism, apathy, exploratory behavior, groping, compulsive manipulation of objects and tools, or grasp reflex reported in callosal-frontal cases. She likewise exhibited the intermanual conflict seen in callosal cases and exhibited no signs of callosal disconnection. Despite the right temporal-parietal-occipital region abnormalities on MRI, this patient was similar to the reported cases of the alien hand syndrome associated with posterior lesions. Such cases have involved multiple loci of cerebral dysfunction (e.g., cortical and subcortical) caused by single or multiple infarcts, which perhaps independently, produced the subjective and behavioral symptoms of the alien hand syndrome. Two previously reported cases of the alien hand syndrome involved non-dominant hemisphere thalamic infarcts in conjunction with additional cortical, sensory impairment. Cases of posterior alien hand syndrome arising from single or multiple infarcts, which perhaps independently, produced the subjective and behavioral symptoms of the alien hand syndrome, have been reported, involving lesions of the corpus callosum alone, the corpus callosum and the dominant medial frontal cortex, and the temporal or/and parietal cortical/sub-cortical areas, and rarely, non-dominant hemisphere thalamic infarcts.

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At present, there seem to be some broad clusters of behavioral and subjective symptoms subsumed under the diagnosis “alien hand syndrome”. This patient and other examples suggest that the alien hand syndrome may not only be as closely associated with focal, critical lesion sites as its callosal and callosal-frontal counterparts, but also with the temporal-parietal-occipital region as well as with the single temporal or single parietal region in the dominant cerebral hemisphere. Rather, it seems to be a disorder of involuntary movement in the context of alterations in body schema sufficient to cause feelings of estrangement from those movements. These symptoms may occur independently of one another and may occur from either single or multiple lesions. The alien hand syndrome, compared to “frozen feet”, a kind of contradictory phenomenon seen often in Parkinsonism, may also be a symptom of movement disorder so attention should be drawn to basal ganglia dysfunction.

References


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