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Rare disease

Transient foreign accent syndrome

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Summary

Foreign accent syndrome (FAS) is a poorly understood and studied syndrome as it is indeed a rare entity. Since its first description in 1907 by French neurologist Pierre Marie involving a patient who presented with an Alsatian accent, there are approximately only 60 cases reported in the literature. The majority of such cases of FAS have been secondary to cerebrovascular accidents. Of the cases in the literature, none report such a transitory nature of FAS. In this particular case, a 55-year-old male presented with a foreign accent. This FAS was triggered by ischemia and was reversed after a seizure, the first reported in the literature.

BACKGROUND

Foreign accent syndrome (FAS) is a rare syndrome in itself, with only 60 cases reported in the literature. Moreover, this is the first case where the FAS was reversed after a seizure. This rare presentation should be included in published literature so it can aid in understanding FAS, which is truly an enigmatic entity.

CASE PRESENTATION

A 55-year-old Texan male, presented with a day history of another accent. His son noted his pronunciation of words as 'foreign'. He denied issues with writing, reading, or grammar and denied any other systemic symptoms including neurological deficits. His medical history included hypertension, obstructive sleep apnoea syndrome, hypertriglyceridemia and cerebrovascular accidents (CVAs): left frontal infarction (2003) and left parieto-occipital infarction (2007). No medications or allergies were significant to this case. He smoked half pack per day for 20 years and denied illicit drug use. Family history is non-contributory.

Physical examination revealed vital signs within normal limits. His mental status via the Montreal Cognitive Assessment was scored as 29/30. There was no right-left disorientation. He followed complex commands and his speech was clear and fluent. Of note, multiple medical personnel noted that the patient sounded as if he had a Cockney accent. The patient did not recognise his accent as foreign. Language showed minor disruptions as alterations of syllable structure. There was no change in tone, rate, or pitch, no sound substitutions, perseveration, or echolalia, no difficulty with speech initiation and there was preservation of syntax. No alexia or agraphia of mono-syllable and multi-syllable words, phrases, or sentences were noted. Additionally, he could copy a paragraph, and there was no evidence of acalculia. No deficits in executive functioning were noted. No other cortical signs were noted. The rest of his neurological examination did not reveal any deficits.

Within minutes of the examination, the patient exhibited right head version followed by a Jacksonian march up his right arm, which was witnessed. This evolved into a tonic-clonic generalised seizure lasting 30 s. Postictal confusion lasted 30 min.

INVESTIGATIONS

There were no abnormalities on laboratories. An initial emergency department CT of his head showed encephalomalacia in the left parietal lobe, precentral gyrus and middle frontal gyrus, without evidence of acute findings.

Following the seizure, further testing performed included a transthoracic echocardiogram and an electroencephalogram that did not reveal new findings. MRI of his brain revealed restricted diffusion in the left parieto-occipital region and in the left middle frontal gyrus (figure 1A,B). No mass lesions and no evidence of pathologic enhancement were seen. Magnetic resonance angiography of his head was normal appearing.

OUTCOME AND FOLLOW-UP

Subsequently, his neurological examination remained similar to the initial examination except that he reverted to his Texan accent. The patient has not been to Britain and does not have relatives/friends there. This was no re-occurrence of his FAS at discharge. He was lost to follow-up.

DISCUSSION

Only 60 cases report FAS since it was first described in 1907 by French neurologist Pierre Marie. It results from damage to the processes involving articulation. FAS is characterised as a pseudoaccent, where listeners categorise speech alterations as resembling a foreign accent,[1] but actually is an impairment of the finer linguistic abilities such as pauses and vocal stress.[2] FAS is postulated to be due to changes in accentuation of syllables. It is not the result from previous experience from another language and patients with FAS have usually never visited the country their accent resembles.[3]

It has been associated with various aetiologies as surveyed in the literature. The most common aetiology involves a vascular lesion affecting the motor speech areas in the language dominant hemisphere.[4] But it has been linked to other aetiologies such as metastatic breast carcinoma,[5] traumatic brain injury,[1] and even as an initial symptom of multiple sclerosis.[6] Additionally, migraines have been implicated as well.

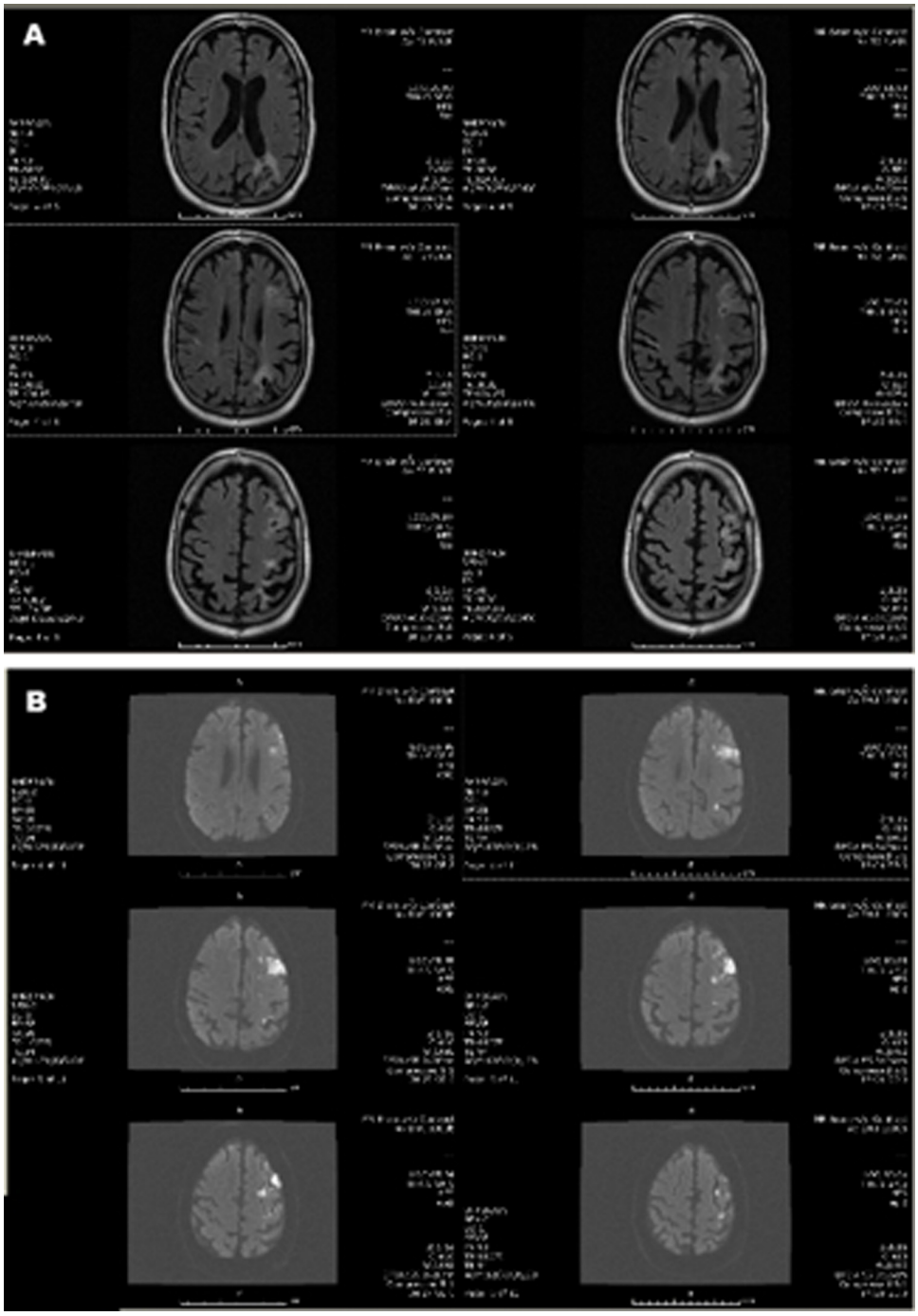

Figure 1 (A,B) MRI brain, selected axial images (Flair, top and diffusion-weighted imaging (DWI), bottom) showing correlated areas of ischemia of the left frontal, involving the middle frontal gyrus, and parieto-occipital regions after the seizure event.

Furthermore, the areas affected in FAS are scattered. The majority of cases involve the motor speech areas, but the dominant precentral gyrus, inferior central gyrus, corpus callosum and insula also have been implicated in FAS.[7 8] Other case reports have shown involvement in the non-dominant rolandic and temporal areas.[6] In a review of 16 cases of FAS, both cortical and subcortical processes were found to be involved.[9] Therefore, there is no clear lesion which can explain FAS consistently. Due to the intricate nature of speech production, possibly a subtle disruption of the motor speech network may lead to FAS, while major disruptions can lead to aphasias. Furthermore, it has been theorised that FAS may be due to a mild deficit in motor planning,[10] where this intricate network is disrupted. In most case reports of FAS, lesion size has been minimal where it is usually less than three centimeters in diameter and typically involves only a single gyrus,[11] this is consistent with the patient's case. Another theory involves the possibility of increased activity of certain areas rather than deficiencies. One report describes an ischemic event of the left putamen in which functional MRI was performed. The authors noted statistically increased activity of the ventral

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angular gyrus and the central sulcus.[12] They concluded these areas are implicated in FAS.

In the differential diagnosis of FAS, apraxia of speech (AOS) is necessary to be ruled out; it is surmised that FAS is not an AOS variant. Unlike AOS, FAS patients do not have difficulties with speech initiation.[13] In diagnosing AOS, the Academy of Neurologic Communication Disorders and Sciences urged that certain clinical characteristics be employed: slow rate of speech, sound distortions resulting in slurred speech, distorted sound substitutions and prosodic abnormalities.[14] These abnormalities were not seen in the patient In diagnosing FAS, certain clinical characteristics have been theorised. These characteristics include the speculation that vowels are more affected than consonants, vowel pronunciation is longer or laxer and lexical stress patterns are affected.[15] Another report states that the main characteristic of FAS was a change in emphasis of syllables.[3]

FAS is a rare phenomenon, but its reversal is rarer. One particular case followed the resolution of FAS. It involved a patient with a left frontoparietal infarct resulting in FAS; subsequently the FAS reversed after a right cerebellar haemorrhage.[8] These CVAs were separated by 3 years. The authors concluded that the mechanisms in accented speech production depend on a delicate bihemispheric balance between cerebral and cerebellar connections.[8] Furthermore, no literature shows the mercurial reversal of FAS after a seizure. The acute ischemia of the left middle frontal gyrus resulted in the patient's FAS. The semiology of the seizure suggests a focus in the vicinity of the new ischemia. This provides the possibility that the increased metabolic demand from the seizure resulted in anoxic damage to the penumbra or even non-adjacent areas containing efferent pathways that fine tuned functions of the initial lesion. This resulted in additional lesions in the described cortical motor speech network,[11] thereby correcting the FAS. This concept has been described,[8] where the first insult lead to FAS and another deficit in the network corrected the aberrations in fine speech control due to another pathway in the network activated or disinhibited. Due to the rapid reversal of the FAS, dedicated speech testing was not performed. The patient declined further language batteries after the reversal and he was lost to follow-up.

Our case describes FAS and its sudden reversal after a seizure, the first reported of its kind. As theorised in the patient, FAS can involve multiple pathways of the complex motor speech network.[11] FAS is a rare phenomenon, one that needs to be better understood.

Learning points

- FAS is a poorly understood phenomenon; it is most likely not a true accent change, rather the perception to the listener as a foreign accent.
- FAS is not a variant of AOS.
- A single cortical area is not responsible for total speech pronunciation, rather it is a complex motor speech network, involving multiple areas.
- This is the first reported case of FAS that was reversed after a seizure.

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Competing interests None.

Patient consent Obtained.

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