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CASE REPORT

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Alien hand syndrome – a rare presentation of stroke

Kelly Le, Christine Zhang and Lisa Greisman

Internal Medicine Residency Program, Greater Baltimore Medical Center, Towson, US

ABSTRACT

Alien Hand Syndrome (AHS) is characterized by intermittent and involuntary movements of a single limb that is not associated with motor dysfunction. AHS may be the initial presentation of serious underlying pathology, such as stroke localized to the non-dominant parietal lobe, corpus callosum damage, or neurodegenerative disease. AHS occurs at a low prevalence in the general population yet represents significant underlying disease burden, necessitating early identification. In this case report, we present an 88-year-old right-handed male with involuntary movement of his left forearm and hand three hours prior to presentation. His symptom corresponded to findings on MRI of the brain, which showed acute infarcts of the right temporal lobe, right parietal cortex, and right parietal subcortex. Infarction of the right parietal cortex accounted for his AHS. The multifocal nature of the infarcts elevated the index of suspicion for an embolic source.

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Alien hand syndrome; frontal; callosal; posterior variant; stroke

1. Introduction

Alien hand syndrome was first described in 1908 by Dr. Hugo Liepmann when he noticed a patient exhibiting an involuntary grasping reflex of the left hand after experiencing a stroke. However, AHS is not a result of a movement disorder. The condition went through several name changes over the years, initially characterized as 'alien' by Dr. Kurt Goldstein in 1908. In 1972, Drs. Brion and Jedynak first coined the term 'alien hand' after evaluating three patients with callosal tumors who were unable to recognize their own hands.[1] AHS is most often caused by stroke, neurodegenerative illness, midline tumor, aneurysm, or damage to the corpus callosum. AHS can affect the frontal, callosal, and posterior regions. In the frontal variant, patients develop disinhibited groping or grasping. Patients are often aware that the limb belongs to them but have difficulty controlling and suppressing such movements. Patients with the callosal variant develop agnostic dyspraxia, in which one hand does not respond to command, and the opposite hand performs the task. [2] The posterior variant causes impaired motor control and non-recognition of the patient's own limb. [2]

2. Case report

An 88-year-old right-handed man presented to the emergency department complaining of acute onset of weakness, numbness, and intermittent involuntary movement of his left forearm and hand (e.g., involuntarily slapping himself while napping). These symptoms began three hours prior to presentation. Head CT revealed no hemorrhage, ischemic event, or mass. All labs were unremarkable except for a mildly elevated troponin level with normal CK-MB. EKG showed nonspecific ST wave changes, but the patient was asymptomatic. He was admitted for further evaluation and started on empiric aspirin and statin therapy. One day after admission, the patient's hand movements spontaneously improved. Brain MRI at that time revealed three areas of acute infarction: the right temporal lobe, right parietal cortex, and right parietal subcortex. An embolic source was the presumed etiology of the multifocal infarcts. Carotid doppler ultrasound and MRA were unremarkable. TTE showed LVEF of 50-55% and grade I diastolic dysfunction. TEE showed hypermobile septum, a probable PFO, and no thrombus. However, after the procedure the patient developed non-sustained atrial flutter, which was previously undiagnosed. A probable history of atrial flutter with thromboembolism was considered as the likely etiology of his CVA. The patient was discharged home on apixaban, aspirin, metoprolol, and atorvastatin.

Images Left to Right: **parietal cortical, parietal subcortical**, and **temporal lobe**

3. Discussion

Diagnosis for this patient was alien hand syndrome. This syndrome occurs more often when patient is fatigued, stress, or has divided attention between concurrent activities. [2] AHS is divided into 3 variants: frontal, callosal and posterior based on their

CONTACT Kelly Le ktle@gbmc.org 6565 N. Charles Street Suite PPE 203 Towson, MD 21204

presentations and locations of brain impairment. The posterior variant can be seen after infraction in the thalamic, occipital, and parietal region of the brain [2], while callosal variant occurs after callosal injury and frontal variant after supplemental motor area, cingulate region, or corpus callosum injury.

The frontal variant of AHS is theorized to occur when the primary motor cortex controlling hand movement is isolated from the premotor cortex's influence. Therefore, the brain is still able to command some body movements but cannot generate self-control over these movements, leading to disinhibited movements. The posterior variant of AHS is believed to occur when disruptions develop between the parietal and temporal cortices. This theory is supported by regional cerebral blood flow (rCBF) studies using single-photon emitted computed tomography (SPECT) and functional MRI (fMRI). In normal patients, fMRI shows activation of multiple extensive neural networks upon initiation of motor activity, whereas isolated activation of the contralateral primary motor cortex is seen in patients with posterior AHS[3]. In a case study reported by Delrieu et al., seven patients with corticobasal degeneration underwent rCBF. Six of the seven patients demonstrated reduced rCBF in the right parietal region compared with controls, and three patients were diagnosed with AHS[4]. In our patient, involvement of the parietal area along with proprioception deficits correlated most closely with posterior-variant AHS. However, signs and symptoms of the three variants commonly overlap, making it difficult to determine precisely which AHS variant is present.

The prognosis of AHS depends on the underlying cause. The duration of signs and symptoms range from 30 minutes to the entire lifespan. There are no known treatments for AHS. Patients and caregivers should receive disease education and counseling to alleviate emotional stress. Coping strategies include use of visual tactics to distract the affected hand with another object, along with cognitive behavioral therapy and hand mittens.

4. Conclusion

AHS is rarely encountered in clinical practice, making it unlikely to be considered in a differential diagnosis. However, this case illustrates AHS as the initial presentation of multiple cerebrovascular infarctions. There should be a low threshold of clinical suspicion for serious brain disorders in patients presenting with AHS. Awareness of this syndrome may allow for earlier detection and effective treatment of the underlying disease.

Disclosure statement

No potential conflict of interest was reported by the authors.

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