Global aphasia without hemiparesis: A case series

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Abstract

**Background:** Global aphasia without hemiparesis (GAWH) is a rare stroke syndrome characterized by the unusual dissociation of motor and language functions. Issues regarding its etico-pathogenesis, lesion sites, and recovery patterns are extensively debated in contemporary neuroscience literature. **Materials and Methods:** Four patients admitted in our hospital between 2005 and 2009 with GAWH caused by ischemic stroke were studied retrospectively with emphasis on number and site of lesions, etiology, and recovery patterns. **Results:** The clinical findings from our subjects showed that GAWH could result from either single/multiple lesions including subcortical lesions. The recovery was rapid, although not complete. One case evolved into Wernicke’s aphasia as seen in earlier studies. Two subjects revealed evolution to transcortical sensory aphasia and one to Broca’s aphasia which is distinct from previous proposals. Two cases showed lack of clinico-anatomic correlation during recovery. **Conclusions:** GAWH could result from both embolic and large vessel strokes and single or multiple lesions. The recovery pattern may be variable and may show lack of clinico-anatomical correlation indicating anomalous cerebral functional reorganization, questioning the conventional teaching of language representation in the brain.

Key Words

Global aphasia without hemiparesis, language, motor function, stroke

Introduction

Global aphasia without hemiparesis (GAWH) is a rare and distinct stroke syndrome involving receptive and expressive language impairment, without the hemiparesis typically manifested by patients with global aphasia after large left perisylvian lesions.\(^1\) Global aphasia occurs after large perisylvian lesions in the territory of left middle cerebral artery (MCA) and is associated with hemiparesis on the contralateral side due to the proximity of the language and motor control areas in the cortex. The limited studies in the past have generated controversies on issues such as the etiology, sites of lesion, and recovery patterns in GAWH.

Site(s) of lesion and etiology in GAWH

Two earlier studies by Van Horn and Hawes \(\text{et al.}\)\(^2\) and Tranel \text{et al.}\(^3\) reported three patients each with GAWH resulting from two discrete lesions in the left hemisphere and suggested it as an indicator of embolic encephalopathy. Legatt \text{et al.}\(^4\) reported two patients with non-embolic etiology among six patients and Hanlon \text{et al.}\(^1\) reported two subjects with non-embolic etiology among ten patients. Deleval \text{et al.}\(^5\) reported two patients with GAWH caused by a single discrete infarct in the posterior part of F2 and F3. In another study by Pundit \text{et al.}\(^6\) among three cases, two had multiple and one had a single anterior perisylvian lesion.

Keyserlingk \text{et al.}\(^7\) reported that global aphasia with hemiparesis was caused by lesions extending to the wall of the lateral ventricle, but lesions in GAWH spared the deep white matter. Bang \text{et al.}\(^8\) using DWI (Diffusion weighted imaging) and SPECT for lesion analysis, sub grouped their patients into (a) classical lesion group (combined inferior frontal and superior temporal gyri); (b) single lesion group (isolated inferior frontal or superior temporal gyrus); and (c) extrasylvian group (subcortical or parieto-occipital).

Language recovery in GAWH

Early studies on GAWH have shown faster and near-total recovery in GAWH, which has been attributed to the circumscribed lesion involving a small portion of the perisylvian region. Nagaratnam \text{et al.}\(^9\) noted that recovery was better in subjects with initial mild language impairment and a single either anterior or posterior lesion. Hanlon \text{et al.}\(^1\) based on language recovery in ten patients with GAWH, proposed the following three distinct subtypes: (a) persistent GAWH – with
dense global aphasia that failed to show any improvements in language functions, (b) GAWH-TCM – where the initial global aphasia recovered to transcortical motor aphasia, and (c) GAWH-Wernicke’s – with recovery to Wernicke’s aphasia.

Materials and Methods

Of 1230 cases of stroke during the period 2005-2009, four cases of GAWH caused by ischemic stroke were included [Table 1]. Inclusion criteria were normal scores for motor power on NIH stroke scale[10] and global aphasia as confirmed by Western aphasia battery (WAB).[11] At admission, all were alert and globally aphasic and underwent CT and/or MR imaging study. MR/CT angiogram was done in patients with suspected large vessel disease. The subtype of ischemic stroke was determined as per TOAST criteria.[12] Aphasia subtypes were confirmed after detailed language evaluation by a Speech-Language Pathologist using the Malayalam and Kannada versions of WAB.[13] The WAB aphasia quotient and subscales of fluency, comprehension, repetition, and naming were used to establish the subtypes of aphasia [Table 2].

Case 1

A 67-year-old, literate, Malayalam-speaking, right-handed male, with no premorbid illness, presented with abrupt onset of inability to express and comprehend speech, without any limb weakness. Neurological examination revealed decreased linguistic fluency and verbal output, impaired comprehension, repetition, and naming. There was no facial paresis or limb weakness. CT scan done at 48 hours showed a left MCA territory infarct located in the anterior frontal region, which was confirmed by MRI brain [Figure 1]. MR angiogram was normal. Echo showed sclerotic aortic valve and trivial mitral regurgitation. Carotid–vertebral Doppler was normal and Holter analysis showed multiple atrial ectopics without any atrial fibrillation. As per the TOAST criteria, he was classified as possible cardioembolic stroke. The initial language assessment was suggestive of global aphasia. The second evaluation (two weeks later) revealed recovery to Wernicke’s Aphasia. The third evaluation, six months later, showed improvement in all domains of language functions, except comprehension, leading to the diagnosis of transcortical sensory aphasia.

Case 2

A 36-year-old, right-handed, Malayalam-speaking, literate gentleman, who was a chronic smoker and alcoholic, abruptly developed inability to speak and impaired comprehension. No motor weakness or facial paresis was noted. He was found to have polycythemia (Hemoglobin -19.3 G%, Packed cell volume -57.4) and dyslipidemia (HDL cholesterol - 21 mg %). The CT scan showed infarcts in the caudate and temporoparietal regions of the left hemisphere [Figure 2]. CT angiogram and carotid vertebral Doppler revealed total occlusion of left internal carotid artery(ICA) suggestive of a large artery stroke as per TOAST classification. Language assessment revealed a global aphasia. A follow-up examination, four weeks later, showed marked improvement in comprehension, indicating the recovery to Broca’s aphasia.

Table 2: Western aphasia battery at admission and follow-up

| Subjects | Case 1 | Case 2 | Case 3 | Case 4 |
|----------|--------|--------|--------|--------|
| Assessment | I | II | III | I | II | I | II | I | II |
| Fluency | 0 | 5 | 9 | 0 | 2 | 1 | 9 | 0 | 5 |
| Comprehension | 0.4 | 5.7 | 0.03 | 4.3 | 2.6 | 6.5 | 0.5 | 2.1 |
| Repetition | 0 | 6.2 | 8.4 | 0 | 2.4 | 1.4 | 9.8 | 0 | 3 |
| Naming | 0 | 1.2 | 8.3 | 0 | 0.3 | 0 | 4.9 | 0 | 1.5 |
| Diagnosis | GA | WA | TSA | GA | BA | GA | TSA | GA | WA |

GA = Global aphasia; WA = Wernicke’s aphasia; TSA = Transcortical sensory aphasia; BA = Broca’s aphasia

All patients were right handed. MRA = MR angiogram; TTE = Transthoracic echo; EF = Ejection fraction
Case 4
A 50-year-old, right-handed male, premorbidity diagnosed to have hypertension and ischemic heart disease, was admitted two years earlier with left hemiparesis and normal language function due to a right MCA perisylvian infarct. Echo showed left ventricular systolic dysfunction and dense spontaneous LV contrast. He was treated as an embolic stroke and had gradually improved over three months to Grade 4 power on the left side. He was again admitted with sudden onset difficulty to speak and understand after he had been noncompliant with medications. The neurological examination showed a significant aphasia with no facial or limb weakness on the right side and Grade 4 power on the left side due to the old stroke. The CT scan performed 24 hours post-stroke showed an old perisylvian infarct in the right hemisphere and a new infarct in the left posterior temporoparietal region [Figure 4]. MRI could not be done due to poor patient cooperation. Echo revealed LV systolic dysfunction and dilated cardiomyopathy. Carotid Doppler was normal. He was classified under cardioembolic stroke as per the TOAST criteria. WAB scores showed global aphasia which evolved into a Wernicke’s aphasia on a follow-up examination four weeks later.

Discussion
Among four patients in our series, two had a cardioembolic source, one was due to internal carotid artery occlusion and other was of undetermined cause conforming to earlier studies that GAWH can result from embolic as well as large vessel strokes. Previous studies have reported that both single and multiple lesions cause GAWH. Specifically, two of our subjects (Cases 1 and 4) showed single lesion and the remaining two showed multiple lesions.

Subcortical lesions causing GAWH have been reported by Van Horn and Hawes and in two patients by Bang et al. Bang et al. suggested that the spared motor skill was due to the functional reorganization to the ipsilateral (i.e., right) cortex subsequent to an earlier lesion of the motor pathway. Our subject with subcortical GAWH did not reveal any history indicative of previous stroke or transient ischemic attack and did not show evidence for ipsilateral functional reorganization.

Three subjects showed marked improvements in their language skills by four weeks, supporting the earlier reports of rapid recovery. Additionally, two subjects in our cohort (Cases 1 and 4) showed a pattern of recovery (GAWH-Wernicke’s)
proposed by Hanlon et al. However, in one of them (Case 1), a reassessment of language functions at six months revealed subsequent recovery to transcortical sensory aphasia, a pattern distinct from Hanlon's proposal. Case 2 showed aphasia recovery to Broca's aphasia and Case 3 recovered to transcortical sensory aphasia which is distinct from earlier reports. These observations ascertain that the recovery pattern may be highly variable in subjects with GAWH.

A novel observation from two of our subjects was the lack of anatomo-clinical correlation during the recovery period. Case 1 exhibited a recovery to Wernicke's aphasia immediately post-stroke and later to transcortical sensory aphasia (6 months post-onset), subsequent to an infarct in the left frontoparietal area. Similarly, Case 2 showed recovery to Broca's aphasia at fourth week post-onset despite the predominant lesion of the posterior linguistic cortices. Lack of such anatomo-clinical correlation maybe considered as an index of complex and anomalous functional organization of linguistic skills in subjects with GAWH.

Limitations

The small number of subjects in the study appears to be a serious limitation, but it merits attention considering the rarity of the syndrome. Second, the current observations are from the acute stage of stroke. However, it has been reported that certain vital clinical observations can be made only in the acute stage, especially when the signs and symptoms are transient and in the process of recovery. Third, none of our subjects were available for a second follow-up, except Case 1. Although MRI would have been better for lesion localization, it could not be done in case 2 and 4 due to poor patient cooperation. Functional imaging could not be employed in the present study due to no availability. In view of the limited numbers, it is difficult to establish conclusively the causal etiological relationship, for example, in case 2, a total internal carotid occlusion does not rule out a distal embolism.

Conclusions

The findings from our study showed that GAWH could result both from single and multiple lesions as well as embolic and large vessel strokes. It also provided evidence for faster recovery of language functions in these patients. However, differing from earlier studies, our findings showed that the recovery pattern in GAWH could be variable among the patients and that the ipsilateral hemispheric control of motor functions may not be the cause of spared motor functions in these patients. The lack of anatomo-clinical correlation during the recovery period is suggestive of the complex cerebral functional reorganization occurring in some patients with GAWH.

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