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Case report

A role for the cerebellum in motor speech planning: Evidence from foreign accent syndrome

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Abstract

A 3 year follow-up study was performed in a patient with foreign accent syndrome (FAS) as the sole cognitive manifestation of a left fronto-parietal stroke. The hypothesis of involvement of the right cerebellum in this motor speech planning disorder was investigated by means of functional neuroimaging (SPECT) and neurobehavioral assessments. Based on the close parallelism between the evolution of FAS symptoms and the perfusional changes in the right cerebellum, it is argued that FAS may result from a disruption of a close functional interplay between the supra- and infratentorial speech centers involved in motor speech planning. © 2005 Elsevier B.V. All rights reserved.

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Foreign accent syndrome [1] (FAS) is a rare motor speech disorder, which results in articulatory distortions that are perceived by native-speakers as a foreign accent. Since the first description [2], the condition has been reported in only 31 cases. The causative lesion mostly involved the prerolandic motor cortex (BA 4), the frontal motor association cortex (BA 6 or 44) or the striatum of the language dominant hemisphere [3].

Clinically as well as anatomically, FAS resembles apraxia of speech (AoS), which is defined as a selective impairment of speech movements following the inability to properly and smoothly convert phonological knowledge into verbal–motor commands [4]. As a result, it has been argued that FAS is a subtype of AoS [5]. Dronkers [6] localized the crucial anatomical seat of AoS in the language dominant insula, directly anterior to the central insular sulcus. Earlier terminology, such as 'ataxic aphasia' and 'cortical dysarthria' [7] suggests that AoS shares some semiological similarities with ataxic dysarthria. Indeed, motor speech symptoms resulting from cerebellar pathology may also be slow, monotonous, staccato, scanned and indistinct. Since these similarities reflect a distorted process of articulatory planning and coordination, FAS, AoS and ataxic dysarthria may have some pathophysiological mechanisms in common. Given the recently acknowledged role of the lateralized linguistic cerebellum in a variety of speech and language processes and the close anatomical connections between the right cerebellum and the motor speech centers of the language dominant hemisphere, we hypothesized that in addition to the perisylvian speech regions, the right cerebellum may be crucially involved in motor speech planning disorders. To test this hypothesis we examined a patient with FAS by means of functional neuroimaging and close neurobehavioral follow-up.

1. Case report

A 53-year-old right-handed native-speaker of Dutch with 14 years of education acutely developed severe speech

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difficulties and weakness of the right hemicorpus. Neurological examination on admission revealed total speechlessness with intact auditory-verbal and written comprehension, normal spelling and normal oral praxis. A moderate right hemiparesis and central facial nerve palsy were found as well. Tendon reflexes were brisker at the right than on the left side of the body. The right plantar response was extensor. Sensory examination was normal. On confrontation, no visual field defects or spatial neglect phenomena were noticed. Medical history was unremarkable. The day after admission speech started to recover and an oral-verbal output syndrome became apparent in which speech was rather slowly and hesitantly articulated. In addition, vowel and consonant lengthenings and a slight flattening of voice volume disrupted speech. Syllables tended to become isochronous and of equal prominence (scanning speech). Although the articulatory distortions occurred rather inconsistently, most errors were close phonemic target substitutions. Articulatory groping and struggling leading to sequential errors and mostly unsuccessful efforts to self-correction alternated at times with 'islands of error-free speech'. Errors significantly increased with word length and were not task dependent. No improvement was found when the patient recited, repeated or read aloud. Apart from the articulatory deviations which resulted in a deficient speaking rate (68.4 words/min), analysis of 26 spontaneous speech utterances recorded in a short conversation (2 min) did not disclose any linguistic deficits. CT scan of the brain on the fourth day of admission revealed an infarction in the left fronto-parietal region. Two weeks post-stroke, MRI displayed a cortical ischemic lesion with discrete hemorrhagic conversion in the territory of the left medial cerebral artery, involving the inferior frontal gyrus (BA 44 and 45), the precentral gyrus (BA 4), the anterior insular cortex (BA 52), the postcentral gyrus (BA 1, 2 and 3) and the supramarginal gyrus (BA 40) (Fig. 1). Aphasic and dysarthric disturbances were formally excluded 3 weeks



Fig. 1. Brain MRI coronal T1-weighted slices (A–F) display the fronto-parietal lesion involving the inferior frontal gyrus, the precentral gyrus, the anterior insular cortex, the postcentral gyrus and the supramarginal gyrus of the left hemisphere.

after onset neurological symptoms by means of standardised language tests including the Dutch version of the Aachener Aphasie Test [8], the Boston Diagnostic Aphasia Examination [9], the Token Test [10], the Boston Naming Test [11,12], a phonological and semantic verbal fluency task, subtests of the Dutch version of the Psycholinguistic Assessments of Language Processing in Aphasia [13] and Frenchay Dysarthria Assessment [14]. However, the patient herself as well as her direct acquaintances and hospital staff noticed that her speech sounded odd as if she spoke with a French, German or even Russian accent. This was striking since she did not master these languages. A neurolinguistic diagnosis of FAS was made and intensive speech therapy was started. Extensive neuropsychological investigations were carried out in the postacute phase (between 3 weeks and 4 months post-onset) by means of the Mini Mental State Examination [15], the Progressive Matrices [16], the Wechsler Adult Intelligence Scale [17], subtests of the Hierarchic Dementia Scale [18], the Wechsler Memory Scale-Revised [19], the Rey–Osterrieth figure [20], the Wisconsin Card Sorting Test [21], the Stroop Colour–Word test [22], the Right–Left Orientation test, the Visual Form Discrimination test and the Judgment of Line Orientation test [23]. On these tests, the patient obtained above average results. After a rehabilitation period of 1 year, FAS as well as a residual right spastic hemiparesis persisted. Three years after the stroke FAS had completely remitted and only very mild AoS symptoms were found. The patient spoke slightly hesitantly and consonant clusters were sometimes articulated with increased effort. Scanning of speech had disappeared but articulatory groping still sporadically occurred.

Tc-99m-ECD SPECT perfusion scans were acquired using a Trionix (OH, USA) Triad three-detector gamma camera equipped with high resolution fan-beam collimators. The



Fig. 2. Tc-99m-ECD SPECT scan of the brain in the acute phase of the stroke displays a severe hypoperfusion in the left frontal motor and parietal cortex as well as a secondary relative hypoperfusion in the thalamus, striatum and anterior temporal region of the left hemisphere (upper row). A relative hypoperfusion of the right hemicerebellum, reflected by contralateral cerebellar uptake reduction of 15% (crossed cerebellar diaschisis) is shown as well (lower row). Visually this activity reduction corresponds to 2–3 colour-steps on the image.



Fig. 3. Repeat SPECT in the late phase of the stroke shows normalisation of the tracer uptake in the right cerebellar hemisphere (lower row). Only a very mild perfusion improvement is observed in the frontal motor area, parietal cortex, thalamus and striatum of the left hemisphere (upper row).

projection data were reconstructed by filtered backprojection using a Butterworth filter (with cut-off frequency of 0.7 cyc/cm and rol-off 5) resulting in trans-axial images with a pixel size of 3.56 mm. The color-scale was subdivided in 16 colors, represented each 6.25% of the maximum activity. Baseline Tc-99m-ECD SPECT scan of the brain performed 33 days post-onset neurological symptoms showed a breach of the left frontal motor cortex. A relative hypoperfusion was found in the thalamus, striatum and anterior temporal region of the left hemisphere. In addition, a relative hypoperfusion of the contralateral right cerebellum was found as reflected by a 15% reduction of tracer uptake (Fig. 2). After clinical remission of FAS (3 years post-onset neurological symptoms) repeat Tc-99m-ECD SPECT showed normalisation of tracer uptake in the cerebellum whereas only mild perfusion improvement was observed in the supratentorial brain regions (Fig. 3). Due to technical reasons the same color scheme could not be used when the second scan was performed.

2. Discussion

Following a short period of total speechlessness in the early acute phase, this patient developed FAS after vascular ischemic damage in the anatomoclinically suspected frontoparietal region of the language dominant hemisphere. Some semiological and anatomoclinical findings deserve further consideration.

Firstly, although FAS is traditionally regarded as a distinct neurolinguistic entity, research has neither been able to identify a coherent system in the speech errors nor to separate it unambiguously from AoS. This has led to the conviction that FAS is a subtype of AoS [5]. Our findings corroborate this view as residual speech symptoms coped with a diagnosis of mild AoS when FAS had resolved in the late phase of the stroke. Secondly, AoS and FAS are remarkably similar to ataxic speech disturbances consequent to lesions involving the superior paravermal region of the cerebellum [7,24]. A slow and irregular articulation, a monotonous, staccato and

scanned oral-verbal output as well as inconsistent misarticulations and phonetic alterations of vowel and consonant production are commonly found in AoS, FAS and ataxic dysarthria. In addition, it has been demonstrated in experimental studies [25] that voice onset time (the time interval between consonant burst and vowel onset) and production and discrimination of vowel length is affected in FAS, AoS and ataxia of speech. These similarities might suggest some shared pathophysiological mechanisms underlying disrupted articulatory planning and deficient speech timing processes. In addition to the long-standing belief that FAS and AoS are produced by lesions of the anterior perisylvian speech areas, functional neuroimaging data obtained in our patient suggest that the cerebellum may also be crucially involved in motor speech planning disorders. The SPECT follow-up study revealed a close parallelism between the remission of the right cerebellar hypoperfusion and clinical recovery of FAS symptoms 3 years after the stroke. In agreement with the recently acknowledged role of the linguistic cerebellum in a variety of non-motor linguistic processes, this observation might indicate that FAS and AoS may result from disruption of a close functional connection between the supra- and infratentorial motor speech centers as reflected by diaschisis. In addition to a substantial amount of clinical and experimental evidence in support of a functionally lateralized linguistic cerebellum [26], the present case at least supports the view that the cerebellum may be involved in motor speech planning disorders, such as AoS and FAS. The hypothesis for a role of crossed cerebellar diaschisis in the pathogenesis of motor speech planning disorders associated with a left lower frontal area lesion merits further prospective research on a larger series of patients. Our study further indicates that functional neuroimaging might be of crucial importance to further explore the functional role of the cerebellum in cognition.

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