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Case Report

Unilateral neglect or alien hand syndrome? A diagnostic challenge

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الملخص

تعتبر متلازمة اليد الغريبة من مضاعفات السكتة الدماغية النادرة. بالرغم من أن حدوث المتلازمة بعد السكتة الدماغية غير شائع، إلا أنها يمكن أن تسبب أثرا معطلا على نمط الحياة والوظيفة لدى المرضى بعد السكتة الدماغية. ومن الصعب التفريق بين متلازمة اليد الغريبة والجلطة المخية النصف كروية غير السائدة مع أعراض العمى الشقي وإهمال النصف الأيسر، حيث إن المرضى بمتلازمة اليد الغريبة يمكن أيضا أن يكون لديهم إهمال وكذلك أعراض سلوكية، إذا كانت تشمل الفص الجبهي. نحن هنا نصف حالة لرجل عمره ٦٢عاما أُدخل إلى قسم إعادة التأهيل وتم علاجه من الإهمال وتعذر الأداء بعد السكتة الدماغية للشريان الدماغي الأوسط الأيمن. بعد المزيد من التقييم، تم تشخيص المريض بمتلازمة اليد الغريبة. وظيفيا، تم تحسنه واختفت الأعراض بعد أربعة أشهر من السكتة الدماغية. في وصف هذه الحالة، قمنا بإبراز صفات متلازمة اليد الغريبة ونهج علاجها الفريد

الكلمات المفتاحية: متلازمة اليد الغريبة؛ السكتة الدماغية؛ الإهمال؛ تعذر العمل؛ إعادة التأهيل

Abstract

Alien hand syndrome (AHS) is a rare post-stroke complication. Although the occurrence of AHS after stroke is rare, it can have a disabling impact on the lifestyle and career of the patients post stroke. It is difficult to distinguish AHS from the non-dominant hemispheric infarction with symptoms of hemianopia and left hemineglect, as patients with AHS can also have neglect as well as behaviour symptoms if the frontal lobe is involved. We report the case of a 62-year-old gentleman who was admitted to the rehabilitation ward and was treated for neglect and apraxia following right middle cerebral artery stroke. After further re-evaluation, the patient was diagnosed with AHS. Functionally, he improved and the symptoms disappeared four months after the stroke. In this case report, we highlight the features of AHS and provide a unique management approach.

Keywords: Alien hand syndrome; Apraxias; Neglect; Rehabilitation; Stroke

Introduction

The alien hand syndrome (AHS) is a rare phenomenon of cardioembolic stroke and is characterised by involuntary and uncontrollable movement behaviour of the arm, not under the control of the patient's mind.[1] AHS can have a non-life-threatening disabling impact on daily activities and may lead to unintentional self-injury (See Figures 1 and 2).[2]

This psychomotor disorder is characterised by the patient's perception of alienation with the involuntary, but seemingly purposeful, movement of the affected limb.[1] It was

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originally proposed by Brion and Jedynac, who described it as two distinctive sensory phenomena characterised by the denial of ownership of one hand despite no sensory loss when it was held by the other hand, secondary to lesions of the parietal lobe and posterior part of the corpus callosum.[2]

The rehabilitation approach of this disorder is adapted to the unique needs of the patient, aimed at increasing the involvement of the patient in everyday life tasks and involves a multidisciplinary approach.

Case description

A 62-year-old man with a history of hypertension, diabetes, and dyslipidaemia was admitted for an elective coronary artery bypass grafting (CABG) after he was diagnosed with three-vessel disease (3VD) six months prior to myocardial infarction. Post operatively, he experienced the complication of right sided middle cerebral artery (MCA) infarction, where he was noticed to be dysarthric, unable to swallow and to follow orders, and also unable to raise his left arm. A Computer Tomographic (CT) brain scan showed acute ill-defined hypodensity of the right frontal, right corona radiata, head of right caudate nucleus, and right pons. Four days after the first stroke diagnosis, he became delirious and had a right-sided gaze preference, was unable to talk, and experienced weakness on the left side of the body. However, there was no evidence of any further evolution of stroke or haemorrhagic transformation in the repeated CT brain.

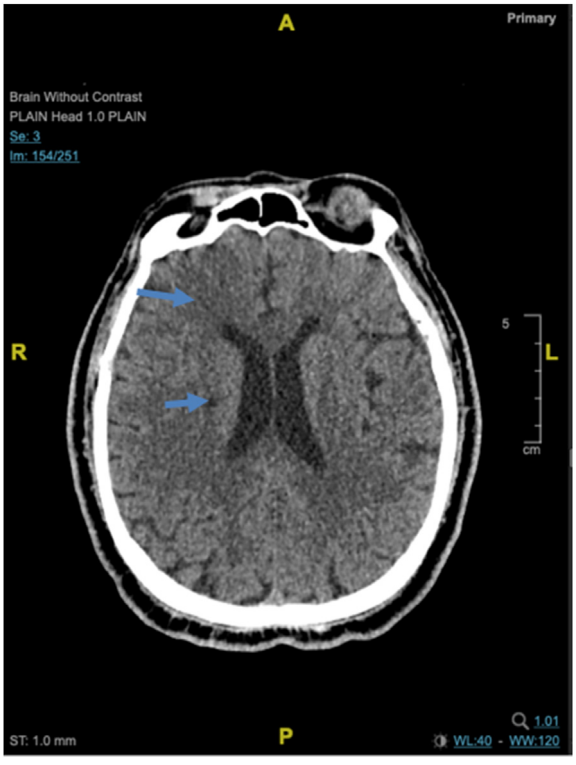

Figure 1: Initial Day 1 Brain CT Scan showing acute ill-defined hypodensity (blue arrows) of right frontal and right corona radiata.

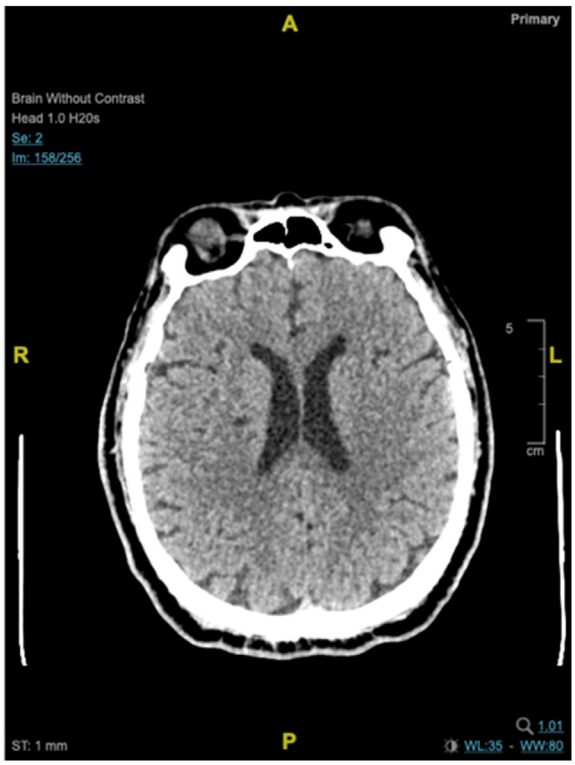

Figure 2: A repeat brain CT at Day 4 post trauma, showing no evidence of any new evolution of stroke or haemorrhagic transformation.

During the initial assessment he had multiple impairments including severe cognitive state as indicated by a Mini Mental State Examination (MMSE) score of 10/30 and was unable to execute tasks despite no lack of ability to obey two-step commands. He had visual spatial sensory and motor neglect, and dysphasia, where he had difficulties identifying and finding term. In term of activities of daily living, he was severely reliant on the caregiver, with the initial Modified Barthel Index (MBI) score of 10/100 (total dependency) and the National Institute of Health Stroke Scale (NIHSS) impairment score of 15.

He underwent intensive rehabilitation therapy daily with the physiatrists, physiotherapists, occupational therapists and a psychologist, aimed at improving his cognitive functions and hemineglect. He was taught cognitive therapy techniques, and underwent learning task therapies and mirror box therapy to improve his condition.

Two weeks after the stroke, he demonstrated functional progress where he was able to take oral feeding after intensive dysphagia management by the rehabilitation team. He was still having difficulty identifying objects, and had a depressed mood. At discharge, his MBI score was significantly improved from 10/100 (total dependency) to 55/100 (severe dependency).

Two months post stroke and bypass surgery, upon reviewing him in his first outpatient visit at the clinic, his hemineglect was found to be improved and the assessments of neglect were normal (Table 1). He was able to walk unaided with a normal gait and had good left hand dexterity with no spasticity.

Table 1: Summary of hemineglect assessments.

	Initial Assessment (1 week post stroke)	Second assessment (2 months post stroke)
1) Line bisection test (normal value < 6 mm mean deviations)	20 mm mean deviations (abnormal result)	3 mm mean deviations (within normal range)
2) Star Cancellation test (normal value > 44/54)	23/54 (abnormal result)	52/54 (abnormal result)
3) Behaviour Inattention Test		
i) Figure and shape copying	- Major omission and gross distortion from the given image.	- Minor omission and no gross distortion from the given image.
ii) Address and sentence copying	- Failure to complete the address given with more than 30 letters omitted.	- Able to complete the address given with no mistake.

Despite these positive outcomes, he started to complain of having weird movement of his unaffected right hand, that affected his bimanual activities. He was aware of the movement but was unable to control it. He described the weird movement as "the right hand tries to counteract all movements or activities I perform with my left hand". This was very distressing to him as it affected his daily activities. These movements had not been manifested earlier during the acute and subacute stages of stroke. He denied any similar presentation previously or any similar history in his family. Further observations during his therapy sessions showed similar unintended movements, and he had to keep telling himself to control his right hand. The managing team's tactic was to keep reminding him of the unwanted movements. His MBI was 90/100 (mild dependency) at this time. The diagnosis of AHS was made on the basis of these symptoms and his previous brain CT finding. He was prescribed oral amantadine 100 mg OD to overcome the AHS that disturbed his activities.

After taking the oral amantadine for two months, he reported that the unwanted involuntary movements had disappeared and he was able to carry out his daily activities uninterrupted. At present, four months post stroke, he is independent in all ADL, with MBI of 100/100 (independent), is able to drive and has returned to work successfully.

Discussion

The best classification for AHS was presented in the paper by Hassan and Joseph, who divided AHS into anterior and posterior variants.[3] The anterior variant was further subdivided into frontal variant (which affects the dominant hand and exhibits impulsive groping and difficulty releasing objects, along with the frontal lobe signs) and the callosal variant (which affects the non-dominant hand and presents with callosal disconnection syndrome including apraxia, neglect, alexia, agraphia and anomia). The posterior variant affects the non-dominant hand and is associated with the sensation that the affected hand does not belong to the rest of the body. In the present case, the patient was perfectly matched to the callosal variant of AHS.

Another possible aetiology for AHS includes ischaemic stroke arising from anterior cerebral artery infarction, in which the patient mostly presents with frontal type AHS.[4] Giroud et al. found that two out of eight patients with callosal infarction had AHS and both the patients had the characteristic of frontal alien hand syndrome.[5] Our patient also had the characteristic pattern of callosum AHS. The symptoms appeared in the non-dominant upper limbs and there was no obvious motor deficit.

Most of the symptoms in AHS reported in previous case reports were apparent at the earlier stage of stroke. However, our patient manifested more prominent symptoms during his outpatient clinic follow-up, mainly due to the unestablished AHS during acute and subacute phases of rehabilitation where it was treated as visuospatial neglect.[6] In addition, with the superimpose adjustment disorder at that time, distinguishing AHS and neglect was difficult. The recognition of AHS could have been missed as the patient was discharged early. Levine, Rinn et al. argued that left sided AHS can be seen as a syndrome of the non-dominant posterior cerebral artery territory infarction with symptoms of hemianopia, left hemineglect, and primary sensory loss in the absence of motor neglect or hemiparesis which is later considered a "sensory" or "posterior" form of AHS.[7] Our patient may also be regarded as having non-dominant hemisphere infarction syndrome with similar symptoms.

In managing a patient with AHS, a multidisciplinary approach along with a pharmacological approach is vital. Despite the lack of evidence of pharmacological intervention in the treatment of AHS, amantadine has been documented to be used in frontal AHS therapeutic management.[8] Besides medication, environmental modifications have also been suggested to reduce the risk of injury. Generally, the rehabilitation approach is geared to the individual needs of the patient and targeted at the improvement of the patient's participation in activities of daily living.

Conclusion

Although AHS is a rare stroke syndrome, the knowledge of its clinical presentation is crucial, since early diagnosis of this syndrome can minimise the debilitating effect on the patients, enabling them to be more independent. The role of a multidisciplinary team in managing this condition is also vital as it helps ensure that this disability does not limit the everyday routines of the patient.

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Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

The authors certify that all required patient consent forms have been received. The patient has given his permission for relevant information to be published in the journal. His identity will be hidden, however the anonymity cannot be guaranteed.

Authors' contributions

WYWR collected necessary case information and wrote the original draft. MHH conceptualized the case report and its aims and reviewed the initial draft and final draft. SA and NK provided a critical review and edited the case. IAH did the visualisation. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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