

SHORT REPORT

Posterior alien hand syndrome after a right thalamic infarct

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The alien hand syndrome, as originally defined, should be reserved for cases in which the hand feels foreign "together with" observable involuntary motor activity. These involuntary movements are unusual during or after acute stroke. 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 and subcortical areas. A patient with posterior alien hand syndrome of vascular aetiology is reported. Imaging studies disclosed an isolated infarction of the right thalamus sparing other cerebral regions.

Although several studies have reported on patients with alien hand syndrome since Goldstein's initial description,¹ most of them were attributable to corpus callosum and mesial frontal lesions, either alone or in combination.^{2–4} The alien hand syndrome has also been reported after lesions sparing the corpus callosum.^{5–9} In the classic syndrome, the main symptom is the report of alienness with abnormal movements of the hand. In comparison with the frontal/callosal alien hand cases in which motor plans are abnormally activated and callosal pathology prevents normal transfer to the opposite hemisphere, our patient had a complicated sensory-ataxic disorder with self stimulation by the left arm.

CASE REPORT

A 64 year old right handed woman with a medical history of coronary artery disease and longstanding type I diabetes mellitus awoke to find her left arm was acting as if under someone else's control. She reported that she was astonished and afraid of it because her left arm seemed to act independently and she could not control "his" movements.

At admission she was fully alert, oriented, and cooperative. On neurological examination cranial nerve function was normal. Speech was fluent and language abilities were functionally intact for conversation with no evidence of dysarthria. No homonymous hemianopia was detected. She had normal tone and strength on both sides. There was left hemiasomatognosia and mild left spatial neglect with impaired visuospatial processing. Other neuropsychological dysfunction was absent. Apraxia was not present. The patient presented a prominent hemisensory defect affecting both "superficial" (tactile, temperature, and pain) and "deep" (position and vibration) modalities. She also showed delayed and distorted perception, inability to localise sensation, and astereognosis. She had an hemiataxic disturbance: right arm movements were normal, but those on the left were inaccurate in trajectory and irregular in speed and amplitude. The limb showed features of an approach as compared with an avoidance bias, which is a distinguishing feature separating the posterior from the anterior

frontal form. These intermittent movements had an irregular speed with slow and smooth onset but became more jerky and rapid before hitting her body. The left arm sometimes struck objects on her face. Movements often seemed delayed or "stuck" with episodes of uncontrolled levitation that could not be diminished voluntarily. She made no attempt to correct the movements visually at the onset of the process. Optic ataxia was not present. Plantar responses were flexor bilaterally. She presented bilateral achilles areflexia.

These spontaneous movements were observed frequently and episodically: she described how she saw a curtain moving alone finding to her own surprise that was her left arm handling it. A second episode took place while she was undressing. The left limb grasped and pulled up her dressing gown in a perseverative way. She had to sit down and wait until this involuntary behaviour stopped. She many times denied ownership and attributed an own personality to the limb. Her left hand, with eyes closed, was confused with examiner's hand or a third party's hand although recognised like a hand when performing manoeuvres recommended to explore alienness.¹⁰ Diagnostic dyspraxia, mirror movements, and intermanual conflict were not observed.

The patient received antiaggregant treatment with ticlopidine.

Initial brain computed tomography performed at admission showed a right thalamic infarct. Follow up magnetic resonance imaging three days after onset revealed T1 (TR 700 ms, TE 20 ms) and T2 (TR 2700 ms, TE 95 ms) weighted images, an infarct in the ventral posterolateral and dorsal portion of the right thalamus sparing the posterior limb of the internal capsule and the mesencephalon. No lesions were detected in the corpus callosum or other cerebral regions (fig 1).

On the fifth hospital day her misreaching with either hand and sensory loss improved. Left arm dysmetria was less prominent. By the end of the first hospital week, she no longer feared her left hand and she soon came to treat it as a misbehaving child. By her discharge, 16 days after onset, the involuntary movements had reduced considerably in frequency but the sense of alienness persisted unchanged. She improved slightly during the next two months. The ataxia was still severe but she now used visual guidance to direct her hand helping her to suppress the involuntary movements. She died one year after discharge. Throughout, the involuntary movements persisted but reduced considerably in frequency and the patient still interpreted these movements as alien in origin.

DISCUSSION

Alien hand syndrome is defined as unwilling, uncontrollable movements of an upper limb together with the verbally expressed feeling of foreignness from these movements and frequent personification of the arm.^{10–11} This involuntary motor activity is not part of an identifiable movement disorder. According to this definition, our patient fulfils the criteria for

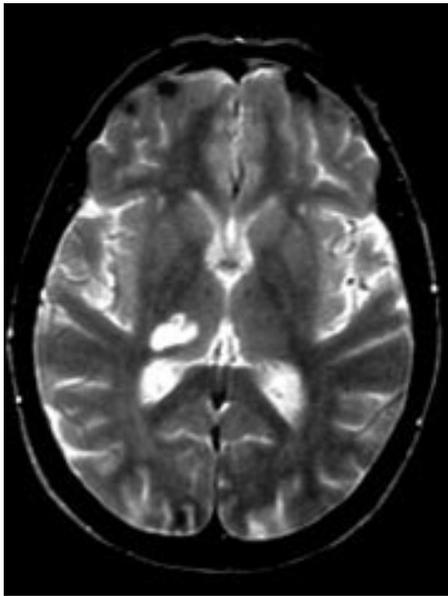


Figure 1 Axial magnetic resonance image (TR 2700 ms, TE 95 ms) shows high signal indicating a subacute infarction in the ventral posterolateral and dorsal portion of the right thalamus.

alien hand syndrome. To the best of our knowledge, a patient with a sustained alien hand syndrome attributable to an isolated right thalamic infarction has not been reported previously.

This particular sign may be a component of various constellations of syndromes; at least three: the callosal and the frontal forms in which the alien hand syndrome is secondary to a corpus callosum lesion alone or in combination with a lesion in the frontal area, sometimes associated with lesions in the basal ganglia or thalamus,^{4 6 8} and the posterior or parieto-occipital form resulting either from corticobasal degeneration involving primarily posterior cortical degeneration or from cerebrovascular event.^{6 8 12-15}

The callosal alien hand form involves the non-dominant hand and is characterised primarily by intermanual conflict with typically little evidence of limb paresis, whereas the callosal-frontal variety implies a more grasping behaviour and compulsive manipulation with a crural paresis of the limbs contralateral to the affected hemisphere. The posterior form generally involves the non-dominant hand, which tends to levitate into the air displaying ataxic, non-purposeful and non-conflictual movements with relevant sensory impairment, evidence of visual and somatosensory dysfunction as well as body schema distortion.

Fisher provides¹⁶ an extensive review of the literature of the motor abnormalities that have come to be included under the designation of alien hand phenomena with the addition of six personal cases, emphasising on the clinical description of the abnormal movements.

We attribute the alien hand syndrome in our patient to the right thalamic lesion. As compared with the frontal/callosal form, our case is similar to reported cases of alien hand syndrome associated with posterior lesions.

The patient had a complicated “sensory-ataxic” disorder with self stimulation from the neglected side. Similar clinical syndromes were reported by Levine and Rinn⁶ in a right handed woman with a homonymous hemianopia, hemianesthesia, and opticosensory ataxia attributable to proximal right posterior cerebral artery infarction (right basal temporo-occipital and small ventral posterolateral thalamic infarcts) and by Ay *et al*¹³ after subacute infarction in the right thalamus, hippocampus, inferior temporal lobes, splenium of

corpus callosum, and occipital lobe attributable to right posterior cerebral artery occlusion. These authors suggested that this syndrome should be considered as a “sensory” or “posterior” form of the alien hand syndrome to be distinguished from the “motor” or “anterior” form described more commonly.

Ventura *et al*⁸ described a patient with an alien hand syndrome attributable to a right capsulothalamic haemorrhage with mesencephalic extension without a detected lesion in the corpus callosum, showing in a PET scan right frontoparietotemporal cortical hypometabolism, most pronounced in the sensorimotor area. The report of Bundick and Spinella¹² included a patient with a posterior variant of the alien hand syndrome and an infarct in the right parietotemporooccipital region sparing the right basal ganglia and the corpus callosum but involving both the medial and lateral frontal cortices. They suggested that posterior alien hand syndrome may not be closely associated with focal, critical lesions sites as its “anterior” counterparts and that it seems to be an involuntary movement in the context of changes in body schema sufficient to cause feelings of estrangement from those movements. Martí-Fàbregas *et al*⁹ reported a patient with an isolated non-dominant parietal infarction without corpus callosum involvement and a contralateral alien hand syndrome. They speculate that the right parietal infarction in their patient could cause a loss of sensory integration and feedback interfering with motor function.

How can we explain the right thalamic lesion account for both the feeling of alienness and the involuntary movements presented by our patient? Lesions at any level of the sensory pathways may cause ataxia of the contralateral limbs. Inability to distinguish her own left arm from someone else’s with her eyes closed could be attributed to the sensory loss. The feeling of lack of control over spontaneous left arm movements may have been related to the “sensory ataxia”. This “sensory ataxia” has been associated at necropsy with lesions of the ventral posterolateral thalamus and also the splenium of the corpus callosum and the temporo-occipital lobes.^{6 13} Dejerine believed this to be a sensory ataxia caused by destruction of the somatosensory nuclei of the thalamus.¹⁷

On MRI lesion mapping, the infarct involved the ventral posterior nucleus and the ventral lateral nucleus (thalamo-geniculate or inferolateral territory) and also the dorsomedian nucleus (tuberothalamic artery). It is important to note that in cases of thalamic infarction, the extent of the lesion is not restricted to individual nuclei or groups of nuclei but is attributable to the complex and variable supply from the internal carotid, posterior communicating, and posterior cerebral arteries.¹⁸

Sensory loss in our patient may be easily explained by involvement of the ventral posterolateral and ventral posteromedial nuclei on the right thalamus. The abnormal movements were unintentional and she lacked the sensory feedback to determine their authorship once their exaggerated character brought them to notice. The personification of the arm, sometimes hostile, may have been an elaboration of the sense of lack of control or foreign control of the limb. The fact that she appeared to be using visual guidance to control the left hand movements, rather than relying on somatosensory kinesthetic feedback from the limb, seemed to “help” her to suppress the involuntary motor behaviour, which was much more prominent early on after the onset of her stroke. Perhaps this could be achieved by a reorganisation of sensory input used to stabilise the motor cortex output that had become unstable because of its “de-afferentation” resulting from the lesion to somatosensory thalamic nuclei.

The explanation for thalamic ataxia may be more controversial. Involvement of the ventral lateral nucleus, with interruption of dentatorubrothalamic fibres, may be a possible cause for ataxia in our patient who showed involvement of that nuclear group on MRI. In patients with ataxic hemiparesis

and internal capsule involvement, the ataxia has been linked to the interruption of reciprocal fibres connecting the ventral lateral nucleus and the precentral cortex.¹⁹ This emphasises the possibility that the ventral lateral nucleus of the thalamus may play a key part in the genesis of "thalamic ataxia".

Crossed cerebellar diaschisis¹⁹ describes a metabolic depression in the cerebellar hemisphere contralateral to supratentorial infarction. Although crossed cerebellar diaschisis has been reported in patients with thalamic stroke the correlation with clinical ataxia has only been anecdotal. However, the remote effect of thalamic injury in prefrontal cortex has been demonstrated by several functional neuroimaging studies.^{8–13} Lesions in a given thalamic nucleus may produce cognitive deficits similar to those seen after injury to the primary area of cortical connectivity. Associations have been established between injury to specific thalamic nuclei and the development of hemispatial neglect. Much less is known about the relation between the thalamus and complex executive behaviours associated with frontal lobe function. Nevertheless, impaired visuospatial performance and hemineglect are not rare in thalamic infarctions, especially in the territory of the non-dominant tuberothalamic artery.^{13–18–19} Partial involvement of the ventral and dorsomedial nucleus may explain these manifestations.¹⁸ Ay *et al* performed a PET study in a patient with left hemineglect and an alien hand syndrome showing altered metabolism in the right frontotemporoparietal regions, suggesting a remote effect of a thalamic lesion ("diaschisis").¹³

With regard to the complex movement disorder; complicated combinations of more than one movement disorder may be observed after thalamic lesions¹³ and is easy to suggest an extrapyramidal origin for the episodic, paroxysmal, and involuntary movements seen in our patient.²⁰ One important element in the posterior alien hand syndrome is self stimulation by the left arm. Left arm ataxia was severe in both our patient and the patients of Levine and Rinn⁶ and Ay *et al*.¹³ We believe that there was mainly sensory ataxia, the movements often seemed delayed or stuck and this does not characterise the type of ataxia typically seen in cerebellar ataxia. In the same way, the ability to effect a partial amelioration of the behaviour over time through the incorporation of visual guidance would suggest that this was not a straightforward cerebellar form of ataxia.

We speculate that the isolated right thalamic infarct produced the combination of both the sensory ataxia and a hemineglect syndrome and suggest that these phenomena together with the involuntary movement disorder produced the alien hand syndrome. The involuntary movements in the context of alterations in body schema seem to be sufficient to cause feelings of estrangement from these movements.

A posterior alien hand syndrome may arise from a single thalamic lesion. We agree with other authors^{6–13–21} that this syndrome should be considered a "sensory" or "posterior" form of the alien hand syndrome, as distinguished from the more commonly described "anterior" or "motor" form.

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