

SEVERE DYSARTHRIA DUE TO HYPONATREMIA AND EXTRAPONTINE DEMYELINATION: A SINGLE CASE STUDY

Contributions:

A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
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Abstract

Following an unfortunate iatrogenic incident, a 65 year old woman suffered hyponatremia and extrapontine myelinolysis which left her with severe disabilities, including quadriplegia and dysarthria.

This case report describes acoustic analysis of her voice, speech, and language which was done using PRAAT software. She had severe dysphonia, and her speech involved slow and inaccurate articulator movements. Hypernasality was observed with an overall reduction in speech intelligibility. Hyperadduction of the true and false vocal cords during voice production was also observed. Speech therapists, as part of a multidisciplinary health team, are of great importance in the functional rehabilitation of such patients.

Key words: voice • speech • swallowing • extrapontine myelinolysis • hyponatremia

POWAŻNA DYZARTRIA SPOWODOWANA HIPONATREMIĄ I DEMIELINIZACJĄ POZAMOSTOWĄ – STUDIUM PRZYPADKU

Streszczenie

Po nieszczęśliwym zdarzeniu iatrogenicznym u 65-letniej kobiety wystąpiły hiponatremia i mielinoliza pozamostowa, w wyniku których powstały poważne upośledzenia, w tym tetraplegia i dyszartria. Studium przypadku opisuje analizę akustyczną głosu, mowy i języka pacjentki przeprowadzoną z użyciem programu PRAAT. Badania wykazały poważną dysfonię. U pacjentki podczas produkcji mowy występowały powolne i niedokładne ruchy artykulatorów oraz nosowanie, co skutkowało ogólnym zmniejszeniem zrozumiałości mowy. Natomiast podczas produkcji głosu u kobiety zaobserwowano nadmierne przywodzenie fałdów głosowych i przedśionkowych. Zaanżowanie logopedy jako członka multidyscyplinarnego zespołu terapeutycznego ma ogromne znaczenie dla funkcjonalnej rehabilitacji takich pacjentów.

Słowa kluczowe: głos • mowa • przełykanie • mielinoliza pozamostowa • hiponatremia

Introduction

Osmotic demyelination syndrome (ODS) is characterized by partial damage to myelin sheaths within the central part of the pons. In the outer regions (extrapontine myelinolysis, EPM) [1–3], the acute demyelination is often caused by abrupt fluctuation of serum osmolality involving severe hyponatremia and its excessively rapid correction [2,4]. ODS is therefore an iatrogenic disease.

Hyponatremia is defined as serum sodium less than 135 mEq/L [5]. Epidemiologically, findings of hyponatremia are often seen in a hospital setting, especially in intensive care units [4]. Hyponatremia results from failure of the kidney to eliminate excess water intake, causing cells to swell. When brain cells swell, damage can result

[6]. Hyponatremia is characterized by gastrointestinal tract symptoms such as nausea, vomiting, loss of appetite, neurologic abnormalities, gait disturbances, movement, and behavioral disorders. Expressive language impairment, dysphagia, and dysarthria can also be observed [7–9]. This case report describes the voice, speech, and language outcomes presented by an older adult affected by extra pontine myelinolysis due to excessively rapid hyponatremia correction.

Case report

A 65 year old female came to the department complaining of slurred speech and difficulty swallowing. She had had hypertension for the past 4 years and was under medication for it. Informed consent was obtained from the

subject prior to collection of samples. On 2018/02/02 she was admitted to a private hospital complaining of generalized body pain, tiredness, intermittent fever, and had been vomiting for 2 days. She was diagnosed with enteric fever and enteritis accompanied by severe hypocalcemia, severe hyponatremia (Na 98), and hyperuricemia. She was treated for sodium correction after 3 days of hospitalization. On 2018/02/06, the client again developed loose stool with dehydration and low sodium level (Na 109). Sodium levels returned to normal after being corrected by 3% NaCl. The client was noted to have oral candidiasis and was treated for it. Event-related potentials on 2018/02/10 revealed age-related diffuse cerebral atrophy, and a thin section helical CT showed right otomastoiditis. MRI suggested the presence of hyper intense signals throughout the brain and an extrapontine myelinosis.

Her current physical status showed a reduction in limb movements due to quadriplegia resulting from post-hyponatremia. Although she was normally right-handed, post hyponatremia she predominantly used her left hand. She required complete assistance to fulfill her daily routine. She exhibited signs of emotional instability, frustration, anxiety, depression, and low self-esteem. She also had severe concern about her speech and swallowing difficulties.

Premorbid language history revealed that the client had good comprehension and expression of Tamil and Urdu, both in reading and writing. Post morbid, however, she could comprehend simple commands and execute them but her vocal expression was poor. Verbal imitation of /va/ and /po/ was present, but she expressed her needs through facial gestures and unclear utterances. Her ability to read was well preserved (silent reading). Post morbid writing skills were affected as the client had spastic quadriplegia.

On informal evaluation with the Oral Peripheral Mechanism Examination (OPME), findings revealed that the structure of the lip, jaw, and tongue deviated to the right and the teeth were misaligned; all articulator functions were inadequate for speech production. All vegetative skills such as sucking, blowing, and swallowing were functionally restricted, except that chewing and biting were normal.

Formal assessment methods

A complete test battery tapping the domains of speech and language was carried out. Language assessment was done using the Western aphasia battery, and motor speech was assessed using Frenchay's Dysarthria Assessment (FDA) which includes respiratory and phonatory tests. Subjective voice analysis was carried out with the GRBAS scale and voice was objectively assessed using PRAAT software version 4.4.33 installed on an Acer laptop (Windows 8.1) and recorded using an Xpro-106 Clair microphone. The subject was instructed to sit upright and perform tasks that included sustained phonation of the mid vowel /a/, a conversation task, soft and loud phonation of the vowel /a/, intermittent phonation of vowel /a/, and a glide from low to high. All the tasks were recorded, and parameters such as F_0 in phonation, F_0 in speaking, I_0 , low I_0 , high I_0 , dynamic range, average intensity while speaking, jitter, shimmer, rise

time, and fall time were analysed and an overall dysphonia severity index was calculated.

Informal assessment of swallowing ability revealed no difficulty in consuming liquids, but there was difficulty for solids. Both the client and her caregiver had concerns in this area. On assessment using EAT-10, the scores revealed the client's inability to swallow efficiently and safely.

Language results

The client was able to understand simple commands and perform a few repetitions. However, she could not express herself verbally. Instead, she expressed her needs through simple facial gestures and unclear utterances. Her ability to read was well preserved (silent reading). Non-verbal imitation was present. The Western aphasia battery revealed good performance on auditory verbal comprehension, spontaneous speech, repetition and naming, calculation, and apraxia. On this basis, the client was diagnosed as having Broca's aphasia. Frenchay assessment pointed to severe dysarthria.

Speech results

Evaluation of the respiratory system showed that the coordination of respiration for swallowing and speech were inadequate. Breathing patterns for speech and non-speech activity were observed to be abdominal. The Maximum Phonation Duration (MPD) task revealed inadequate maximum phonation duration. Her speech was slurred, and she had slow and imprecise articulator movements. It was classified as dysarthric. Hyper nasality was also observed.

Voice profile

No harmful vocal and non-vocal habits were reported pre-morbid. On Perceptual Analysis of Voice, the voice's quality was observed to be strained and strangled, the pitch was inappropriate (low) for the client's age and gender, with limited pitch range, and pitch breaks were present. Loudness was inadequate for spontaneous speech.

Subjective evaluation of the voice was done using the clinician rating evaluation scale (Consensus Auditory Perceptual Evaluation of Voice, CAPE-V) and revealed severe deviance of overall quality (85/100), severe roughness (80/100), and severe strain (70/100), whereas there was mild deviance for pitch (20/100), loudness (20/100), and breathiness (10/100). Resonance was hypernasal. The GRBAS scale indicated an overall severe degree (score of 3), with subscales of severe roughness (3); mild breathiness (1); moderate asthenia (2); and moderate strain (2).

With objective evaluation of the voice, the dysphonia severity index revealed severe dysphonia. The acoustic analysis of fundamental frequency showed 138 Hz, and a value of 54 dB for intensity. Jitter and shimmer were 5.0% and 12.2%, respectively, highly deviant and unstable.

Discussion

This paper describes a case report of extrapontine myelinolysis related to rapid hyponatremia correction in an older

Table 1: Language history of client pre and post morbid

Language	Pre morbid				Post morbid			
	Comprehension	Expression	Reading	Writing	Comprehension	Expression	Reading	Writing
Tamil (L1)	✓	✓	✓	✓	✓	×	✓ (Silent)	NA
Urdu	✓	✓	✓	✓	✓	×	✓ (Silent)	NA

* NA- Spastic Quadriplegic

Table 2: Result of GRBAS

GRBAS	Scoring
Grade	3 (Severe)
Roughness	3 (Severe)
Breathiness	1 (Mild)
Asthenia	2 (Moderate)
Strain	2 (Moderate)

Table 3: Result of Consensus Auditory Perceptual Evaluation of Voice – CAPE-V

Parameters	Scoring	Indication
Overall severity	85/100	Severe deviant
Roughness	80/100	Severe deviant
Breathiness	10/100	Mild deviant
Strain	70/100	Severe deviant
Pitch	20/100	Mild deviant
Loudness	20/100	Mild deviant
Resonance	Hypernasal	

adult presented with hypertension. Several cases have been described previously, and these indicate that lesions can take place in areas of the brain such as the thalamus, mesencephalon, basal ganglia, cerebellum, central and temporal white matter, external and extreme capsules. Generally, the damage is referred to as extrapontine myelinolysis [2,4,12]. Central pontine and extrapontine myelinolysis are united under the name osmotic demyelination syndrome and are frequently related to rapid hyponatremia correction [2,4]. Our client was also subject to rapid correction of hyponatremia.

The osmotic myelinolysis is the syndrome of inappropriate secretion of the antidiuretic hormone. Such alteration of the hormone responsible for water absorption will increase the amount of water in the blood serum, causing abnormal serum osmolality such as hyponatremia, hypo magnesium, and hypo potassium. In the treatment of these regions, electrolyte fluctuation occurs easily in the context of hormone

alteration. Hyponatremia is commonly observed in patients with electrolyte disturbance, which tends to worsen with the administration of steroids. During hyponatremia, electrolytes are transported from the intracellular to the extracellular environment to prevent the entraining of water in the cell. From this rapid hyponatremia correction, the incapacity of maintaining intracellular hypertonicity is due to the inability to regenerate intracellular osmolality resulting in cell reduction and death. Oligodendrocytes are particularly vulnerable to destruction by cell volume shrinkage, resulting in myelin loss, ultimately resulting in extrapontine demyelination [11,12]. The referred patient suffered an excessively rapid correction of serum sodium level, which went from 98 mEq/L to 109 mEq/L in less than 24 h. The client was provided with a 3% NaCl correction in the month of February.

Studies of clients with EPM have shown very similar features. Two cases were children with EPM associated with rapid hyponatremia correction in which areas of the basal ganglia and adjacent cerebral cortex were affected [10]. Reported neurological deficits included dysarthria and dysphagia, emotional ability, and spastic quadriplegia. The patients gradually improved, but the neurological deficits were not wholly reversed. EPM can occur postoperatively after a suprasellar arachnoid cyst resection. MRI results show damage in the basal ganglia and there is generalized dystonia, dysarthria, and dysphagia [13].

Considering that ODS is an iatrogenic disease, severe neurological deficits have significant impacts on the speech and language subsystem and significantly affect quality of life.

Conclusions

Extrapontine myelinolysis (EPM) causes severe neurological deficits, and in this case quadriplegia and speech and swallowing disorders, with severe impacts on verbal communication, eating, and quality of life. Broca's aphasia, dysphagia, and dysarthria are disorders commonly described in EPM cases, and they were present in this case. In terms of the voice, they gave rise to a strained and strangled voice with all pitch elements, loudness, and quality being affected. As part of a multidisciplinary health team, speech therapists are of great importance in the functional rehabilitation of such patients.

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