

Alien Hand Syndrome: A Case Report

Decoding the Enigmatic Limb

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ABSTRACT

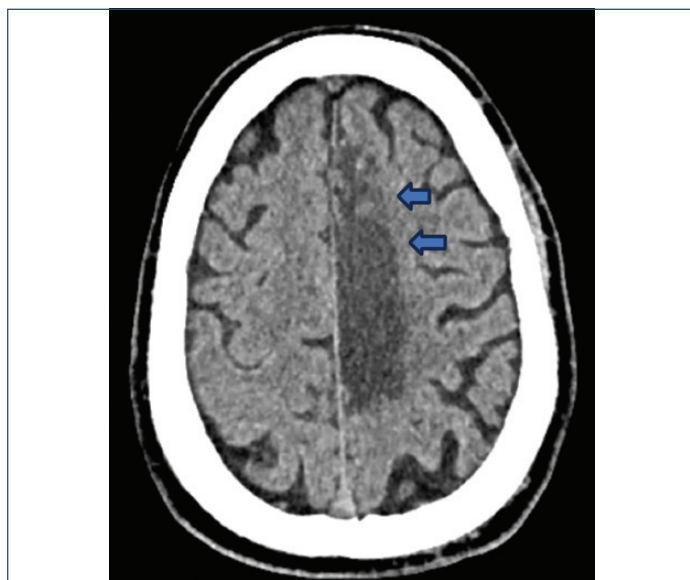
Alien Hand Syndrome (AHS) is a rare condition characterised by involuntary hand movements occurring without the patient's intention. While AHS has various causes, it is frequently associated with ischemic stroke, post-corpus callosotomy, degenerative conditions, and has more recently been observed in cortico-basal degeneration. This case report highlights a 62-year-old male patient experiencing involuntary movement in his right hand three days after admission with an ischemic stroke involving the left fronto-parietal lobe and corpus callosum. The predominant symptoms included compulsive groping, inter-manual conflict, and hemi-anaesthesia in his right hand. The callosal subtype arises from corpus callosum damage and often involves inter-manual conflict. However, callosal infarction is uncommon due to the abundant blood supply. This case report describes a case of AHS caused by callosal infarction, confirmed through magnetic resonance imaging, specifically showing infarction in the left corpus callosum. The patient exhibited symptoms consistent with the anterior variant based on radiographic findings. However, features of the posterior variant were also observed.

Keywords: Autonomous Hand, Callosal Stroke, Cortico-Basal Syndrome, Dr. Strangelove Syndrome, Involuntary movements

CASE REPORT

A 62-year-old male patient, was admitted to the emergency department for a new onset of right-sided weakness affecting both upper and lower limbs for the last two days. Initial symptoms included slurring of speech and right-sided mouth deviation. The patient had a history of a stroke five years ago resulting in right hemi-paresis. On neurological examination, his nutrition and bulk were moderate, with 3/5 power in the right upper and lower limb, hypertonic tone, brisk deep tendon reflexes, and extensor plantar response. Hemi-sensory loss was present on the right side of his body. The patient had a past medical history of hypertension and diabetes mellitus for the past seven years under regular medication and follow-up. There was no history of smoking, alcohol, or illicit drug use.

An emergency Computed Tomography (CT) Brain showed an acute infarct in the anterior cerebral artery territory involving the left frontal lobe and basal ganglia, with a focal area of hypodensity in the left centrum semi-ovale [Table/Fig-1].



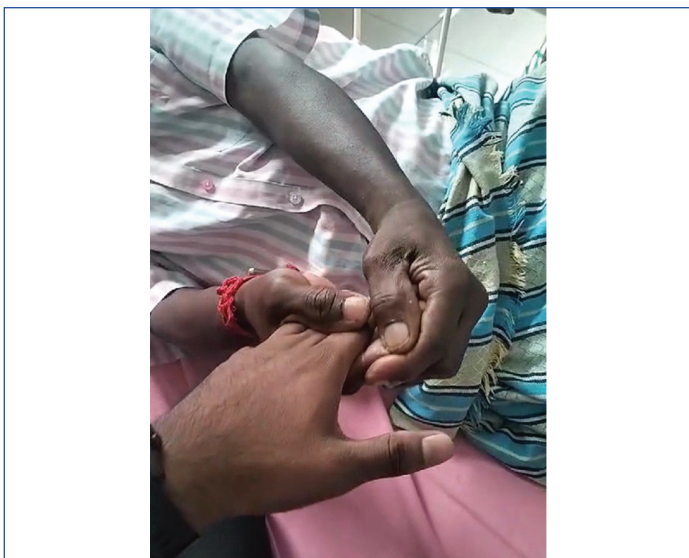
[Table/Fig-1]: Computed Tomography (CT) brain: The axial view reveals a prominent infarct in the left frontal lobe at the left paramedian aspect.

No midline shift or haemorrhagic transformation was observed. In light of recurrent strokes, further evaluation was performed.

Comprehensive blood investigations, including complete blood count, serum electrolytes, renal function test, liver function test, Erythrocyte Sedimentation Rate (ESR), C-reactive Protein (CRP), serum homocysteine, D-Dimer, activated Partial Thromboplastin Time (aPTT), Prothrombin Time- International Normalised Ratio (PT-INR), coagulation profile, peripheral smear, and Fluorescent Treponemal Antibody- Absorption Test (FTA-ABS), yielded normal results, except for a mild elevation in serum creatinine of 1.4 mg/dL and an elevated triglyceride level of 387 mg/dL. An echocardiogram revealed a global hypokinesia of the left ventricle with mild left ventricular systolic dysfunction, and an ejection fraction of 56%. The patient was noted to have 80% stenosis of the left carotid artery on carotid-vertebral doppler. An Electro-cardiography (ECG) revealed sinus rhythm. The patient was started on Dual anti-platelet therapy (Aspirin 75 mg QD and Clopidogrel 75 mg QD), Rosuvastatin 20 mg QD, Perindopril 4 mg BD, and a Basal-Bolus insulin regimen.

On the third day of hospitalisation, the patient reported involuntary movements of his right hand, experiencing jittery movements, and difficulty releasing his shirt, which he involuntarily grabbed, requiring the use of his left hand to control it [Table/Fig-2,3].

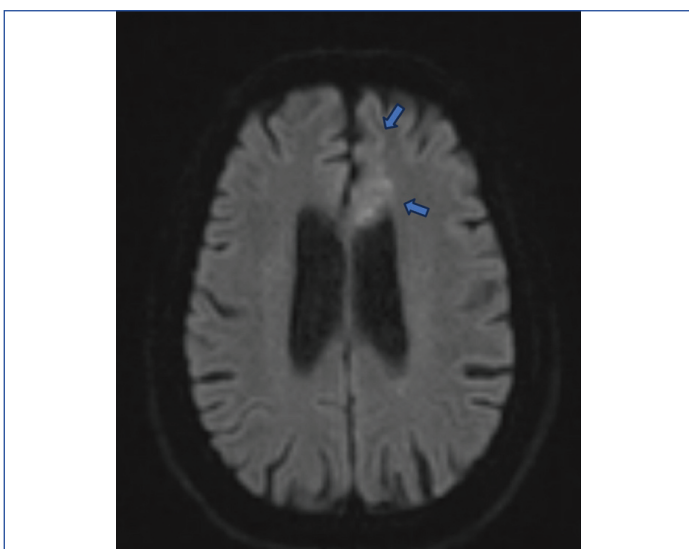
Distressed by the abnormal hand movement, he expressed his emotional anguish and asserted to his wife that these movements were not under his control, and pleaded for help. An Magnetic Resonance Imaging (MRI) of the brain was conducted, revealing a sub-acute infarct in the genu of the corpus callosum, left fronto-parietal cortex, and peri-ventricular white matter [Table/Fig-4]. AHS was diagnosed based on the patient's history and neurological examination. Initially, cognitive behavioural therapy was recommended for the patient. Due to persistent stress and agitation, verbal cue rehabilitation was initiated after further reviews. In this approach, the patient's attendant guided and re-assured the patient while experiencing conflicting movements to complete the intended movement while ignoring the disturbances caused by the alien hand. The patient adhered to the verbal cue rehabilitation for one week during his in-patient stay, leading to reduced agitation and improved functional normalcy by Day 13 of hospitalisation. This



[Table/Fig-2]: The patient involuntarily gropes the examiner's hand and attempts to remove it with his left hand.



[Table/Fig-3]: The patient involuntarily grasped the blanket and attempted to remove it.



[Table/Fig-4]: Magnetic Resonance Imaging (MRI) Brain: Diffusion Weighted Imaging (DWI) reveals sub-acute infarcts in the genu of the corpus callosum and left fronto-parietal cortex.

positive progress allowed for the patient's discharge on Day 15, with a scheduled follow-up visit after one week. During the review visit, the patient reported an overall improvement in his well-being, although the alien hand movements persisted, albeit with a slight decrease in frequency. Clopidogrel was discontinued, and the patient continued on aspirin and rosuvastatin.

DISCUSSION

The condition of AHS is characterised by autonomous purposeful movement of the hand without the patient's intention [1]. Individuals

with this condition experience a temporary loss in their ability to control their hand movements. In this condition, the patient's limb moves autonomously, as if it possesses its cognitive and processing capabilities. The patient is aware of the disparities between their intentions and the action carried out by their hand, experiencing a sense of diminished autonomy in hand movements. For instance, when attempting to eat with the dominant hand, the alien hand may seize control, diverting it away from the mouth [2,3].

Similarly, if the individual tries to pick up an object with their unaffected hand, the alien hand may intervene and obstruct the intended action. It manifests through multiple aetiologies such as stroke, corpus callosotomy, hypoglycemic encephalopathy, multiple sclerosis, and posterior reversible encephalopathy syndrome. Cortico-basal syndrome patients have a high incidence rate of 50%, making it a prevalent cause [2]. AHS is not classified as a movement disorder; rather, it is associated with the region of the brain involved. AHS was first defined by Dr. Kurt Goldstein in 1908 [4,5].

AHS is classified into anterior and posterior variants [4]. The anterior variant has frontal and callosal subtypes. The frontal subtype involves lesions in the supplementary motor area, cingulate cortex, and dominant prefrontal cortex, leading to hand behaviours such as groping, grasping, and compulsive disturbances in work-oriented activities [3]. The callosal subtype results from damage to the anterior corpus callosum causing inter-manual conflict, where the alien hand opposes the functions of the normal hand, as seen in our patient. Patients may find it amusing or frustrating, developing counter mechanisms like sitting on the hand or tying it to a bedrail [2,3]. In the case report by Gao X et al., the patient with the callosal subtype exhibited involuntary movements in the right hand, opposing the actions of the left hand. Inter-manual conflict emerged as the primary symptomatic concern, mirroring the presentation observed in this case [6].

The posterior variant involves lesions pertaining to the parietal lobe, occipital lobe, and thalamus, leading to sensory deficits without weakness, accompanied by hemi-anaesthesia, hemianopia, and optic ataxia [3,5]. The affected hand may exhibit unusual posturing, such as hyperextended phalangeal joints and involuntary levitation while performing a task [3]. Each variant has distinguishable features, although in some cases, these features may overlap. The precise pathophysiology of AHS remains elusive, but there is a consensus linking the symptoms to the lesions in specific brain regions [3,7,8]. In all variants, patients may experience the resolution of stroke symptoms yet persist in exhibiting alien hand movements.

Currently, no definitive treatment exists for AHS. However, certain studies [3,9,10] indicate modest clinical improvements with trials of clonazepam and botulinum toxin, but with limited evidence. A multi-disciplinary approach involving rehabilitation has demonstrated increased benefits. This approach focuses on educating patients and teaching them coping strategies [11].

These strategies help patients understand their condition, remember movement sequences, and gain control over their irritability and frustration associated with involuntary movements. In Bru I et al.'s case report, multi-disciplinary rehabilitation demonstrated better outcomes compared to pharmacological interventions. Methods such as cognitive-behavioural therapy, verbal cues, distracting tasks, and visualisation techniques resulted in improved functional outcomes, along with a reduction in inter-manual conflict [11].

In a case report by Kang Q et al., a patient with the callosal-frontal variant exhibited symptoms of compulsive groping and inter-manual conflict. The gradual reduction of symptoms, particularly in inter-manual conflict, was observed through verbal cues [12]. In this case, the lesion was in the left fronto-parietal lobe and genu of the

corpus callosum, presenting with involuntary grabbing of clothes and jitteriness. However, the patient reported loss of sensation and difficulty performing tasks in his right hand. Nevertheless, his hemianaesthesia was challenging to ascertain whether it stemmed from a prior stroke or the current acute episode. Following rehabilitation with verbal cues resulted in a significant improvement in inter-manual conflict and functional outcomes.

CONCLUSION(S)

The AHS although infrequently encountered in clinical settings, stands out as a unique neurological condition that does not fall under the category of movement disorders. In the presented case, the alien hand exhibited groping of clothes and catching the examiner's hand, coupled with sensory deficits predominantly seen in the posterior variant and had improved outcomes through cognitive-behavioural therapy and verbal cue rehabilitation. This case report highlights an intriguing observation: a patient with an anterior variant of AHS displaying clinical features suggestive of a posterior variant. Evidence of optimal treatment for AHS is limited; however, specific exercises and patient education play a significant role in managing the condition. The key takeaway is that when confronted with unusual extremity movements post-stroke, clinicians should consider AHS as a potential differential diagnosis.

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PLAGIARISM CHECKING METHODS: [\[Jain H et al.\]](#)

- Plagiarism X-checker: Nov 15, 2023
- Manual Googling: Jan 23, 2024
- iThenticate Software: Jan 29, 2024 (6%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 06, 2023**

Date of Peer Review: **Jan 08, 2024**

Date of Acceptance: **Feb 05, 2024**

Date of Publishing: **Mar 01, 2024**