

# The Alien Hand Sign—Case Report and Review of the Literature

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## Abstract

*The alien hand sign refers to a group of signs which include a feeling that the hand is foreign together with autonomous activity, as if the hand is driven by an external agent. These features are commonly associated with frontal and callosal lesions. We report the alien hand sign in a man with left medial cortical infarct, presenting with autonomous grasping and groping with his right upper limb, denial of ownership of his limb, and dissociative phenomena including self restriction and intermanual conflict.*

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*Key words: Autonomous activity, Callosal lesion, Frontal lesion, Grasp reaction, Grasp reflex, Supplementary motor area*

## Introduction

The alien hand sign or “strange hand sign”, first coined by Brion and Jedynak in 1972,<sup>1</sup> refers to a feeling that the hand is foreign, together with autonomous activity which is perceived as independent of voluntary control, as if the hand is driven by an external agent.<sup>2</sup>

The description of the entity dates back to 1908 where Goldstein<sup>3,4</sup> reported a woman whose left hand had “a will of its own”, which at times even grabbed and choked her throat, and in which it took great strength to pull it off. To date, at least 19 cases have been reported in the literature.<sup>1-8</sup> Lesions were found most commonly in the medial frontal cortex following contralateral frontal strokes and the corpus callosum following corpus callosum infarction, corpus callosotomy, or in haemorrhagic or traumatic lesions affecting both the corpus callosum and medial frontal area. In addition, combination of a posterior corpus callosum lesion and contralateral thalamic sensory lesion,<sup>9</sup> bifrontal penetrating cerebral injury<sup>10</sup> and corticobasal degeneration have been associated.”

We report a case of right-sided alien hand sign in a right-handed man with left medial cortical infarct.

## Case Report

A 59-year-old right-handed man with a background history of hypertension, hypercholesterolaemia and gout was admitted to the acute ward with sudden onset of right hemiparesis. CT scan revealed infarct of the left

frontal lobe corresponding to the distribution of the left anterior cerebral artery. There was another area of subacute ischaemia seen in the left posterior parietal lobe in the periventricular white matter. Numerous other lacunar infarcts were seen in the left internal capsule, left putamen and external capsule. Ultrasound duplex of the carotids was normal.

Examination on the 26th post-stroke day revealed a quiet, reticent gentleman, who seldom initiated conversation. His answers to questions were mainly 2- to 3-word phrases or short sentences. Repetition, naming and comprehension were intact.

Examinations of the cardiovascular and respiratory systems were essentially normal. Motor power on the left side was normal, while that on the right upper limb was 4/5 on the Medical Research Council scale and 3/5 in the right lower limb. Tone was mildly increased with an upper motor neuron pattern on the right side. The grasp reflex was absent. Sensations to touch and pinprick were intact.

During the course of the examination, the patient demonstrated involuntary grasping movements of the right upper limb, reaching out to objects on the right side for example, the rail of the bed and the edge of the table. On grasping the object, he then found it difficult to let go, and often used his left hand to ungrasp his right hand. The patient was aware of and frustrated at this behaviour, and often used his left arm to hit his right.

In addition, his right arm would demonstrate other

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involuntary movements, for example, reaching out to grasp the back of his neck, or scratch his cheek. The patient would sometimes leave these movements alone, or use his left hand to hold on to his right in an attempt to check this movement.

The patient volunteered that his right hand felt "funny". When asked to whom the right hand belonged, he replied "devil's hand". Similarly, for his leg he replied "devil's leg". For the left hand and leg, he replied "my left hand" or "my left leg", respectively. He would also volunteer that his right hand felt as if it did "not belong" to him, as he "cannot control it".

The patient was very reluctant to attempt to write. With the right hand, he declared "cannot control", and with his left, he said, "cannot write". However, later during the course of his stay, he managed an attempt to write his name with his left hand.

The patient was able to cross localise stimulation of fingertips fairly accurately, and cross replicate postures with both hands. Tactile and visual naming functions were intact.

When asked to pour a cup of water from a jug using both hands, there was an initial tendency to use his left hand. When he was told to use his right hand, he was able to grasp the jug. He then had difficulty letting go of the jug with his right hand. On drinking, he used his left hand to hold the cup to his mouth, while still grasping the cup with his right hand. When he finished drinking, he used his left hand to push the cup away from his right hand, which continued to grasp the cup.

During his stay in hospital, the patient gradually showed increasing ability to release the involuntary grasp especially during less agitated moments, and regain more control of his right hand. He was able to execute commands with this hand, especially when concentrating on the movement. On discharge, he was able to feed, dress and bathe himself with minimal assistance, and walk with supervision.

## Discussion

Characteristics reported to be associated with the alien hand sign range from a failure to recognise ownership of one's limb when visual cues are removed,<sup>1</sup> a feeling that one's body part is foreign,<sup>2</sup> to personification and autonomous activity.<sup>2,4</sup> Additional characteristics associated include involuntary complex motor activities including motor perseveration, subtle speech and language abnormalities and a wide range of complex motor behaviour ranging from groping and grasping<sup>12</sup> to self destructive activity or intermanual conflict.<sup>13</sup>

Our patient had several features consistent with those of the alien hand sign. Firstly, he denied ownership of his hand on several occasions, saying it did not belong to him. Although Brion and Jedynak<sup>1</sup> originally described

"alienation" of the limb as the inability of patients to differentiate their own hand from the examiner's by feeling with their normal hand, this has not served as the definitive feature of alien movement. In addition, it has limited application with bilateral lesions and when somatosensory loss in the affected hand influences the outcome.<sup>5</sup>

Doody and Jankovic,<sup>2</sup> in their review of cases, proposed that the alien limb be defined as a feeling that one limb is foreign or has a "will of its own", together with observable involuntary motor activity. The description in most published reports supports this definition.<sup>5</sup> The feeling of alienation can even be to the extent of personification of the limb as another personality altogether.<sup>2</sup>

Neglect syndromes secondary to parietal disease or optic ataxia secondary to occipital pathology may induce a sense of foreignness or the feeling that a limb does not "follow commands". Neglect syndromes, however, seem to be related to impaired interpretation of body perception,<sup>2</sup> while dissociative phenomenon or a feeling of "foreignness" appear to be a misinterpretation of body action.<sup>13</sup> There is a dissociation between intention and action wherein patients deny ownership of the limb because of its tendency to involuntary movements which is not controllable intentionally. There were involuntary movements in our patient which comprised automatic grasping and groping movements.

Autonomous movements associated with the alien hand sign noted in the literature include action induced patterned and rhythmical movement, non-goal directed grasping and groping behaviour, or goal directed activities like utilisation behaviour and self destructive acts.<sup>2,5</sup> Grasping (or groping) actions and intermanual conflict are the most commonly reported.<sup>5</sup>

Instinctive grasp reactions refer to a range of behaviours including a tendency to take hold of object stimulus.<sup>14</sup> These reactions can be evoked by visual stimuli, without previous tactile stimulation, especially when they are located in the same half of the visual space as the responding hand and are close to it.<sup>15</sup> Once grasping has occurred, the patient is then unable to release the object by opening the hand. These deficits have been associated with lesions restricted to the supplementary motor area on the medial frontal region.<sup>16</sup>

The grasp reflex refers to a stereotyped simple response, typically the closure of the patient's hand on the examiner's fingers. Attempts to withdraw the fingers through the thumb-index cleft result in an increase of the strength of the grasp.<sup>17</sup> It is widely accepted to be an index of frontal lobe pathology.<sup>17</sup>

"Self restriction", referring to attempts by the patient to control the alien limb and dissociative phenomena with some degree of intermanual conflict was seen in our patient. Self restriction is described in most reported

cases of the alien hand sign.<sup>5</sup> Intermanual conflict, where one hand acts at cross-purposes to the other, has most frequently been described after commissurotomy.<sup>18</sup> Two hands are observed to engage in actions whose purposes are opposed.

There was a paucity of spontaneous speech production with preserved ability to repeat in our patient, a pattern often designated as transcortical motor aphasia.<sup>12</sup> This is not uncommonly described in patients who have sustained damage to the left medial cortex.<sup>19</sup> Evidence from regional cerebral blood flow studies suggests that the supplementary motor area participates in the programming of human speech and is especially important for organising vocalisation that is self initiated and generative (spontaneous propositional speech) rather than exogenously triggered and nonpropositional (externally instructed repetition).<sup>20</sup>

Symptoms associated with the alien hand sign have been reported to vary according to the location of the lesion. Feinberg proposed a distinction between “frontal” and “callosal” forms of the alien hand sign. Frontal alien hand sign, resulting from damage to the supplementary motor area (SMA), anterior cingulate gyrus and medial prefrontal cortex of the dominant hemisphere and anterior corpus callosum, is associated with an apparently purposeful and uncontrollable tendency of the hand contralateral to the lesion to grope and grasp objects.<sup>21</sup> The callosal alien hand syndrome, on the other hand, causes hemispheric disconnection which is manifested during behaviours requiring dominant-hemisphere control. It is typically characterised by intermanual conflict, and manifestation requires only an anterior callosal lesion. In addition, they have been reported to show disconnection syndrome with unilateral tactile anomia, unilateral agraphia, unilateral apraxia, difficulty copying drawings, and abnormalities of somaesthetic transfer.<sup>22</sup>

Potential deficits arising from anterior cerebral artery (ACA) infarction are protean and depend on the exact site of the occlusion. The supplementary motor area is frequently damaged by cerebral infarction in the territory of the ipsilateral anterior cerebral artery or its branches. At the same time, diverse clinical deficits of speech, language, motor function and praxis seen following ACA infarction have frequently been ascribed to SMA damage despite the fact that there are a variety of cortical regions and subcortical structures that are often involved in ACA territory infarctions in addition to the SMA. The majority of patients following ACA infarction have lesions of the anterior corpus callosum and cingulate gyrus in addition to SMA damage. In a series of 27 consecutive patients with ACA territory infarction assessed by CT scans, some involvement of the corpus callosum was present in 20.<sup>23</sup>

Bilateral lesions have also been described. In one report, bilateral frontal lobe damage from primary cerebral lymphoma produced bilateral utilisation and imitation behaviour in a right-handed man. He also had bilateral grasp reflexes and left motor neglect. He would reach for and grasp whatever attracted his attention, whether visually or tactually.<sup>24</sup> Other reports of bilateral utilisation and imitation behaviour have usually involved extensive bifrontal lesions, including orbital cortices.<sup>25,26</sup>

The alien hand sign has also been reported in association with corticobasal degeneration and Alzheimer’s histopathology. Patients with corticobasal degeneration have been reported to have the alien hand syndrome in association with ideomotor apraxia, an akinetic rigid syndrome and other involuntary movements such as focal dystonia and myoclonus. Dysfunction in the supplementary motor area is believed to be the cause of the ideomotor apraxia and the alien limb behaviour in these patients.<sup>27</sup>

Various mechanisms for the alien hand sign have been proposed. In the normal brain, volitional movements have been demonstrated to emanate from the medial premotor systems, consisting of the supplementary motor area and cingulate gyrus, and reactive movements from lateral (arcuate premotor area) and premotor systems.<sup>13,22</sup> In one hypothesis, both medial and lateral premotor systems are normally balanced through mutual inhibition and are highly interactive. After unilateral medial frontal cortical dysfunction, alien movements, which are reactive in nature, are postulated to be produced from the disinhibited lateral premotor system of the contralateral hemisphere. Anticipatory control of the affected hand is controlled by the intact supplementary motor area of the hemisphere ipsilateral to this hand.<sup>5,10</sup> A callosal lesion may be postulated to produce alien movements by dissociating the affected supplementary motor area from contralateral supplementary motor area input. The affected hand becomes “free-running”, producing reactive movements in response to environmental changes or spontaneous motor activity.<sup>5</sup> This frequently results in intermanual conflict.

An alternate conceptualisation of the alien hand sign can be based upon a single medial premotor system in the supplementary motor area that normally controls anticipatory and reactive movements differently. In the alien hand sign, damage to the supplementary motor area may selectively impair initiating operation more than the monitoring operation, thereby impairing reactive and anticipatory movements differently.<sup>5</sup>

Alien hand movements have typically reported to be transient, disappearing within the first year after stroke, but persisting when the lesion involved bilateral cortical and subcortical structures.<sup>5,10</sup> It is possible that the alien

hand behaviour is reduced through a compensatory process in which the intact hemisphere ipsilateral to the affected hand extends control over segmental mechanisms through ipsilateral projections from premotor regions that may operate in parallel with the primary motor cortex. Recent blood flow studies have demonstrated significant bihemispheric cerebral reorganization in patients who have recovered movement after stroke-related hemiparesis.<sup>28</sup> This may explain why with bihemispheric damage, the alien hand sign is less likely to be ameliorated with time.<sup>13,29</sup>

### Conclusion

The alien hand sign with denial of one's limb and dissociative phenomena, could be interpreted as faulty initiation, execution and inhibition of pre-existing motor subroutines which are coupled normally to "external and internal inputs", of which the exact pathophysiology remains to be delineated. Physicians managing patients with neurological disease, especially with lesions in the anterior cerebral artery territory and corpus callosal region, should be cognisant of the existence of this interesting phenomena.

### REFERENCES

Brion S, Jedynak C I. Troubles du transfert interhemispherique. *Revue Neurol (Paris)* 1972; 126:257-66.

Doody R S, Jankovic J. The alien hand and related signs. *J Neurol Neurosurg Psychiatry* 1992; 55:806-10.

Goldstein K. Zur Lehre der motorischen Apraxie. *J fur Psychologie und neurologie* 1908; 11:169-87.

Goldstein K. Dermakroskopische Hirnbefund in meinem Falle von linksseitiger motorischer Apraxie. *Neurol Centralbl* 1909; 28:898-906.

Gasoquoine P G. Alien hand sign. *J Clin Exp Neuropsychol* 1993; 15:653-67.

Leiguarda R, Starkstein S, Nogues M, Berthier M, Roberto A. Paroxysmal alien hand syndrome. *J Neurol Neurosurg Psychiatry* 1993; 56:788-92.

Ball J A, Lantos P L, Jackson M, Marsden C D, Scadding J S, Rossor M N. Alien hand sign in association with Alzheimer's histopathology. *J Neurol Neurosurg Psychiatry* 1993; 56:1020-3.

Bogen J E. The callosal syndrome. In: Heilman KM, Valenstein E,

editors. *Clinical Neuropsychology*. New York: New York University Press, 1979:308-59.

9. Levine D N, Rinn W E. Opticosensory ataxia and alien hand syndrome after posterior cerebral artery territory infarction. *Neurology* 1986; 36:1094-7.

10. Banks G, Short I, Martinez AJ, Latchaw R, Ratcliff G, Boller F. The alien hand syndrome. *Arch Neurol* 1989; 46:456-9.

11. Gibb W R G, Luther P J, Marsden C D. Corticobasal degeneration. *Brain* 1989; 112:1171-92.

12. Goldberg G, Mayer N H, Togliola J U. Medial frontal cortex infarction and the alien hand sign. *Arch Neurol* 1981; 38:683-6.

13. Goldberg G, Bloom K K. The alien hand sign: Localization, lateralization and recovery. *Am J Phys Med Rehabil* 1990; 69:228-38.

14. Seyffarth H, Denny-Brown D. The grasp reflex and the instinctive grasp reaction. *Brain* 1948; 71:109-83.

15. Magnani G, Mazzucchi A, Poletti A, Scoditti U, Parma M. Involuntary grasping and groping responses to space-related visual stimuli. *Mov Disord* 1987; 2:9-23.

16. Penfield W, Welch K. The supplementary motor area of the cerebral cortex. *Arch Neurol Psychiatry* 1951; 66:289-317.

17. Renzi E D, Barhieri C. The incidence of the grasp reflex following hemispheric lesion and its relation to frontal damage. *Brain* 1992; 115:293-313.

18. Bogen J E. The callosal syndrome. In: Heilman KM, Valenstein E, editors. *Clinical Neuropsychology*. London: Oxford Univ Press, 1979:308-59.

19. Masdeu J C, Schoene W C, Funkenstein H. Aphasia following infarction of the left supplementary motor area. *Neurology* 1978; 28:1220-3.

20. Jonas S. The supplementary motor region and speech emission. *J Commun Disord* 1981; 14:349-73.

21. Feinberg T E, Schindler R, Flanagan N G, Haber L D. Two alien hand syndromes. *Neurology* 1992; 42:19-24.

22. Leiguarda R, Starkstein S, Berthier M. Anterior callosal haemorrhage: a partial interhemispheric disconnection syndrome. *Brain* 1989; 112:1019-37.

23. Bougouslavsky J, Regli F. Anterior cerebral artery territory infarction in the Lausanne Stroke Registry: clinical and etiologic patterns. *Arch Neurol* 1990; 47:144-50.

24. Fukui T, Hasegawa Y, Sugita K, Tsukagoshi H. Utilization behaviour and concomitant motor neglect by bilateral frontal lobe damage. *Eur Neurol* 1993; 33:325-30.

25. Lhermitte F. "Utilization behaviour" and its relation to lesions of the frontal lobes. *Brain* 1983; 106:237-55.

26. Lhermitte F, Pillon B, Serdaru M. Human anatomy and the frontal lobes. Part I. Imitation and utilization behaviour: a neuropsychological study of 75 patients. *Ann Neurol* 1986; 19:326-34.

27. Gibb W R G, Luther P J, Marsden C D. Corticobasal degeneration. *Brain* 1989; 112:1171-92.

28. Brion J-P, Demeurisse G, Gapon A. Evidence of cortical reorganization in hemiparetic patients. *Stroke* 1989; 20:1079-84.

29. Banks G, Short I, Martinez A J, Latchaw R, Ratcliff G, Boller F. Alien hand syndrome: clinical and postmortem findings. *Arch Neurol* 1989; 46:456-9.