Subjective experience, involuntary movement, and posterior alien hand syndrome

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Abstract
The alien hand syndrome, as originally defined, was used to describe cases involving anterior corpus callosal lesions producing involuntary movement and a concomitant inability to distinguish the affected hand from an examiner’s hand when these were placed in the patient’s unaffected hand. In recent years, acceptable usage of the term has broadened considerably, and has been defined as involuntary movement occurring in the context of feelings of estrangement from or personification of the affected limb or its movements. Three varieties of alien hand syndrome have been reported, involving lesions of the corpus callosum alone, the corpus callosum plus dominant medial frontal cortex, and posterior cortical/subcortical areas. A patient with posterior alien hand syndrome of vascular aetiology is reported and the findings are discussed in the light of a conceptualisation of posterior alien hand syndrome as a disorder which may be less associated with specific focal neuropathology than are its callosal and callosal-frontal counterparts.

Keywords: alien hand syndrome; involuntary movement

Alien hand syndrome is a loosely defined cluster of symptoms characterised by involuntary movement of an upper limb in conjunction with the experience of estrangement from or personification of the movements of the limb itself. Alien hand syndrome was initially used to describe cases involving a disconnection of the hemispheres via corpus callosal lesion.1,2 Although such early reports suggested considerable neuropathological specificity (a lesion of the anterior corpus callosum), a reliable anatomically derived definition of alien hand syndrome has been elusive.3 More recent acceptable usage of the term alien hand syndrome requires only the “feeling that one limb is foreign or “has a will of its own,” together with (emphasis in original) observable involuntary motor activity.”4 A reason for this broadened acceptable usage has been the identification of theoretically dissociable subtypes. Two frontal varieties of alien hand syndrome have been described—one associated with lesions of the language dominant medial frontal cortex and the anterior corpus callosum affecting the dominant hand and the other with of the corpus callosum alone affecting the non-dominant hand.1,5,6 The behavioural differences between the two, although not absolute, are that the callosal-frontal alien hand engages in more exploratory grasping behaviour and compulsive manipulation, whereas the callosal alien hand presents primarily with intermanual conflict.1 Some disagreement exists as to whether the callosal-frontal variety is the product of disinhibited posterior dominant hemispheric environmental exploratory drive (producing dominant hand involuntary movement)7,8 or of disrupted medial frontal inhibition of lateral frontal exploratory drive which exists in both hemispheres.9 In the second case, the infrequency of non-dominant hand callosal-frontal alien hand syndrome would be attributable to hypokinesia, which often follows non-dominant medial frontal lesions, and which would tend to negate any released exploratory drive.9 A few cases of alien hand syndrome have been reported after posterior lesions5,10 resulting either from corticobasal degeneration involving primarily posterior cortical degeneration or from cerebrovascular accidents. Generally, though not exclusively, these cases have involved the non-dominant limb. The sustained involuntary movements are typically non-purposeful and non-conflictual, and include such behaviours as arm levitation and finger writhing.11,12

We present a case of the second, posterior variety of alien hand syndrome and discuss considerations regarding its aetiology and differences from the two anterior varieties.

Results
A 73 year old, right handed woman developed left side weakness and loss of consciousness after having felt “odd all day.” On admission to hospital, she was reportedly severely dysphagic and moderately dysarthric, and exhibited a moderate left hemiparesis. Severe left hemi-inattention was also noted. Initial brain CT without contrast showed probable infarct in the right parietal-temporal-occipital region involving the cortex and adjacent white matter. Follow up MRI at 11 days postonset showed additional low density in the right frontal grey and white matter with sparing of the corpus callosum.
The infarct extended subcortically, sparing the right basal ganglia but involving both the medial and lateral frontal cortices. Extensive vasogenic oedema was also noted in the right posterior temporoparietal region. T1 weighted sagittal and fluid attenuated inversion recovery (FLAIR) axial MRI images from the follow up study are shown in the figure. Carotid ultrasound performed at 7 days postonset showed bilateral plaque formation with bilateral internal carotid artery stenosis of about 50%.

When the patient entered rehabilitation 51 days postonset, her upper limb hemiparesis had largely resolved, although she had only minimal antigravity movement in her lower limbs. She complained often that her left arm “doesn’t do what its supposed to do,” that it “has a mind of its own,” and that it “acts up.” She eloquently added, “I talk about it like it’s an entity of its own.” The nature of the involuntary movement in the arm was primarily uncontrolled levitation with intermittent writhing of her fingers. The patient complained that she had awakened at least once to find her arm elevated at a 90 degree angle from her bed. No exploratory or self stimulating behaviour (grasping skin, clothing) or intermanual conflict was ever noted to occur, and the patient and her therapists denied such occurrences on repeated questioning. She often restrained her left arm with her right hand. Although she could move the left arm and hand to command, her movements were ataxic and she initiated little spontaneous voluntary movement. She could perform bimanual tasks (for example, folding a sheet of paper and placing it into an envelope) without evidence of intermanual conflict, although her use of the left arm in such tasks was minimal, as when using it to hold an envelope firmly against a table top. The patient did not exhibit grasp reflex or apraxic symptoms in either upper limb.

The patient’s spontaneous verbal production was nearly normal in rate and frequency of utterances, but her mild dysarthria persisted. Whereas the earlier reported dense left hemianopsia had largely resolved, responses to stimuli in the left lower quadrant were inconsistent. The patient exhibited pronounced left visual neglect on line bisection and target cancellation tasks, although she did look to the left spontaneously when addressed from that side and would follow commands to look leftward. Left upper and lower extremity tactile sensation was severely impaired, although she made inconsistent allaesthetic responses to firm touch of the left hand. Proprioception and kinaesthesia were both severely impaired bilaterally, worse in the left than right upper limb. The patient often reported that she could not find her left arm, a difficulty compounded by the involuntary movements. 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 impossible with the left hand, and were impaired with the right. Severe visuospatial and constructional impairment was noted (for example, the patient could not approximate the
first item on the WAIS-R block design subtest). During the block design subtest, the patient attempted to use her left hand to help her dominant right hand, but it inadvertently knocked several of the blocks from the table surface due to ataxic movement. This occurrence was not interpreted by the patient or the examiner as representing intermanual conflict. After this incident, she did not attempt again to use the left hand, and it slowly elevated itself, bent at a 90°, to shoulder level.

By her discharge 75 days postonset, the levitation and complaints of avolitional movements had reduced considerably in frequency, although visual neglect, lower limb weakness, upper limb ataxia, and hemianesthesia remained. Throughout, the patient seemed perplexed rather than angered or frightened by her involuntary movement.

Discussion
Given this patient’s involuntary movements and her verbal expressions of perplexity and estrangement from the movements, she clearly manifested alien hand syndrome as defined in recent studies. She did not, however, exhibit the exploratory behaviour, groping, compulsive manipulation of objects and tools, or grasp reflex reported in callosal-frontal cases. She likewise never exhibited the intermanual conflict seen in callosal cases and exhibited no signs of callosal disconnection. Finally, callosal involvement was not suggested by MRI.

Rather, despite the frontal and medial frontal abnormalities on MRI, her case is similar to reported cases of alien hand syndrome associated with posterior lesions. Such cases have involved multiple loci of cerebral dysfunction (for instance, cortical and subcortical) caused by single or multiple infarcts which, perhaps independently, produced the subjective and behavioural symptoms of alien hand syndrome. Two previously reported cases of alien hand syndrome involved non-dominant hemisphere thalamic infarcts in conjunction with additional cortical, sensory impairment. Ay et al11 suggested that their patient’s involuntary movement was triply determined. They suggested that a thalamic infarct produced both cerebellar ataxia and sensory ataxia, while a posterior cortical infarct contributed to optic ataxia. They hypothesised that the body schema distortions and hemineglect produced alien feelings regarding the involved limb, and that together, these phenomena produced alien hand syndrome. Cases of posterior alien hand syndrome arising from corticobasal degeneration may be similarly multideterminate.

We speculate that our patient’s feelings of estrangement from her non-dominant upper limb and its movements were similarly produced by body schema distortion and hemineglect secondary to the non-dominant parietal lobe infarct. The precise aetiology of her involuntary movement, however, is not as clear. It is possible that disrupted cortico-striato-thalamic functioning (in this case resulting from more widespread right hemispheric infarcts) produced involuntary movement due to sensory ataxia and poor proprioceptive awareness.

Another possibility is of unvisualised basal ganglia dysfunction. Other authors4 have found abnormal basal ganglia cerebral blood flow in a patient with hemidystonia when CT, EEG, and angiography were normal. That the involuntary movements are not specifically associated with any particular, theoretically critical neuropathology represents a fundamental difference between posterior alien hand syndrome and the callosal and callosal-frontal varieties.

Remission of alien hand syndrome of this type may occur with improvement of either the involuntary movements, alterations in the body schema, or both. Our patient’s involuntary movements had largely resolved on her discharge, whereas her alterations in body schema (visual neglect, hemianesthesia, and proprioceptive impairment) remained. It may be reasonable to expect that, given this context, any future occurrence of sustained involuntary movements (for example, as related to further cerebrovascular compromise) could produce a recurrence of alien hand syndrome.

At present, there seem to be three broad clusters of behavioural and subjective symptoms subsumed under the diagnosis “alien hand syndrome.” This patient and other examples suggest that posterior alien hand syndrome may occur with a thalamic infarct. Cranial computed tomographic lesion sites as its callosal and callosal-frontal counterparts (callosal and callosal plus medial frontal lesions). 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 arise independently from one another and may arise from either single or multiple lesions.

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References