CORTEX 45 (2009) 870–878

available at www.sciencedirect.com

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Research report

Foreign accent syndrome as a developmental motor speech disorder

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ARTICLE INFO

Article history:
Received 9 April 2008
Reviewed 14 July 2008
Revised 14 August 2008
Accepted 31 October 2008
Action editor Mike Anderson
Published online 21 November 2008

Keywords:
Foreign Accent Syndrome
Specific language impairment
Apraxia of speech
Language
Motor speech
Cognition

ABSTRACT

Introduction: Foreign Accent Syndrome (FAS) is a relatively rare motor speech disorder in which the pronunciation of a patient is perceived by listeners of the same language community as distinctly foreign. FAS has been well documented in adult patients with etiologically heterogeneous, though mostly vascular brain lesions affecting the motor speech network of the language dominant hemisphere. In addition, reports exist of adult patients in whom FAS was due to a psychiatric illness. Although FAS has been reported in children, such accounts are rare and have remained largely anecdotal in that there have been no formally documented cases of FAS as a developmental motor speech disorder.
Methods and results: For the first time, we describe the clinical, cognitive and neurolinguistic findings in two patients who in the absence of a history of psychiatric illness or acquired brain damage already presented with FAS at an early stage of speech and language development. In the first patient ''developmental FAS'' was associated with a dysharmonic distribution of neurocognitive test results indicating slight underdevelopment of visuo-spatial skills and visual memory. The second patient presented with ''developmental FAS'' associated with specific language impairment (SLI). Independent support for a diagnosis of FAS in both patients was obtained in an accent attribution experiment in which groups of native speakers of (Belgian) Dutch assessed the type of foreign accent of a sample of the patients' conversational speech. Both patients were judged as non-native speakers of Dutch by the majority of participants who predominantly identified the accent as French.
Conclusion: This paper for the first time documents two patients who presented with FAS on a developmental basis. The finding that FAS does not only occur in the context of acquired brain damage or psychogenic illness but also exists as developmental motor speech impairment requires a re-definition of FAS as a clinical syndrome.

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doi:10.1016/j.cortex.2008.10.010

1. Introduction

The condition in which listeners of the same language community perceive a motor speech disorder as a foreign speech accent was first described in 1907 by the French neurologist Pierre Marie in a patient who developed a regional accent when recovering from anarthria following a subcortical left hemisphere stroke (Marie, 1907). Since then, more than 60 adult patients with a wide variety of, mostly vascular, etiologies have been described who presented FAS either in isolation or, more commonly, in association with other speech and language disorders such as aphasia, apraxia of speech (AoS) or dysarthria. In the majority of patients, FAS resulted from lesions in the perisylvian speech regions involving the prerolandic motor cortex (BA 4), the frontal motor association cortex (BA 6 or 44) or the striatum (Dankovicova et al., 2001). Only a few adult patients have been described who developed FAS in the absence of structural brain damage (e.g., Critchley, 1962; Moonis et al., 1996; Coelho and Robb, 2001; Hwang et al., 2001; Reeves and Norton, 2001; Van Borsel et al., 2005; Verhoeven et al., 2005; Laures-Gore et al., 2006; Ryalls and Whiteside, 2006; Poulin et al., 2007; Reeves et al., 2007). In most of these cases FAS resulted from a psychogenic cause (psychogenic FAS).

A full 100 years of multidisciplinary research has neither been able to identify the pathophysiological substrate of this syndrome nor to identify a coherent system in the speech errors that may separate FAS unambiguously from AoS (anarthria, verbal apraxia, speech apraxia) or ataxic dysarthria. It has to be conceded that FAS, AoS and ataxic dysarthria share a slow, monotonous, staccato, scanned, indistinct, remarkably irregular, jerky, explosive, slurred, and laboured verbal output. On the basis of this close semiological resemblance it has been argued that FAS is a sub-type of AoS (Whiteside and Varley, 1998). In addition, a direct link of FAS and AoS with cerebellar speech pathology is also suggested by earlier terminology for FAS – '*ataxia* of the prosody faculty' (Monrad-Krohn, 1947) – as well as for AoS – '*ataxic* aphasia' or 'cortical *dysarthria*' (Whitty, 1964). Since the motor speech symptoms of these conditions basically result from distorted articulatory planning and coordination processes, it has been hypothesized that the cerebellum may also be crucially implicated in the pathophysiology of FAS and AoS (Whitaker, 1982; Cole, 1971; Mariën et al., 2006; Mariën and Verhoeven, 2007).

Although FAS has been well documented in adult patients, reports of FAS as an acquired motor speech disorder following structural brain damage are very rare in a paediatric population and have remained largely anecdotal (Jha, 2007). In addition, there are, to the best of our knowledge, no formal reports in which FAS is described as a developmental motor speech planning disorder. This paper is the first description of the clinical, cognitive and neurolinguistic findings in two patients who did not acquire FAS in a context of structural brain damage or psychiatric illness but who already presented FAS in an early stage of speech and language development. In the first (adult) patient, FAS was noted in association with developmental AoS, while the second (paediatric) patient presented with FAS in association with specific language impairment (SLI).

2. Case reports

2.1. *Case 1 (TL)*

2.1.1. *Clinical history*

TL is a 29-year-old right-handed women who is a native speaker of (Belgian) Dutch (Verhoeven, 2005). She consulted the neurological department because of 'strange pronunciation' that had been characteristic of her speech since early childhood. Apart from the perceptual impression of a foreign accent, developmental milestones were unremarkable. Medical history was not contributive. She was born at term after normal gestation and labour and there had been no perinatal or postnatal problems. Scholarly achievements had always been average and there was no family history of developmental language problems or learning disability. This patient's parents and siblings were monolingual speakers of (Belgian) Dutch. She had an educational level of 14 years and worked as a secretary. At school she had learned French, English and German, but she only seldom used these languages and their use was restricted to professional contexts. After careful heteroanamnestic inquiry, no indications were found for an acute onset of deviant speech features: close relatives reported that the foreign accent had always been characteristic of her speech and had not changed over time. Neurological examination was unremarkable. Brain MRI and an EEG were normal.

2.1.2. *Neuropsychology and neurolinguistics*

Formal assessment of handedness by means of the Edinburgh Inventory (Oldfield, 1971) showed a strong and consistent right hand preference, reflected by a laterality quotient of +100. The Wechsler Adult Intelligence Scale (WAIS-III; Wechsler, 1997) revealed a normal total intelligence quotient (IQ) of 88 with a discrepancy of 16 points between the verbal (VIQ = 97) and performance level (PIQ = 81; −1.2 SD) (Table 1). At the performance level, deviant scaled scores (≥−2 SD) were obtained for 'picture completion' and 'picture arrangement'. The Wechsler Memory Scale–Revised (Wechsler, 1987) was also characterized by a significant discrepancy between the verbal (=100) and visual memory index (=74; −1.7 SD). Low scores were obtained on verbal and visual working memory tasks (digit span). The Rey–Osterrieth figure (Osterrieth, 1944) as well as the copying tasks of the Birmingham Object Recognition Battery (BORB) (Riddoch and Humphreys, 1993) and Hierarchic Dementia Scale (HDS) (Cole and Dastoor, 1987) were normal. The ability to conceptually rearrange pictures was unimpaired (Hooper, 1983). Visual object identification and visual semantics were normal (Riddoch and Humphreys, 1993; Mariën et al., 1998). As demonstrated by the Wisconsin Card Sorting Test (Heaton et al., 1993), frontal planning and problem solving were normal. The ability to inhibit a competing and more automatic response set was also normal (Golden, 1978). On tasks evaluating visual search and sequencing the patient scored in the lower range (Reitan, 1958). Mood and behaviour were normal. No indications were found for mood or behavioural abnormalities during the clinical assessments and careful inquiry of the patient's close relatives confirmed these findings. Examination of bucco-labio-lingual praxis by means of a Dutch