

Case report

Dominant posterior-variant alien hand syndrome after acute left parietal infarction

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ABSTRACT

Alien hand syndrome (AHS) is a complex clinical disorder in which patients develop a sense of estrangement from, and loss of volitional control of, an affected limb and non-purposeful complex motor actions of the same limb. Several forms of AHS exist, including frontal, callosal, and posterior types with a series of different associated neuroanatomical lesions and clinical symptoms. Most commonly, the lesions associated with AHS occur in the frontal lobes and corpus callosum. Rarely, lesions in the parietal lobes may be associated with AHS, and most often occur within the non-dominant hemisphere. We describe a 57-year-old patient who developed symptoms of posterior AHS after an acute infarction in the left (presumably dominant) parietal lobe. A review of the different clinical features of AHS and the underlying mechanisms is also presented.

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1. Introduction

Alien hand syndrome encompasses a variety of clinical symptoms and neurological lesions and involves a sense of disconnection between a patient's will and action. It was first described in the early twentieth century in a 57-year-old woman who developed a "feeling of estrangement" from her left upper limb and associated motor abnormalities that were distinct from both ataxia and paresis [1]. Roughly 60 years later, three patients were reported with similar symptoms, which the authors termed "le signe de la main étrangère" or alien hand sign [2]. Since that time, this terminology has come to describe numerous different clinical and pathologic entities involving areas of the frontal lobes, corpus callosum, and more recently the parietal lobes, occipital lobes, and thalamus [3]. When the parietal lobes are involved, the lesions are almost exclusively found in the non-dominant hemisphere. Here we present a case of a patient who developed AHS as his presenting symptom following an acute left (presumably dominant) parietal infarction.

2. Case report

The patient is a 57-year-old, right-handed man with a history of hyperlipidemia, hypertension, obesity, tobacco use, and chronic obstructive pulmonary disease who noted abnormal movements of his right arm, while driving his car. The patient described feeling that he had lost control of his arm and felt like it had "fallen

off." He also reported that his right arm grabbed the steering wheel "without me knowing it." His symptoms continued intermittently over the next several hours and the patient subsequently presented to the emergency department. The patient denied motor or sensory problems in any other limb, visual or hearing changes, loss of consciousness, or hallucinations. On examination, he had a temperature of 97.8 F, blood pressure of 149/88, heart rate of 68, respiratory rate of 18 and an oxygen saturation of 99% on room air. His general physical examination revealed no significant abnormalities. On neurological examination, he was alert and oriented to person, place and time. He had spontaneous and fluent speech, normal comprehension and was able to follow commands. His cranial nerves were intact. On motor examination, the patient demonstrated normal strength bilaterally. However, the patient's right arm demonstrated spontaneous waving movements while his eyes were closed. He denied knowledge or control of these movements. On sensory examination, he had decreased sensation to pin prick testing on his right hand and arm. He was also noted to have decreased graphesthesia and stereognosis on his right upper extremity compared to his left. His gait was normal.

MRI examination demonstrated a subacute infarction in the left anterior parietal lobe along the posterior lip of the postcentral gyrus. This is expected to involve the posterior aspect of the primary sensory strip (likely Brodmann's area 2), as well as the tertiary somatosensory cortex in the superior parietal lobule (likely in the location of Brodmann's area 5) (Fig. 1A and B). No other major lesions or infarcts were seen, though note was also made of a very few punctuate foci of diffusion hyperintensity in the left motor/premotor region. The remainder of the brain appeared normal. The patient also underwent time-of-flight MR angiography of

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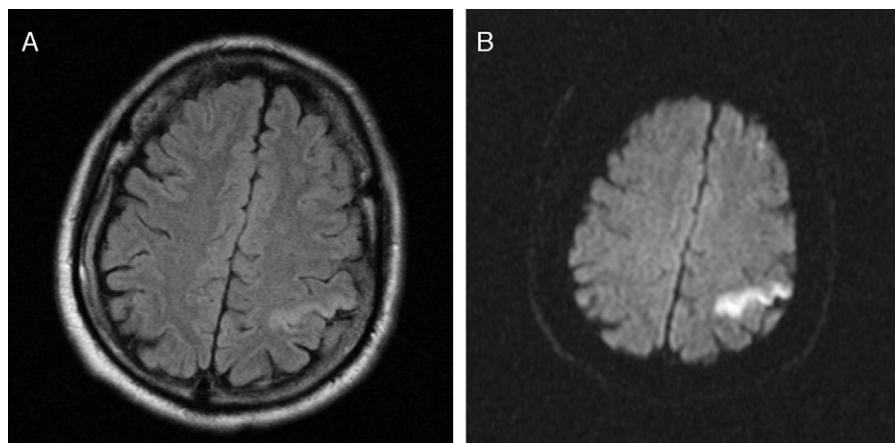


Fig. 1. (A) FLAIR demonstrating hyperintensity in the left anterior parietal lobe along the posterior lip of the postcentral gyrus. (B) Diffusion weighted images demonstrating corresponding hyperintensity in the left anterior parietal lobe.

the circle of Willis and neck vessels that demonstrated no significant intracranial stenoses. Neck MRA showed moderate stenosis of the left internal carotid artery.

His symptoms continued to improve during his hospitalization, but were still present upon discharge 5 days later. The patient was seen again in neurology clinic eight days after his discharge, when he noted moderate improvement in his symptoms with residual discomfort using his right hand.

3. Discussion

Alien hand syndrome (AHS) is one of the most fascinating disconnection syndromes in neurology. It is a complex disorder that may result from a variety of central nervous system lesions that may present with several different signs and symptoms. The common factor between all cases of AHS is the patient's experience of dissociation between his intention and the subsequent motor activity [4,5]. Typical symptoms include a sense of foreignness of a limb, failure to recognize ownership of a limb, and autonomous motor activity perceived as involuntary [6].

Several different mechanisms of neurologic injury may cause AHS including infarction, surgical resection, seizure, and corticobasal degeneration [5,7,8,4]. Early research in AHS divided the disorder into two clinical and pathologic categories [9]. The first type presented in patients with damage to the dominant hemisphere in the supplementary motor area, anterior cingulate gyrus, or medial prefrontal cortex. These patients tended to develop motor signs of AHS that included grasping or groping activities with their right hand. The second type of AHS presented in patients with damage to the anterior corpus callosum, and presented primarily with complex movements of the affected limb that counteracted the movements of the unaffected limb, often referred to as intermanual conflict. For example, in one case a thirsty patient filled a glass of water with one hand, only to pour it out with the other [10].

However, it later became clear that there existed a group of patients who demonstrated symptoms of AHS, but lacked an identifiable lesion in the anterior structures typically thought to produce the syndrome. Often, these patients also presented with more prominent feelings of estrangement from the affected limb and less complex motor activity, such as limb levitation, than in the other types of AHS [3,11–14]. This variant became known as posterior AHS [10,12,15].

Though the specific pathways remain unknown, several authors have proposed theories of the neuronal mechanisms underlying the posterior forms of AHS. The inferior parietal lobule has been described as a multimodal association area that receives input

from multiple neuronal pathways and coordinates motor output [5,15,16]. Thus, disruption to this system may lead to the interruption of normal motor output, such as that seen in AHS. Alternatively, some have hypothesized that posterior AHS may represent a variation of neglect syndromes [12]. In typical neglect syndromes patients suffer damage to the parietal lobe and subsequently ignore the contralateral hemispace. In comparison, posterior AHS patients may suffer damage to a portion of the parietal lobe and subsequently disregard the activities of their own contralateral limb, while maintaining normal interactions with the contralateral external world.

Most reports of the posterior-variant AHS have involved the right parietal lobe and the left hand [11,14,17]. There have been very few reports of isolated lesions in the left (presumably dominant) parietal lobe causing posterior AHS of the right upper extremity [16]. In one case, a patient developed seizures and subsequent right limb AHS after surgical removal of an arteriovenous malformation from the left parietal lobe [5]. Other published reports have described posterior AHS symptoms in patients with non-ischemia related atrophy in the left parietal lobe [11,18].

In our review of the literature, we were able to identify only one more case of a patient with AHS whose imaging demonstrated an infarct in the left parietal lobe [19]. This patient was a 63-year-old woman, who presented with acute right hemiparesis and dysarthria. Several months later, after substantial improvement in her hemiparesis, she began demonstrating symptoms of posterior AHS, including a feeling of estrangement from her limb and involuntary limb levitation.

Our patient differs in two important ways from the other reported cases of dominant parietal lobe AHS. Firstly, to our knowledge, no other cases of left parietal AHS have been reported in the setting of acute infarction. Second, though commonly described in the frontal and callosal forms of AHS, complex motor movements are quite rare in the posterior form. Though some authors have described simple groping movements in patients with posterior-variant AHS, more frequently the motor component of posterior AHS involves non-purposeful movements or limb levitation [3]. In contrast, our patient manifested complex movements, including seemingly purposeful manipulation of the steering wheel of his car.

It is unclear what mechanisms underlie the symptoms of AHS in our patient. Though his anatomic lesions are posterior, our patient's clinical symptoms are more similar to those described in the anterior forms of AHS. This combination of findings raises the possibility that AHS disorders may represent a spectrum ranging from purely frontal to purely posterior forms. More specifically, our patient's case provides interesting correlations because the infarct is rela-

tively discreet along the cortical ribbon of the posterior lip of the postcentral gyrus, rather than a massive lesion which would preclude specific neuroanatomic correlation. In our patient, one of the main structures involved is expected to be the posterior portion of the sensory strip (e.g., Brodmann's area 2), which subtends the kinesthetic sense (as opposed to Brodmann's areas 3a and 3b in the anterior portion of the sensory strip, which subtend sensations of pain and temperature) [20]. Another main structure involved would appear to be Brodmann's area 5, just posterior to Brodmann's area 2, corresponding to a portion of the tertiary somatosensory cortex, which subtends stereognosis [21]. Thus, the lesions in our patient seem to involve a sense of where the limb is and what it is touching, logically correlating to an alien limb syndrome presentation, as opposed to a simple hypesthesia.

Further imaging research is needed into the neural pathways that are disrupted in patients like the one presented here to more clearly elucidate the neuroanatomic correlates. However, our patient illustrates that alien hand symptomatology may occur in the right upper extremity from a left parietal acute infarct, and may present acutely with complex motor movements once thought typical only of anterior-variant alien hand syndromes.

References

- [1] Goldstein K. Zur lebre der motorischen apraxia. *Z Physiol Neurol* 1908;11:169–87.
- [2] Brion CP, Jedynak S. Troubles du transfert interhémisphérique. A propos de trois observations de tumeurs du corps calleux. Le signe de la main étrangère. *Rev Neurol* 1972;126:257–66.
- [3] Assal F, Schwartz S, Vuilleumier P. Moving with or without will: functional neural correlates of alien hand syndrome. *Ann Neurol* 2007;62:301–6.
- [4] Fischer CM. Alien hand phenomena: a review with addition of six personal cases. *Can J Neurol Sci* 2000;27:192–203.
- [5] Leiguarda R, Starkstein S, Noguez M, Berthier M, Arbelaz R. Paroxysmal alien hand syndrome. *J Neurol Neurosurg Psychiatry* 1993;56:788–92.
- [6] Doody RS, Jankovic J. The alien hand and related signs. *J Neurol Neurosurg Psychiatry* 1992;55:806–10.
- [7] Kikkert MA, Ribbers GM, Koudstaal PJ. Alien hand syndrome in stroke: a report of 2 cases and review of the literature. *Arch Phys Med Rehabil* 2006;87:728–32.
- [8] Levine DN, Rinn WE. Opticosenory ataxia and alien hand syndrome after posterior cerebral artery territory infarction. *Neurology* 1986;36:1094–7.
- [9] Feinberg TE, Schindler RJ, Flanagan NG, Haber LD. Two alien hand syndromes. *Neurology* 1992;42:19–24.
- [10] Akelaitis AJ. Studies on the corpus callosum. IV. Diagonist dyspraxia in epileptics following partial and complete section of the corpus callosum. *Am J Psychiatry* 1944;101:594–9.
- [11] Carrilho PE, Caramelli P, Cardoso F, Barbosa ER, Buchpiguel CA, Nitrini R. Involuntary hand levitation associated with parietal damage. *Arq Neuropsiquiatr* 2001;59:521–5.
- [12] Martí-Fàbregas J, Kulisevsky J, Baró E, Mendoza G, Valencia C, Martí-Vilalta JL. Alien hand sign after a right parietal infarction. *Cerebrovasc Dis* 2000;10:70–2.
- [13] Ventura MG, Goldman S, Hildebrand J. Alien hand syndrome without a corpus callosum lesion. *J Neurol Neurosurg Psychiatry* 1995;58:735–7.
- [14] Marey-Lopez J, Rubio-Nazabal E, Alonso-Magdalena L, Lopez-Facal S. Posterior alien hand syndrome after a right thalamic infarct. *J Neurol Neurosurg Psychiatry* 2002;73:447–9.
- [15] Goldberg G, Bloom KK. The alien hand sign. Localization, lateralization and recovery. *Am J Phys Med Rehabil* 1990;69:228–38.
- [16] Jeannerod M. The neuronal and behavioral organization of goal-directed movements, vol. 15. Oxford: Clarendon Press; 1988.
- [17] Scepkowski LA, Cronin-Golomb A. The alien hand: cases, categorization, and anatomical correlates. *Behav Cogn Neurosci Rev* 2003;2:261–77.
- [18] Rohde S, Weidauer S, Lanfermann H, Zanella F. Posterior alien hand syndrome: case report. *Neuroradiology* 2002;44:921–3.
- [19] Gondim FA, Oliveira GR, Cruz-Flores S. Position-dependent levitation of the dominant arm after left parietal stroke: an unreported feature of posterior alien limb syndrome? *Mov Disord* 2005;20:632–3.
- [20] Baehr M, Frotscher M. Duus' topical diagnosis in neurology. Stuttgart: Thieme Verlag; 2005. p. 375.
- [21] Blumenfeld H. Neuroanatomy through clinical cases. Sunderland: Sinauer Publishers; 2002. p. 31.