

Alien Hand Syndrome: Introduction, literature review and overview**Hassan I. Osman *(1), Rudaina I. Osman (2)*****(1) Medical student, Napata College, Khartoum, Sudan****(2) Medical student, Napata College, Khartoum, Sudan****Corresponding Author:****Hassan I. Osman, Medical Student, Napata College, Khartoum, Sudan****Email: hassanismail603@gmail.com****hassan.io@live.co.uk****Article type: Review Article****Abstract**

Few disorders/syndromes are surrounded by as much mystery as Aline Hand Syndrome (AHS) – a rare neurological disorder of connection which ergo results in misinterpretation and a loss of ownership of a limb (usually upper). In this paper, we aim to bring forth enough light on AHS so that more clinicians can correctly diagnose AHS, research efforts increase and awareness. It is, undoubtedly, misdiagnosed en masse on a daily basis as a result of our failure to properly introduce it in medical literature. A phenomenon of which we have a fair amount of understanding, yet have yet to teach it in medical references. In our search for AHS in medical references, we came across a total of only 1 reference which discussed AHS. It is important that we note that the number of references we searched was 16 and that these were well-known references which medical students and practitioners read on a daily basis all around the globe.

Introduction

Very few disorders which affect humans are as frightening to experience as AHS (also known as Dr. Strangelove syndrome),^[2] for measure – imagine this:

You're riding in a bus headed somewhere, when all of a sudden a hand approaches you attempting to grasp your trousers. At first, you assume you're being invaded by another passenger; to your unpleasant surprise; you realize the foreign hand was actually your own, you had lost your sense of ownership of said limb. The previous scenario isn't fictitious in nature, but is 100% true and was lived by a 65-year old man in a reported case^[1].

The man suffered a frightening experience of AHS as a result of focal atrophy in his left medial frontal cortex^[1]. As we shall later explore, AHS has multiple forms and may manifest itself in multiple patterns.

History of AHS

First described back in 1908 by K. Goldstein who published a case report discussing involuntary movements of the left hand following a stroke;^[4,5,8] AHS has managed to grasp the interest of all those who have encountered it. Although first explained in 1908; it wasn't until 1972 that the term AHS was coined^[4,6]. That was done by Brion and Jedynak who had observed three patients with callosal tumors who were failed to recognize their own hands.^[4,6] In 1991^[4,7], Dell Salla had made a breakthrough which helped make AHS clearer to understand, Salla explained that more than one form of AHS existed, they classified AHS into:

- 1) Acute, and
- 2) Chronic

Acute AHS was a result of callosal lesions, while chronic AHS was a result of callosal and anteromedial frontal lesions^[4,7]. Later in this article, we will be discussing the aforementioned types of AHS as well as a 3rd one which was relatively recently discussed.

Clinical manifestations & Variants of AHS

The most prominent clinical manifestation of AHS is the sudden involuntary movement of the limb and the inability to stop these horrifying uncontrolled movements as well as autonomic activity.^[1]

The feeling of the hand not being their own is a major complaint in AHS patients, and when the affected limb levitates without their will.^[1]

Some patients even feel like they have an extra limb.^[1]

In some extreme cases, the “alien hand” could even suffocate the patient,^[2] showing us how dangerous this could be.

Patients are usually above the age of 60, and throughout their life had a neurological disorder affecting the regions of the brain, like the corpus callosum, parietal region, or frontal region,^[2] ischemic attacks and CVA occlusion.

Until now it has been given 3 subtypes frontal, callosal and posterior AHS subtypes.^[3]

As aforementioned, a total of 3 variants of AHS exist in the literature, with the posterior variant being a relatively recent discovery.^[4,9]

Both the frontal and colossal variants are sometimes collectively termed anterior variants; however, it is important to note that

they differ exponentially in their clinical manifestations, ^[4,1] in their malfunctioned areas and in their treatment. Because of that we will be using the 3 classification method of AHS. So, what are the 3 variants of AHS?

- 1) Frontal,
- 2) Colossal, and
- 3) Posterior ^[3,4,9]

Each, as we shall now observe, with different clinical manifestations, brain imaging results, and treatment.

First, we will be discussing the affected areas of the brain, after which we will take a quick glance over the clinical manifestations of each and every area.

AHS is classified as caused by a frontal lesion when that lesion affects the SMA, cingulate cortex and dominant medial prefrontal cortex. ^[4]

AHS is classified as caused by a callosal lesion when that lesion affects the corpus callosum

AHS is classified as caused by a posterior lesion when that lesion affects thalamic, posterolateral parietal, or occipital lobe.

Each of these variants manifests itself in a different manner; clinically speaking.

Anterior AHS will most likely manifest itself as “groping (where the hand seems to be constantly searching for nearby objects), grasping, or compulsive manipulation of tools.” ^[4,10]

Callosal AHS will most likely manifest itself as intermanual conflict (commonest in lesions of the anterior 3rd of the rostrum). ^[4,11,12] Geschwind et. al., ^[4,13] reported in 1995 the case of a 68-year old woman who experienced interhemispheric disconnection syndrome following the experience of intermanual conflict which had manifested following a callosal infarction. Her IDS manifested itself in a triad of symptoms:

- 1) tactile anomia

- 2) ideomotor apraxia, and
- 3) agraphic aphasia

As hypothesized, AHS possess the capability to cause psychological distress to patients experiencing. The literature proves this hypothesis ^[4,11]; as the referenced papers state that AHS “can exhibit autocriticism” ^[4] which is a state of distress caused by intermanual conflict. Sometimes patients will actually sit on their alien limb to avoid the looks of bystanders. ^[4,11] You might expect this to occur as a result of callosal lesions only, but surprisingly it does occur in cases of frontal AHS, albeit rarely. ^[4,11]

Regarding the posterior version of AHS, it will most likely manifest itself in the form of avoidance response, a failure to coordinate hand movements or involuntary levitation. ^[4,14] In some instances, accompanying symptoms might manifest themselves, these include hemianesthesia, hemianopia, visuospatial neglect, ^[4,15] and optic ataxia. ^[4,16]

Treatment

As far as we can tell, the treatment of AHS seems to be highly dependent on the morbidity and mortality of whatever disorders might have brought it forth to begin with. So far, it seems that following successful treatment of the cause will result in the disappearance of AHS. Psychiatric consultation might prove necessary in particularly frightening manifestations of AHS.

Clinical cases from the literature

The following are copied from different case reports; they aim to increase awareness of the possible manifestations of AHS.

- 1) Paroxysmal alien hand syndrome ^[1]

Case 1:

'A 65-year-old right handed man reported the following: "While I was travelling on a bus I noticed that a hand was approaching me on the right from behind, trying to catch me. After grasping my trouser leg, the hand did not release it. First, I thought somebody was assaulting me, but then I realised that it was my own right hand, although I did not feel it belonged to me. Thereafter, the fingers developed creeping movements, and repetitive jerks involving the whole arm soon followed. I was unable to control my right hand and I had to grasp and hold it with my left hand. My right arm felt heavy and awkward. I was very anguished, anxious and frightened, and had palpitations. . ." The episode lasted for a few minutes, and recurred briefly that evening at the patient's home: "I couldn't sleep at all that night because I was terrified my right hand would assault me while sleeping." Neurological, neuropsychiatric, and general examinations yielded normal results. Laboratory analyses (haemoglobin concentration, white 7 blood cell count, erythrocyte sedimentation rate, blood glucose, blood urea, plasma electrolytes, calcium ions, Venereal Disease Research Laboratory test, lipids, liver function test, protein electrophoresis, coagulation studies, T3 and T4, urinalysis, electrocardiography, Doppler ultrasound of neck and intracranial vessels, echocardiogram, and interictal electroencephalography) also yielded normal results

Computed tomography showed focal atrophy restricted to the left medial frontal cortex. The patient was prescribed carbamazepine (800 mg a day), and the episodes did not recur during the next two years.'

Case 2:

This 54-year-old right handed man experienced two transient episodes of seemingly purposeful movements of his left arm which he interpreted as foreign ("the arm acted independently of my own will, certainly it was not mine, the arm was driven by somebody

else, it felt quite superfluous"). The first episode took place while he was driving. The patient was forced to stop the car because his left arm had an uncontrollable tendency to grasp and pull the steering wheel in a chaotic way. The second episode took place a week later while he was undressing. The left limb grasped and pulled up his trousers in a perseverative way, and he had to sit down until this involuntary behaviour stopped. He was very distressed and frightened and had profuse sweating. On examination, the patient was CTscan showing enhanced right frontal parasagittal lesion mainly affecting supplementary motor area. alert, oriented, and cooperative. Cranial nerves and motor, sensory, and cerebellar functions were all normal. Interictal electroencephalography yielded normal results. CT showed a right parasagittal intra-axial mass just in front of the motor strip. Surgery was performed, and a diffuse lymphocytic lymphoma was removed. The post operative period was uneventful with a mild residual weakness in the arms that disappeared after a few weeks. He was treated with diphenylhydantoin and had no symptoms during the next three years.

References

- Guide = Author(s); year; title;
Journal/Publisher
- 1) Ramon Leiguarda,et. al.; 1993;
 Paroxysmal alien hand syndrome ;
Journal of Neurology, Neurosurgery, and Psychiatry;
 56:788-792
- 2) Ragesh Panikkath, et. al.; 2014; The
 alien hand syndrome; *Proc (Bayl Univ Med Cent)*; 27(3):219–220
- 3) Bekir Enes Demiryu`rek1; 2016;
 Paroxysmal posterior variant alien
 hand syndrome associated with
 parietal lobe infarction: case

- presentation; *Cogn Neurodyn* 10:453–455
- 4) Harini Sarva, et. al.; 2014; Pathophysiology and Treatment of Alien Hand Syndrome; *Tremor and Other Hyperkinetic Movements*
 - 5) Brainin M, Seiser A, Matz K.; 2008; The mirror world of motor inhibition: The alien hand syndrome in chronic stroke.; *J Neurol Neurosurg Psychiatry*;79:246–252, doi:<http://dx.doi.org/10.1136/jnnp.2007.116046>
 - 6) Brion S, Jedynak CP.; 1972; [Disorders of interhemispheric transfer (callosal disconnection). 3 cases of tumor of the corpus callosum. The strange hand sign].; *Rev Neurol (Paris)*; 126:257–266.
 - 7) Biran I, Chatterjee A.; 2004; Alien hand syndrome. *Arch Neurol*; 61:292–294, doi: <http://dx.doi.org/10.1001/archneur.61.2.292>.
 - 8) Yong Won Park; 2012; Alien Hand Syndrome in Stroke- Case Report & Neurophysiologic Study -; *Ann Rehabil Med*; 36: 556-560 pISSN: 2234-0645 • eISSN: 2234-0653 <http://dx.doi.org/10.5535/arm.2012.36.4.556>
 - 9) Kloesel B, Czarnecki K, Muir JJ, Keller AS.; 2010; Sequelae of a left-sided parietal stroke: Posterior alien hand syndrome. *Neurocase*; 16:488–493, doi: <http://dx.doi.org/10.1080/13554794.2010.497154>.
 - 10) Huang Y, Jia J.; 2013; Corpus callosum hematoma secondary to cerebral venous malformation presenting as alien hand syndrome.; *Neurocase*;19: 377–381, doi: <http://dx.doi.org/10.1080/13554794.2012.690420>.
 - 11) Brainin M, Seiser A, Matz K.; 2008 The mirror world of motor inhibition: The alien hand syndrome in chronic stroke.; *J Neurol Neurosurg Psychiatry* 2008;79:246–252, doi: <http://dx.doi.org/10.1136/jnnp.2007.116046>.
 - 12) Feinberg TE, Schindler RJ, Flanagan NG, Haber LD.; 1992; Two alien hand syndromes. *Neurology*;42:19–24, doi: <http://dx.doi.org/10.1212/WNL.42.1.19>.
 - 13) Geschwind DH, Iacoboni M, Mega MS, Zaidel DW, Cloughesy T, Zaidel E.; 1995; Alien hand syndrome: Interhemispheric motor disconnection due to a lesion in the midbody of the corpus callosum. *Neurology*; 45:802–808, doi: <http://dx.doi.org/10.1212/WNL.45.4.802>.
 - 14) Kloesel B, Czarnecki K, Muir JJ, Keller AS.; 2010; Sequelae of a left-sided parietal stroke: Posterior alien hand syndrome. *Neurocase*; 16:488–493, doi: <http://dx.doi.org/10.1080/13554794.2010.497154>.
 - 15) Yuan JL, Wang SK, Guo XJ, Hu WL.;2011; Acute infarct of the corpus callosum presenting as alien hand syndrome: Evidence of diffusion weighted imaging and magnetic resonance angiography.; *BMC Neurol* ;11:142, doi: <http://dx.doi.org/10.1186/1471-2377-11-142>.
 - 16) Levine DN, Rinn WE.; 1986; Optic sensory ataxia and alien hand syndrome after posterior cerebral artery territory infarction. *Neurology*; 36:1094–1097, doi: <http://dx.doi.org/10.1212/WNL.36.8.1094>.

Abréviations :

AHS = Alien Hand Syndrome

SMA= Supplementary Motor Area

IDS = Interhemispheric Disconnection Syndrome

Key Words:

Alien Hand Syndrome, AHS, Stroke, Neurology, Rare

The authors hereby declare no conflicts of interest.