LETTER TO THE EDITOR

Aphemia: an isolated disorder of speech associated with an ischemic lesion of the left precentral gyrus

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Sirs,

Aphemia, also termed apraxia of speech, cortical anarthria or pure word mutism, is an isolated disorder of coordinated speech articulation that results in severe affection of verbal motor output [8, 9]. This rare and possibly underdiagnosed syndrome was first postulated by Paul Broca in 1861 [1]; however, the definition remained imprecise during the course of history. Especially the differentiation between aphemia and aphasia—the latter of which was shortly after described by Armand Trousseau—as well as dysarthria has been in discussion since then.

A 61-year-old right-handed dentist with treated hypertension awoke with acute loss of speech and saliva running out of the right corner of his mouth. He wrote "stroke" on a piece of paper for his wife, who called the ambulance. In the emergency room, he was nearly mute due to grossly distorted motor output, but he had full comprehension of speech, could communicate through gestures and intact, fluent writing (Fig. 1a). Speech was very difficult with strangled vowels, severe phonematic paraphasia and abnormal prosody that was not facilitated by singing, reading or repetition. This and the constant effort to correct himself cumulated in visible frustration, whereas, surprisingly, the patient was able to produce-apparently emotionally triggered—short, but intact and fluently articulated commentaries of the situation (e.g., "It's not working!", "That's terrible!"). There was a mild right facial palsy without further affection of coordinated buccofacial movement (whistling, etc.) or oropharyngeal sensibility (NIHSS 4). Diffusion-weighted MRI performed shortly after presentation to the emergency room showed an acute ischemic lesion of the left precentral gyrus that was not yet visible on T2-weighted FLAIR images. The lesion extended slightly to the medial part of the premotor cortex (Fig. 2). Ultrasound and MRA demonstrated a highgrade stenosis of the left internal carotid artery (ICA). As extensive stroke workup failed to show an alternative source of embolism, symptomatic ICA stenosis was considered the most likely cause of stroke, and the patient was treated with endarterectomy on day 4. Under speech therapy, the symptoms improved gradually, and speech became more and more fluent after day 3. In the beginning, syntax presented incompletely because of the effort of speech, but became intact by day 3. From day 1, repeated dictations and spontaneous essays (see Fig. 1b) could demonstrate the constantly fluent and intact written language. During observation, dysarthric components never occurred. At 1 week, the patient recovered completely, including the right facial palsy.

Still not clearly classified as an articulatory or language disorder, aphemia is today understood as an isolated disorder of the planning of motor articulation of speech. These patients, even when mute, can write correctly and have no difficulty in the production of verbal sequences as long as they do not have to articulate them, distinguishing the disorder from motor aphasia. The presentation of abnormal prosody, "false starts," self corrections and the occurrence of undisturbed "insulas" in speech can help to differentiate it from dysarthria. Often, right-sided hemiparesis, limb apraxia, mild buccofacial apraxia and central right facial palsy are associated symptoms. The lesion of the precentral gyrus corresponds to recent data of the few similar cases published [2, 5, 7]. These case studies have linked the

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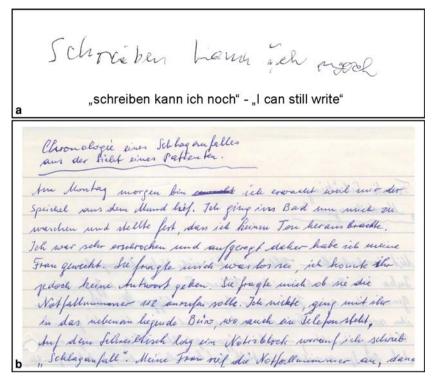
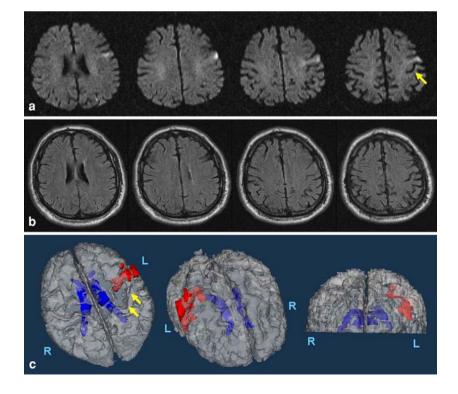


Fig. 1 Examples of intact written language: in the emergency room, the nearly mute patient wrote "I can still write" on a piece of paper that he held in the palm of his hand—which explains the somewhat unsteady handwriting (a). On day 2, speech being still grossly distorted and verbal communication not possible, the patient could give a full written report of the events entitled "Chronology of stroke—from a patient's point of view" (b). "Monday morning I awoke because saliva was running out of the corner of my mouth. I

went to the bathroom to wash myself and noticed that I could not make a sound. I was very frightened and nervous, and so I woke my wife. She asked what the matter with me was, but I was not able to give an answer. She asked if she should call the 112—the emergency number. I nodded and went with her to the office next door where the telephone is. There was a notepad on the desk on which I wrote down the word 'stroke.' My wife called the emergency number..."

Fig. 2 Diffusion-weighted MRI (a) shows a circumscribed hyperintense acute ischemic lesion in the left precentral gyrus, not yet visible on T2-weighted FLAIR images (b). Yellow arrows mark the central sulcus. Three-dimensional visualization of lesion causing aphemia attained through segmentation, reformation and registration of two-dimensional data sets: transparent brain tissue, ventricles in blue, lesion in red (c)





control of articulation to a cortical region adjacent to the face motor cortex. Similarly, functional imaging studies have shown representational sites of tongue, lip and articulation movements located in the pre- (M1) and postcentral (S1) gyrus without significant activation in Broca's and Wernicke's areas [4]. Lesions of the premotor cortex [6], the insula and Broca's area [3] in the dominant hemisphere have also been observed causing aphemia-like syndromes. This widespread lesion pattern could suggest an underlying network involving motor cortex, Broca's area and supplementary motor areas engineering the articulation of speech.

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