

Ego-Syntonic Alien Hand Syndrome After Right Posterior Cerebral Artery Stroke

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ABSTRACT. Groom KN, Ng WK, Kevorkian CG, Levy JK. Ego-syntonic alien hand syndrome after right posterior cerebral artery stroke. *Arch Phys Med Rehabil* 1999;80:162-5.

The alien hand syndrome classically consists of involuntary movements accompanied by a feeling of foreignness and personification of the affected limb. Autocriticism, in which patients criticize and express astonished frustration with the behavior of the autonomous limb, is a commonly noted feature. Most cases of alien hand are associated with lesions of the supplementary motor areas of the frontal lobes, the corpus callosum, or both. The authors report on a 79-year-old man who sustained a right posterior cerebral artery distribution infarction and developed alien hand syndrome in the absence of callosal involvement. Also unique is that the patient perceived the alien hand as acceptable and positive, not hostile and repugnant. This case suggests not only that the neuroanatomic regions responsible for alien hand syndrome may require re-examination, but also that its definition may need expansion.

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THE ALIEN HAND syndrome (AHS) was first described in 1908 by Goldstein¹ as a type of apraxia.¹ The knowledge of this clinical entity advanced little for 64 years, until the condition was characterized as *main étrangère*, or *alien hand*, by Brion and Jedynak,² in patients with cerebral neoplasm. In recent decades, this syndrome has been the subject of much interest and debate. The critical components necessary to make the diagnosis of AHS are described with great variability. Recent reports have converged around the following criteria: (1) a feeling of foreignness, or lack of recognition of the affected limb, and (2) involuntary movements of the limb, which give rise to personification of the affected body part.^{3,4} In addition to these classic features, a variety of associated features are often present, such as intermanual conflict/diagnostic dyspraxia (ie, the two hands work at cross purposes), mirror movements of the unaffected hand, grasp reflex, impulsive groping, compulsive manipulation of tools, self-restriction, callosal signs, ideomotor and construction apraxia, and motor perseveration.^{5,6} Autocriticism, in which patients criticize and express astonished frustration with the behavior of the autonomous limb, is also associated with AHS.⁴ Indeed, reports exclusively suggest AHS as *ego-dystonic*, a psychiatric

term that characterizes a set of thoughts, feelings, or behaviors as repugnant or inconsistent with the patient's total personality. This is in contrast to the term *ego-syntonic*, in which thoughts, feelings, or behaviors of the patient are construed as acceptable and consistent with his or her total personality.

Most cases of alien hand are associated with lesions of the supplementary motor area of the frontal lobe, the corpus callosum, or both.⁴ Although each of these regions appears sufficient to produce AHS in the absence of injury to the other, no etiologic theory has emerged that can explain all cases.^{7,8} For example, patients with corticobasal degeneration who have manifested AHS have shown cell loss in the substantia nigra, locus ceruleus, and nucleus ambiguus.⁴ With regard to more posterior neuropathology, only one case has been reported—that of a patient with a right posterior cerebral artery (PCA) infarction who manifested AHS within the context of opticosensory ataxia.⁹ Doody and Jankovic¹⁰ have stated that a large posterior infarct alone may be sufficient to produce AHS, but such a case has not been reported. We present a case of an individual with an ego-syntonic variant of AHS after right PCA stroke that did not affect the corpus callosum.

CASE DESCRIPTION

A 79-year-old right-handed man underwent percutaneous placement of an aortobiliac endovascular stent for an abdominal aortic aneurysm. Postoperatively, he was found to have left-sided weakness and a left visual field defect. Computed tomography of the head revealed a right PCA distribution infarction. After stabilization, he was transferred to a rehabilitation center near his home. On arrival there, however, he was found to have decreased pulses in his right foot, requiring immediate transfer back to the hospital. The patient underwent right iliac stent placement, and after 1 week was admitted to the rehabilitation unit in the hospital. On the day after his admission, however, he developed pain in his right leg with progressive cyanosis and mottling. He was taken again to the vascular lab where he underwent thrombolysis and stent placement in the right iliac artery. When the arteries showed continued patency, the patient was admitted again to the rehabilitation unit for comprehensive rehabilitation. Magnetic resonance imaging of the brain confirmed an ischemic lesion in the distribution of the right PCA occupying the medial and posterior portions of the right temporal lobes and extending into the right posterior parietal and occipital lobes. However, the corpus callosum was spared (fig 1).

On admission to the rehabilitation unit 2 weeks after initial onset of stroke, the patient had regained most of the motor function in his left side and was fully alert and oriented. Cranial nerve function was intact except for the finding of a left homonymous hemianopsia. He also evidenced neglect of items in his left hemispace despite coaching to "look left." In fact, when attempting to correct for this tendency, he tended to overshoot and look around behind himself to his left side. Speech was fluent but circumlocutory with no evidence of dysarthria, and comprehension was intact. He had normal strength on the right and 4/5 strength on the left. Sensation to

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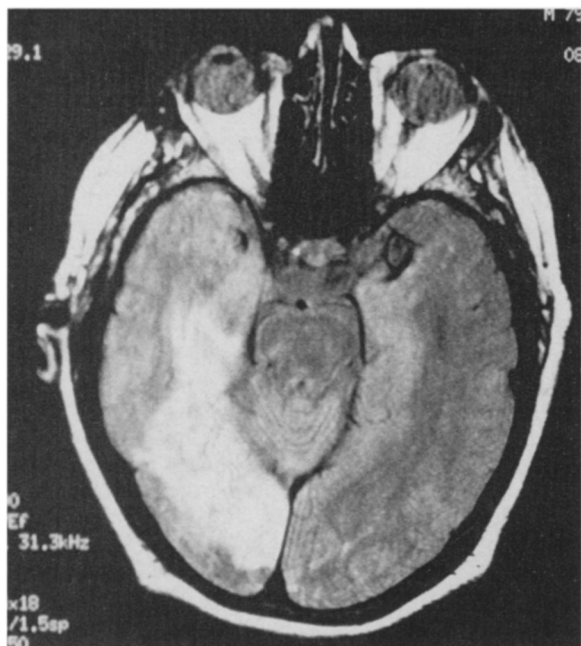


Fig 1. MRI of the brain (approximately 5 weeks after onset of stroke) clearly demonstrates an ischemic lesion in the distribution of the right posterior cerebral artery.

light touch and proprioception was decreased on the left. He also showed pronator drift on the left. Cerebellar functioning was normal on the right but displayed prominent dysmetria on the left.

Affectively, the patient was quite friendly, talkative, and lighthearted. He could usually be observed smiling as he made puns and told stories to staff on the unit. He was anosognostic as to his limitations, acquired only a vague understanding of his medical status, and had difficulty appreciating the need for continued rehabilitation with respect to optimizing long-term outcome. Early in the hospitalization, thought content focused on his hemisensory loss and the antics of his left arm.

Without prompting, the patient introduced his left arm into the examination as "my little buddy." He described how he (his left arm) could do things for him (the patient) like massaging his head. After this statement, his left hand indeed was noted to grope ataxically until it found his scalp and began massaging his head. He acknowledged his lack of control of the arm by stating that "he gets a little rambunctious at times." At one point, when he complimented the examiner's tie, his left hand immediately and impulsively reached for the tie, somewhat to his own surprise. Additionally, mirror movements were observed in the left hand when he was asked to write with his dominant right hand.

In occupational therapy, the patient was encouraged to integrate both hands as he practiced performing activities of daily living. Therapists found it useful to bring his left hand into his right hemispace to increase visual feedback as he worked on coordination. Therapists also discouraged the patient from referring to his hand in the third person and used his verbal abilities to compensate for visual-motor problems. His involuntary movements evolved in conjunction with ongoing therapy. Initially he displayed hemiballistic-type movements in the left arm and akathisia of the left leg, which progressed to self-restriction of the left arm by the right and no involuntary left leg movements by 4 weeks poststroke. He regained volitional, yet

ataxic, movement of the arm by week 6, but with continued personification. On outpatient follow-up at 10 weeks post-stroke, bimanual coordination was only mildly impaired, but he still referred to his left arm in the third person.

Neuropsychologic testing. As is frequently seen with a right-hemisphere stroke, intellectual assessment revealed an extreme disparity between an estimated verbal IQ of 113 (81st percentile, high-average range) and performance IQ of 72 (3rd percentile, mildly impaired range).

Visual spatial processing. Assessment of visual perception revealed severely impaired visual discrimination of simple geometric figures and low-average visual closure. Visual sequencing of pictures in a logical, story format was low average. Visual motor integration was moderately impaired for block constructions and putting puzzles together. Visual abstract analogic reasoning was discontinued secondary to a strong response bias toward choosing answers on the right side of the page. It was unclear as to what proportions of sensory effects from left hemineglect and homonymous hemianopsia, and primary visual problem-solving difficulty contributed to the impairments, but it is likely that both hindered performance.

Attention/memory. Attention was average for verbal information such as forward and backward repetition of digits. Immediate graphomotor recall of line drawings was severely impaired, whereas recall after a 30-minute delay was mildly impaired. However, immediate and delayed recall of verbal information presented within a story format were both very superior. Similarly, paired associative learning of likely and unlikely word combinations was high average.

Language. Language abilities were functionally intact for conversation, and there were no paraphasias. Apraxia was not present in either hand for buccofacial or ideomotor commands. The patient demonstrated high-average sight-word reading vocabulary and superior expressive vocabulary, both of which are consistent with at least a high-average premorbid IQ. Despite occasionally skipping lines, he was able to read the newspaper aloud. Cognitive production rate as measured by verbal fluency was average.

Executive. Executive visuomotor sequencing of numerals was severely impaired, as was set shifting for sequencing numerals alternating with letters, presumably due to difficulties with visual searching to the left. Speed of reading color names in columns was severely impaired. Speed of identification of colors was borderline, whereas performance of the task in the face of interfering verbal stimuli was average. This pattern is indicative of better preserved frontal lobe functions in the presence of cognitive slowing and more posterior pathology.

DISCUSSION

AHS has been postulated as a manifestation of an interhemispheric disconnection of the motor and supplementary motor areas.⁷ A classic disconnection syndrome involves a lesion in a neuronal pathway that disrupts the transfer of information from one region of the brain to another.¹¹ Such is the case in conduction aphasia, wherein auditory signals from the Wernicke area are interrupted en route to the Broca area. Another paradigmatic disconnection syndrome is seen in patients after cerebral commissurotomy. Traditional views state that surgical severance of corticocortical pathways in the corpus callosum leads to two hemispheres operating in isolation from one another. However, recent reports suggest that, despite a callosal transection, interhemispheric transfer of information can take place subcortically, albeit at a more implicit, unconscious level.^{11,12} This implies that alternate pathways do exist between the two sides.

One may then question whether a corpus callosum lesion is essential in AHS. Most authors have emphasized lesions in the corpus callosum or the medial frontal lobe in AHS.^{4,7,13} Even the patient described by Levine and Rinn,⁹ who had right PCA occlusion, had involvement of the splenium of the corpus callosum. In contradistinction to these cases, our patient demonstrated that lesions in the PCA distribution may produce AHS without damage to these areas. Similarly, Ventura and associates⁸ described a case of AHS in a patient with a thalamic hemorrhage but no callosal lesion. Perhaps the disconnection occurs indirectly. An example is the medial paralimbic system, which is believed to contribute to the perception of volitional control over actions. Damage to the medial paralimbic fibers can lead to a loss of intentionality and a sense of foreignness of the affected limb.¹⁴ The tracts originate from the hippocampal region of the medial temporal lobe and from the basal ganglia and extend to the anterior cingulate gyrus, which abuts the supplementary motor area. The supplementary motor area, in turn, connects with its contralateral counterpart via the corpus callosum. Thus, a lesion within this network may, in effect, act like a corpus callosum lesion. Alternatively, the perception of foreignness in our patient may be the result of a parietal lesion. The sensory and visuospatial deficits in this patient are akin to hemiasomatognosia, which results from damage to the right inferior parietal region. The syndrome includes denial of motor deficits, hemineglect, sense of strangeness of a limb, and feeling that a body part does not belong.¹⁵ An inspection of the neuroimaging of our patient reveals that parietal and medial temporal fibers were affected by this infarct (fig 1). These interpretations may provide the basis for a possible mechanism of dysfunction in our patient.

The patient met all major criteria for the diagnosis of AHS: lack of recognition of his hand, involuntary purposeful movements outside his volitional control, and personification of the hand.¹⁰ Additionally, he showed the associated features of self-restriction, impulsive grasping and groping, impaired bimanual coordination, and mirror movements.⁶ Nevertheless, this may be the first known case of an ego-syntonic perception of one's alien hand that was viewed as acceptable and benign, albeit as an autonomous force.

Levine⁹ suggested that premorbid personality characteristics may predispose one to manifest AHS.⁹ Our case begs the more pertinent question of whether the neurobehavioral symptoms associated with brain pathology of particular regions makes one more likely to experience AHS and, when experienced, to flavor the presentation in unique ways. Although our patient was quite perplexed by the behavior of his hand, he had no reason to think it intended to harm him and thus formed a cooperative alliance with it as his "little buddy." The right-hemisphere-stroke syndrome is well documented and includes a unique pattern of neurobehavioral syndromes including anosognosia, denial, secondary mania, and *la belle indifférence* reactions.^{16,17} These neurobehavioral phenomena make it intuitively less likely that a patient who presents with AHS will view the manifestation in ego-dystonic, negative terms.

One must also wonder if the valence of the attribution toward the hand, whether it is positive or negative, has any relevance with regard to the course of the syndrome and patient prognosis. In theory, the ego-syntonic manifestation of AHS may, by virtue of creating less distress, interfere to a lesser extent than the ego-dystonic type. Patients who are overtly frightened may have a natural motivation to normalize their appraisal of the hand. In contrast, our patient appeared to have little reason to abandon the notion of personification due to a lack of distress

caused by the condition, and his AHS remained present even at follow-up 10 weeks postonset. Indeed, as a form of treatment, Levine⁹ instructed his patient to make friends with the arm by talking to it as a misbehaving child.

Review of the previously described attributions of patients toward their alien hand reveals certain distinct negative reactions and beliefs. Reactions include terror, anguish, anxiety, fright, distress, profuse sweating,¹⁷ astonishment, frustration,¹³ and unpleasant sensations.⁶ Bizarre reports of an alien limb holding hostile motivations⁸ or attempting to break into a house have been made.³ Leiguarda and coworkers¹⁸ stated that their patient believed that the alien limb was "like a monster" and "wanted to hurt" her.

Clinicians themselves have attributed harmful motivations to alien hands. For example, Kuhn and colleagues¹⁹ state that their patient's arm performed "destructive movements such as pulling on a feeding tube." This appraisal implies that the arm, of its own will, intends to do harm to the patient. Della Salla²⁰ even includes the negative attributions "unpleasant and frightening" in his definition of the syndrome. However, the fact that several, if not most, case reports of AHS do not report the patient's attributions regarding the alien hand suggests that it is not a standard component of the diagnosis.

The presentation of this unique case requires a re-examination and possible expansion of the location of brain lesions thought to be responsible for AHS to include a large posterior infarct not affecting the corpus callosum. Additionally, as medical nosologists clarify and refine the definition of AHS, they may wish to consider the positive as well as negative attributions that patients make toward their alien members and, ultimately, the potential impact such attributions may have on rehabilitation and outcome.

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