Case Report

Foreign Accent Syndrome in a Patient with Posturemic Encepalopathy

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Abstract

Foreign accent syndrome (FAS) is a rare, poorly understood speech disorder. It is characterized by the patient speaking their native language in a different accent foreign to both the speaker and the listener. A majority of previously reported cases have been described in patients with diagnosed organic brain damage and a handful of other psychiatric disorders. FAS was not the result of language experience in our index patient, and there is no history of the patient ever visiting the United States of America. This case is presented because it is the first-ever seen case in the environment.

Keywords: Dialysis, foreign accent syndrome, uremia

Introduction

Foreign accent syndrome (FAS) is a rare motor speech disorder which causes patients to speak their “native” language with an accent which is perceived as nonnative by speakers of the same native community.[1,2] It was first described in 1907 by a French neurologist.[3] Although only relatively few number of cases have been described, FAS has been attributed to various causes ranging from neurological damage to psychological and psychiatric causes. Mixed types involving neurological damage as well as psychogenic disorders have also been described.[4,5] The most common causes of FAS have been shown to include cerebrovascular disorders, multiple sclerosis, Parkinson’s disease, mental disorders such as schizophrenia, and sarcoidosis among others. First described in 1907, the term FAS was first coined by Whitaker in 1982, with an initial set of diagnostic criteria. It included four features: (a) the accent is considered by patient acquaintances and examiner to sound foreign, (b) it is unlike the patient’s native dialect before the

Résumé

Le syndrome de l’accent étranger (SAF) est un trouble de la parole rare et mal compris. Elle se caractérise par le fait que le patient parle sa langue maternelle dans un accent différent étranger à la fois au locuteur et à l’auditeur. La majorité des cas précédemment rapportés ont été décrits chez des patients diagnostiqué des lésions cérébrales organiques et une poignée d’autres troubles psychiatriques. Le SAF n’était pas le résultat d’une expérience linguistique dans notre index patient, et il n’y a pas d’antécédents de visite du patient aux États-Unis d’Amérique. Ce cas est présenté parce que c’est le premier jamais vu cas dans l’environnement.

Mots-clés: Dialysis, foreign accent syndrome, uraemia

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cerebral lesion, (c) it is temporarily correlated to the central nervous system (CNS) lesion, and (d) there is no evidence that the patient is a foreign language speaker. However, even rare cases of nonorganic FAS have been described. This includes schizophrenia, posttraumatic stress disorder, obsessive-compulsive disorder, and conversion disorders. In this very rare case report, we present the first-ever documented FAS in our environment.

Case Report

A 39-year-old woman presented with an established diagnosis of systemic lupus erythematosus (SLE) with lupus nephritis. She was being managed at a tertiary health care center in South Nigeria for approximately 4 years. She had been on conservative management but had been having a progressive worsening of her biochemical parameters with a progressive increase in her serum urea and creatinine and serum potassium.

In addition, the patient was also getting progressively anemic. The patient eventually became uremic with uremic encephalopathy, uremic gastritis, and pulmonary edema. On account of the patient’s clinical condition, doctors at the tertiary hospital where the patient was being managed had prescribed hemodialysis, but the patient refused hemodialysis because she was advised to have a blood transfusion prior to hemodialysis on account of severe anemia. She refused transfusion on account of being a Jehovah’s Witness and discharged herself from the tertiary hospital to a private dialysis center.

On examination at presentation, the patient was in respiratory distress, markedly pale (packed cell volume [PCV] 15%) with marked edema. The patient also had bilateral mid-zone and low-zone crepitations. The abdomen was distended, full with epigastric tenderness. The liver was palpably enlarged 3 cm below the right costal margin. The spleen and kidneys were not palpably enlarged.

CNS examination showed that the patient was well oriented in time, place, and person. There was no focal neurologic deficit, but there was moderate asterixis.

Initial electrolyte, urea, and creatinine results were as follows: urea – 215.8 mg/dL, creatinine – 7.45 mg/dL, Na – 132 mmol/L, K – 5.24 mmol/L, Cl – 108 mmol/L, and HCO₃ – 13.2 mmol/L.

The patient was on prednisolone, azathioprine, hydroxychloroquine, hydrochlorothiazide, and valsartan.

Computed tomography (CT) done previously did not reveal any deficit.

The scanogram was unremarkable. Five millimeter pre- and post-intravenous contrast-enhanced axial CT slices were taken from the base of the skull to the vertex. There was no shift of the midline brain structures. The lateral ventricles, third and fourth ventricles, as well as the basal cistern appeared grossly normal. The paranasal sinuses as well as the mastoid air cells appeared grossly normal. The orbit was grossly normal with no demonstrable defect in the bony orbit or intraocular mass lesion. The overlying soft tissue appeared grossly normal.

Conclusion: Normal cranial CT scan.

PCV done was 15%.

The patient and her relations were counseled for dialysis and blood transfusion. They revealed that they left the tertiary health center when they were told that the patient needed a blood transfusion before she could be dialyzed which they declined on religious grounds. They claimed even in life-threatening conditions; they would not accept a blood transfusion.

The patient was dialyzed, and over the next few weeks, she gradually improved clinically even though hemodialysis was markedly irregular.

The patient continued to improve and returned to the tertiary hospital for the continuation of dialysis since the PCV was now 23% and could be dialyzed without a mandatory transfusion.

The patient presented again in a few weeks with a different complaint. She was speaking English with a distinctly American accent. It had started 3 days prior to presentation. She claimed that the accent was distressing and embarrassing and a source of worry to herself and her family.

The examination revealed a calm patient. There was no neurologic deficit. Mini-mental state examination was within reference values. The patient scored 26. The speech had normal content, but the accent was American and markedly different from her previous speech pattern. The patient had a slightly elevated mood.

Her past medical history showed that she had a previous episode 6 years earlier when she recovered from a coma at the University of Benin Teaching Hospital Intensive Care Unit. She claimed that the symptoms resolved spontaneously after a few days. There has been no other episode since that time.

As stated earlier, a previous CT did not reveal any abnormality. PCV was 27%. All other systems were grossly normal.

Urea and creatinine results showed urea of 126 mg/dL and creatinine was 4.5 mg/dL. There was no asterixis. There were also no gross abnormalities on cranial CT scan, and uraemic encephalopathy had resolved. On account of these, a diagnosis of FAS post uraemic encephalopathy with possible hypomania was made.

The patient was referred to the mental health physician and was eventually placed on haloperidol 2.5 mg/dL and symptoms resolved gradually over the next 2–3 weeks.

Haloperidol was gradually stopped over a 1 month period, and there has been no relapse over the past 10 months. The
patient has been stable clinically without any relapse to a foreign accent.

**Discussion**

This article discusses the case of a patient who developed FAS, a rarely encountered speech disorder, in the absence of demonstrable damage to the CNS. However, as more cases were discovered, another variant markedly similar to this index patient was added that did not conform to the criteria set by Whitaker. This was described as the variant in which the foreign accent of the patient is grounded in underlying psychological issues. It is also referred to as nonorganic functional or psychosomatic.

The index patient had both psychological issues from issues of chronic kidney disease, issues of the psychological effects of needing hemodialysis and issues of refusal to accept transfusion as a result of religious beliefs in the Jehovah’s witnesses sect; and psychosocial issues of an altered body image ad a result of severe bloating (anasarca) due to marked fluid retention. This could have caused a possible psychosomatic problem.

There was also the effect of a general metabolic dysfunction due to marked increases in uremic toxins, urea at a presentation in this patient was 126 mg/dL, and creatinine was 4.5 mg/dL which are surrogate markers of uremia and uremic toxins.

CT scan did not reveal any abnormality. Magnetic resonance imaging (MRI) could not be done.

However, the issue of FAS did not develop in this patient until about 4 weeks after her biochemical parameters had normalized as a result of repeated hemodialysis.

Of note, however, is that the patient had a similar episode 6 years ago after she recovered from a coma secondary to lupus cerebritus and uremic encephalopathy.

Her PCV had also risen to 27% as at the time she presented with the speech abnormality, so it was difficult to postulate a cause and effect model as a result of the uremic encephalopathy she initially presented with.

FAS had also been reported in patients with schizophrenia. Clinically, this patient did not have the clinical features of schizophrenia.

Affective disorders have also been reported as a possible cause of FAS. Haloperidol has been used in similar cases. Treatment with haloperidol of 2.5 mg daily was given to the patient for a few days, and the symptoms resolve. It is inconclusive because it is difficult to project whether the symptoms may have resolved spontaneously. This patient is still being followed up, and there has been no relapse so far.

**Conclusion**

FAS is a rare and yet to be completely understood disorder, and there is no report of any previous cases in our environment.

This index patient, a known SLE patient with CKD, secondary to Lupus Nephritis, did not have any pathological brain damage on CT or clinical examination. Therefore, a psychogenic cause was assumed for which the patient was commenced on haloperidol. The patient has since improved and the symptoms completely resolved.

Whether the haloperidol was responsible for the noted resolution of symptoms is debatable as it is of note that the previous episode that occurred to this patient after a coma 6 years earlier resolved spontaneously.

There is a need for further research into this syndrome and this patient.

Neuroimaging techniques such as MRI, positron-emission tomography, and single-photon emission computerized tomography might have been very useful, but these are not readily available in our environment in Benin City.

In addition, there is a need to be on the lookout for such rare clinical cases like FAS.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. The patients (and/or other educated relative(s), if applicable) gave his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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