

Case Report

The Alien Hand Syndrome: Report of a Case and Review of the Literature

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The term "alien hand syndrome (AHS)" comprises many clinical signs of which the common features are the involuntary motor movement of the affected limb and the denial of limb ownership. It can result from several diseases involving corpus callosum or medial frontal cortex. Two major types of AHS were previously classified, callosal and frontal types. Moreover, posterior subtype of which the lesions do not involve corpus callosum have been reported. In the present report, the authors describe a 57-year-old man with AHS, aggressive behavior and hemispatial neglect which are the rare manifestations of callosal damage. Neuroimaging demonstrated subacute infarction of entire corpus callosum from the rostrum to splenium. A review of the literature on these abnormalities is included in the present paper.

Keywords: Alien hand syndrome (AHS), Corpus callosum

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Alien hand syndrome (AHS) is a distinctive clinical feature which comprises a variety of clinical conditions. The term alien-hand (*la main trang re*) was first introduced by Brion and Jedynek in 1972 to describe patients with midline brain tumors who denied the ownership of one of their hands⁽¹⁾. Formerly, the phenomenon of this sign was described by Kurt Goldstein in 1908⁽²⁾, and years later (1945) it was described again by Akelaitis⁽³⁾. Akelaitis described a patient whose left hand involuntarily performed the opposite of what she wanted her right hand to do following the section of the corpus callosum and he named this feature as "diagnostic dyspraxia". Since then, there have been a number of reports of this syndrome. A lot of new signs have been reported making a variety of this syndrome. In Thailand, there have been only two reports of AHS⁽⁴⁻⁵⁾. This is another report of a patient presenting with AHS, aggression and hemispatial neglect from damage of the whole corpus callosum. These findings are substantially different from previously reported cases.

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Description of the patient

A 57-year old, right handed man had a medical history of diabetes mellitus, hypertension, dyslipidemia, coronary artery disease and peripheral arterial disease. He was admitted for femeropliteal bypass graft. There were no intraoperative and immediate postoperative complications. One week postoperatively, the patient's caregiver and nurses observed that he was unmotivated as well as disinterested and spoke much less than he used to do. However, he could speak and comprehend correctly. His caregiver also complained of the patient's verbal and physical aggressiveness. They thought that the patient might have depressed mood, hence they did not inform the attending physician promptly. After several days of observation, the patient rarely cooperated to ambulate, performed the basic activities of daily living independently. He had intermanual conflict which was observed by his caregivers occasionally. For instance, his left hand grabbed a television remote-control from his right hand and he complained that his left hand robbed it. He occasionally had visual hallucinations. No other medical conditions interfering with cognitive functions were revealed, then neurological consultation was performed.

Neurologic examination revealed normal cranial nerve, motor, and sensory function. The myostatic reflexes were normal. No pathological reflexes could be elicited. The sensory system was normal. Verbal fluency was markedly reduced, however, he could follow and cooperate with commands well. There were some literal paraphasia. The patient had no alexia but he had agraphia of his left hand. His left hand always mimicked the movement of his right hand. When asking the patient to perform some actions with his left hand, he always used his right hand to do the tasks instead. Moreover, he had finger agnosia, graphesthesia and astereognosia on his left hand. He could not perform complex motor action on verbal command, pantomime and use real objects by his left hand. The clock drawing test is shown in Fig. 1. The tactile and visual double stimulation tests were positive on the left side.

Cranial magnetic resonance imaging (MRI) performed after two weeks of symptoms demonstrated mixed low and high signal intensity on T1W and high signal intensity on T2W at corpus callosum (rostrum, body and splenium) which was markedly enhanced after gadolinium injection suggestive of subacute infarction. Frontal lobes had mild cortical atrophy. Cranial MRA demonstrated mild irregularity of supraclinoid right ICA more than left ICA. The carotid duplex ultrasound and MRA of carotid arteries showed mild stenosis of both sides.

Discussion

The term “alien hand” is used to describe the distinctive clinical syndrome in which an upper limb performs autonomous complex movements against the patient’s will. However, this term is usually confusing. There are 5 classifications of “alien hand syndrome (AHS)”⁽⁶⁾ as summarized in Table 1.

Specific alien-hand signs in most reports include⁽¹⁷⁾ 1) intermanual conflict⁽⁷⁾ 2) mirror movement, in which one hand automatically mimics the movement of the other hand 3) enabling synkinesis, in which one hand can perform the action only in unison with the other 4) grasp reflex 5) groping or magnetic apraxia⁽¹⁸⁾, in which the affected hand reaches toward and grasps objects as if drawn to them by a magnet and release of the objects is difficult (6) utilization behavior or compulsive manipulation of tools 7) negative feeling toward the affected limb in most cases.

The present patient had intermanual conflict and reduced verbal output. He also had an agonistic dyspraxia which was shown by the use of his right hand to perform commanded actions of his left hand.

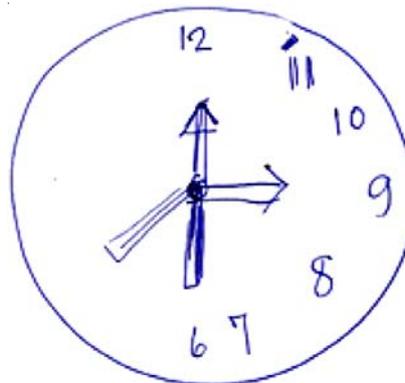


Fig. 1 Clock Drawing Test (with frame) of the patient when he was told to draw 10 minutes past 11 o'clock



Fig. 2 Cranial MRI (T1W with gadolinium injection) shows mixed low and high signal intensity at corpus callosum which is markedly enhanced after gadolinium injection

Table 1. Demonstrates 5 major classifications of AHS

Classifications	Characteristics
Diagnostic dyspraxia and intermanual conflict	the left hand (in right-handed subjects) performs actions contrary or opposite to, or interfere with, the actions of the right hand ^(3,7,8) .
Alien hand sign	usually affects the left hand; subjective feeling that the hand does not belong to the patients ⁽¹⁾ .
Anarchic hand or way-ward hand	no denial of limb ownership but the affected hand, being contralateral to the lesion, performs goal-directed movements that the patient does not perceive as initiated or controlled by his own will ⁽⁹⁻¹¹⁾ .
Supernumerary hands	the patient reports the feeling of having an extra extremity ⁽¹²⁻¹⁵⁾ .
Agonistic dyspraxia	the patient uses the hand on the opposite side of a motor command, while the “instructed” hand remains immobilized ⁽¹⁶⁾ .

There were ideomotor apraxia, agraphia, literal paraphasia, finger agnosia, astereognosia, graphesthesia and visuospatial neglect of the left side. The simultaneous bilateral stimulation in visual and tactile modalities demonstrated the impairment in detecting the stimuli applied to his left side. A lesion in many areas in the brain can cause contralesional hemispatial neglect such as the cingulate cortex, posterior parietal cortex, frontal eye fields, striatum and thalamus⁽¹⁹⁾. To our knowledge, this is the first report of hemispatial neglect in patients with only callosal involvement. Another rare manifestation of callosal damage in the present case is the development of aggression. The etiology of aggression is complex and it can be the result of damage to some focal areas in the brain (brainstem, hypothalamus, amygdala, temporolimbic cortex, and prefrontal cortex), neurotransmitter modulation (serotonin, acetylcholine, catecholamine, and GABA), hormonal changes (testosterone and other androgens), delirium, mood state and provoking stimuli⁽²⁰⁾. There have been only a few reports citing the behavioral and mental problems after callosal damage such as callosal dementia, antisocial behavior, agitation and deficits in social intelligence⁽²¹⁻²³⁾. The present patient was also mildly apathetic, as shown by being unmotivated and disinterested in doing things, and had mild nonfluent verbal output.

There are several causes of AHS such as stroke, tumor, epilepsy, multiple sclerosis, migraine, corticobasal degeneration, Marchiafava-Bignami disease and after callosotomy. Stroke might be from infarction of anterior cerebral artery which is the most frequent vascular involvement in this phenomenon. It

can also be caused by the rupture of aneurysms near the anterior cerebral artery. The anterior cerebral artery supplies blood to medial frontal areas and to the anterior two-thirds of the corpus callosum. The posterior splenic branch of posterior cerebral artery supplies splenium of corpus callosum. The present patient had the rare infarction of the entire corpus callosum (rostrum, body and splenium) without involvement of other sites as demonstrated on the MRI of the brain. The most likely vascular localization should be pericallosal artery with the variation to supply splenium of corpus callosum but the occlusion was not demonstrated because of the lag period after the event and imaging as well as the inferior sensitivity of MRA to detect small arterial occlusion to cerebral angiography. Nevertheless, the MRI of the brain can still demonstrate the lesion with high sensitivity even two weeks after the onset of the symptoms.

The presented patient had callosal type of the ideomotor apraxia. The ideomotor apraxia was classified into three subtypes as shown in Table 2⁽²¹⁾.

There are two main subtypes of AHS, callosal and frontal⁽²⁵⁾. Patients with the former subtype will show disruption of complex willed motor act of the nondominant hand (intermanual conflict or diagnostic dyspraxia). In addition to callosal-subtype alien-hand signs, the frontal-subtype cases will show grasp reflex, groping and compulsive tool manipulation by the dominant hand (frontal-subtype alien hand signs). This may result from the damage of medial frontal lobe which is often accompanied by the damage of the corpus callosum. Hence, the frontal-subtype cases may also show the callosal-subtype alien-hand signs⁽⁷⁾. Most cases

Table 2. Shows the characteristics of three ideomotor apraxia⁽²⁴⁾

Type of apraxia	Lesion location	Apraxic limbs	Hemiparesis	Aphasia
Parietal	Inferior parietal, arcuate fasciculus	Right and left	None	Conduction
Sympathetic	Frontal lobe	Left	Right	Broca's
Callosal	Anterior callosal fibers	Left	None	None

Table 3. Summarizes the differentiation between the two subtypes of AHS

Clinical & anatomical differences	Callosal type	Frontal type
Anatomical lesion	Corpus callosum	Medial frontal cortex (premotor & supplementary motor areas, anterior cingulate gyrus)
Intermanual conflict/ diagnostic dyspraxia of the nondominant hand	Yes	Often
Grasp reflex/ groping/ compulsive tool manipulation by the dominant hand	No	Yes

of alien hand signs are seen in patients who suffered damage to the medial frontal cortex with accompanying damage to the corpus callosum. The clinical and anatomical differences between these two subtypes are shown in Table 3.

Most reports of the alien hand signs are from anterior corpus callosal lesion. However, there are reports of posterior AHS of which the lesions are confined to sites other than corpus callosum such as the thalamus, parietal and temporal cortex⁽²⁶⁻³¹⁾. In these cases, patients usually have “*levitating hand*”, in which the hand contralateral to the lesion levitates aimlessly. Moreover, there are typically other signs showing cortical involvement such as anosognosia (denial of hemiparesis) and left-sided spatial neglect in cases having right parietal lesion and Balint syndrome. However, the details of the behaviors and neurologic findings are less described than that of anterior AHS.

This is one of the cases who developed interhemispheric disconnection syndrome from corpus callosal infarction. The right and left hemisphere are disconnected. Another example of disconnection syndrome is “*alexia without agraphia*” of which the typical lesions are located at left occipital lobe and splenium of the corpus callosum. The right occipital lobe perceives the written language but is unable to transfer it to the left occipital lobe for language translation because of the posterior corpus callosal lesion. The presented patient also had infarction of splenium

but did not show “*alexia without agraphia*” because he had no left occipital lesion.

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กลุ่มอาการเอเลียสแฮนด์: รายงานผู้ป่วย 1 รายและบททบทวนวรรณกรรม

วิรัชศักดิ์ เมืองไพศาล, สิทธิพงศ์ ศรีสัจจากุล, พิพัฒน์ เชี่ยววิทย์

กลุ่มอาการเอเลียสแฮนด์ประกอบไปด้วยอาการแสดงหลายอย่างที่สำคัญคือการเคลื่อนไหวของระยางค์โดยควบคุมไม่ได้และการปฏิเสธความเป็นเจ้าของระยางค์นั้น. สาเหตุที่ทำให้เกิดกลุ่มอาการนี้เกิดได้จากหลายโรคที่มีการทำลายคอร์ปัสคอลลโลซัมหรือสมองฟรอนทัลด้านใน ได้มีการแบ่งความผิดปกติของกลุ่มอาการนี้ออกเป็นสองแบบคือ ชนิดคอลลโลซัมและชนิดฟรอนทัล. นอกจากนี้ยังมีหลายรายงานแสดงให้เห็นถึงความผิดปกติของสมองส่วนหลังที่ทำให้เกิดกลุ่มอาการชนิดนี้โดยที่ไม่มีความผิดปกติของคอร์ปัสคอลลโลซัมหรือสมองส่วนฟรอนทัล ในรายงานนี้คณะผู้นิพนธ์ได้นำเสนอผู้ป่วยชายอายุ 57 ปีที่เกิดกลุ่มอาการเอเลียสแฮนด์ พฤติกรรมรุนแรงก้าวร้าวและไม่สนใจร่างกายและสิ่งแวดล้อมด้านซ้ายซึ่งเป็นภาวะที่พบไม่บ่อยที่เกิดจากรอยโรคที่คอร์ปัสคอลลโลซัม. ภาพเอกซเรย์สมองพบลักษณะที่เข้าได้กับสมองขาดเลือดโดยรอยโรคอยู่ที่คอร์ปัสคอลลโลซัมตั้งแต่ส่วนต้นจนปลาย. ในช่วงทำยรายงานผู้นิพนธ์ได้ทบทวนวรรณกรรมเกี่ยวกับความผิดปกติเหล่านี้ด้วย
