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Case reports

Speech, swallowing and quality of life outcomes in extrapontine myelinolysis: a case report of a teenager with central nervous system germ cell tumor

Achados de fala, deglutição e qualidade de vida na mielinólise extrapontina: relato de caso de uma adolescente com germinoma do sistema nervoso central

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ABSTRACT

This paper is a case report describing the speech-language and quality of life outcomes presented by a teenager with a central nervous system germ cell tumor affected by extrapontine myelinolysis. The extrapontine myelinolysis is an acute demyelinating disease that can be caused by abrupt changes in serum osmolality, as in the rapid correction of hyponatremia. Data were obtained from clinical assessment protocols, pediatric inventory about quality of life and medical records information. The patient presented mutism, dysphagia and dysarthria caused by extrapontine myelinolysis, with significant impacts on their verbal communication, feeding and quality of life. The scores of quality of life after extrapontine myelinolysis decreased compared to the time before the disease. She received speech-language therapy during hospital admission and in the outpatient clinic. We observed changes in patterns of speech, swallowing and mobility of orofacial structures; with partially reverse of neurological deficits. The speech therapists, as part of a multidisciplinary team of health care, are of great importance in the functional rehabilitation of patients affected by this disease.

Keywords: Speech, Language and Hearing Sciences; Speech; Swallowing; Quality of Life; Myelinolysis, Central Pontine; Rehabilitation

RESUMO

O presente trabalho é um relato de caso que descreve os achados fonoaudiológicos e de qualidade de vida apresentados por uma adolescente com um tumor do sistema nervoso central acometida pela mielinólise extrapontina. A mielinólise extrapontina é uma doença desmielinizante aguda que pode ser causada por variações abruptas na osmolaridade sérica, como o que ocorre na rápida correção da hiponatremia. Os dados foram obtidos a partir da avaliação clínica fonoaudiológica, de questionários pediátricos sobre a qualidade de vida e dados contidos no prontuário médico. A paciente apresentou mutismo, disartria e disfagia decorrentes da mielinólise extrapontina, com impactos significativos na sua comunicação verbal, alimentação e qualidade de vida. Os escores de qualidade de vida após a mielinólise extrapontina apresentaram piora quando comparados aos do período anterior à doença. Recebeu atendimento fonoaudiológico hospitalar e ambulatorial. Foram observadas evoluções nos padrões de fala, deglutição e mobilidade das estruturas orofaciais, com reversão parcial dos déficits neurológicos. A atuação fonoaudiológica, como parte de uma equipe multiprofissional de saúde, é de grande importância na reabilitação funcional dos pacientes acometidos por esta doença.

Descritores: Fonoaudiologia; Fala; Deglutição; Qualidade de Vida; Mielinólise Central da Ponte; Reabilitação

INTRODUCTION

The osmotic demyelination syndrome (ODS) is characterized by the partial damage of myelin sheaths in the central part of the basis pontis (central pontine myelinolysis - CPM)[1], or in pontine regions (extra-pontine myelinolysis - EPM)[2,3]. This acute demyelinating disease can be caused by abrupt fluctuation in the serum osmolality, being frequent related to the severe hyponatremia and its rapid correction[2,4].

Clinical manifestations are varied and include neurological deficit related to the affected areas. Movement and behavioral disorders, mutism, dysphagia and dysarthria can be observed[3-5].

This disease features serious progression and often fatal. The recovery can be spontaneous and gradual, with partial or complete recovery of neurologic sequelae[2,4-6]. Best results are described when occurred during childhood[4].

Patients with suprasellar tumors present frequent electrolytic disorders associated to hormone disorders. Osmotic myelinolysis, especially EPM may occur, and the diagnosis is based on clinical presentation and the images of magnetic resonance imaging (MRI)[6].

In this case report we intend to describe the speech-language and the quality of life outcomes presented by a teenager with a central nervous system germ cell tumor in suprasellar region, after being affected by extrapontine myelinolysis associated to rapid hyponatremia correction.

CASE REPORT

We describe a case of a 14-year old girl diagnosed with central nervous system germ cell tumor in suprasellar region, under treatment in the National Institute of Cancer Jose de Alencar Gomes da Silva. The patient whose case is described in this study is part of a major study with project approved by the Committee of Ethics in Research (protocol 492.325/2013), and we obtained the parents' consent form and the adolescent's assent.

Speech, orofacial motricity and swallowing outcomes described in this paper were obtained from speech-language clinical evaluation. Regarding speech, we evaluated features of articulation, speed, intelligibility, pneumophonic coordination and prosody; applying tasks of repetition, nomination and spontaneous speech. About the orofacial motricity, we observed posture, strength and mobility of the structures.

Swallowing evaluation included the observation of the following clinical parameters: bolus capture, oral-motor control, ejection, extra-oral scape, stasis in oral cavity, nasal scape, time of oral transit, hyolaringeal movement, multiple swallowing, cough, throat clearing, "wet voice", dyspnea and cyanosis during the feeding. The evaluation also included the Functional Oral Intake Scale – FOIS[7] use. During the swallowing assessment it was performed cervical auscultation[8] and the oxygen saturation monitoring.

The health team used as instruments of quality of life assessment the PedsQL™ 4.0 Generic Core and 3.0 PedsQLTM Cancer Module protocols Brazilian versions. PedsQL™ Inventory 4.0 is multidimensional, including the assessment of four scales: physical, emotional, social and school functioning[9]. The PedsQL™ 3.0 Cancer Module inventory measures the disease and treatment impact on the quality of life of children with cancer through items divided into eight scales: pain and hurt, nausea, procedural anxiety, treatment anxiety, worry, cognitive problems, perceived physical appearance and communication[10]. Besides data from the assessments, the team also used information contained in the medical chart.

The teenager presented panhypopituitarism and diabetes insipidus (DI) using desmopressin (DDAVP). During the 6th cycle of the chemotherapy treatment which she was submitted (Carboplatin + Etoposide and Ifosfamide + Etoposide), she presented febrile neutropenia, mucositis and superior airways infection, demanding to be hospitalized.

The patient did not presented speech, swallowing and orofacial myofunctional disorders related to the central nervous system tumor, previous to extrapontine myelinolysis.

Throughout her admission period, she presented degradation of the conscious level. It was detected severe hyponatremia with serum sodium level of 103 mEq/L, being initiated the sodium replacement. The patient was referred to the unit of pediatric intensive therapy. After less than 24 hours the serum sodium level went up to 128 mEq/L reaching in 48 hours, level of 150 mEq/L. At this point, the patient was alert with efficient verbal communication and oral diet with multiple consistencies, without swallowing disorders.

Five days after the hyponatremia correction, she began to present a clinical picture of sleepiness, movement and speech disorders, being observed slow speech pattern, with articulatory imprecisions and pneumophonic incoordination.

She presented respiratory patterns disorders, demanding noninvasive ventilatory support (BIPAP). In the following days, her respiratory patterns improve, starting to ventilate in room air. She presented mutism, with ocular opening in the absence of verbal answers to interaction, and emotional lability and constant crying. She was incapable to perform volunteer oral movement. The saliva swallowing was altered, observing accumulation in oral cavity and extra-oral escape.

The brain MRI carried through in this period disclosed hyperintense signal in T2 and FLAIR cortic-subcortical areas showing contrast enhancing and diffusion restriction scanning in some gyrus of frontal, parietal, occipital and temporal regions. Additionally, it was observed hyperintense signal in T2 and FLAIR, also after contrast and diffusion restriction in the basal ganglia bilaterally. The image aspect in correlation with clinical and laboratorial data led to the diagnosis of extrapontine myelinolysis.

RESULTS

Extrapontine myelinolysis, in result of the rapid hyponatremia correction, caused severe functional sequelae related to patient's verbal communication and swallowing. The speech-language therapy in hospital was carried through daily during the attendance, in the unit of intensive therapy and infirmary.

The post extrapontine myelinolysis outcomes related to swallowing included damages to the oral and pharyngeal phase, with difficultness in the bolus capture, oral motor control, chewing and ejection of foods, which caused an increased time of oral transit, extra-oral escape of liquids, oral cavity stasis of foods and cough during feeding; an indication of laryngeal penetration and/or aspiration.

The health team did not carried through swallowing instrumental assessment since the patient was admitted in the unit of intensive therapy with unstable clinical picture, and they considered that, at this point, the outcomes of the instrumental assessment would not change the therapeutic conduct applyed[11].

Due to the picture of sleepiness and the alterations in the swallowing dynamic, featuring a neurogenic oro-pharyngeal dysphagia, the oral diet was derailed by the speech and language team, being indicated the nasoenteric tube (NT) as feeding alternative.

Handling dysphagia, the speech therapists conducted indirect swallowing therapy (oro-myofunctional exercises and maneuvers) and direct therapy using semiliquid foods, and later, liquids. With the intervention, we observed more organized swallowing dynamic, better patterns of bolus capture, oral manipulation and ejection of the food, more adequate time of oral transit and absence of suggestive clinical signals of aspiration, which enabled the return of exclusive oral feeding.

After 8-day speech-language intervention directed to swallowing rehabilitation with improvement to the functional pattern, the team considered to be safe the reintroduction of oral diet and removal of the NT. The alternative feeding via was used during 20 days, since dysphagia was detected.

The patient evolved from level 1 (no oral intake) in FOIS scale - at the beginning of the disorder - to level 5 (total oral intake of multiple consistencies requiring special preparation or compensations) - at the moment of hospital discharge.

Regarding the communication, the initial picture after extrapontine myelinolysis was of mutism being observed, in the absence of verbal communication, head movements and hand gestures in response to the requests. The language comprehension related to simple verbal information was preserved. In this context, the speech-language intervention involved language stimulation conducts aiming to reestablish verbal communication.

Mutism condition last 14 days and it began six days after hyponatremia correction. When reestablishing verbal communication, initially the patient emitted sounds and isolated words being later capable to produce simple sentences. Speech was dysarthric featured by articulatory imprecisions, much reduced speed of speech, altered pneumophonic coordination and prosody. Dysarthria was classified as mixed, considering the damage of more than one motor system (pyramidal and extrapyramidal), with components of the spastic and hypokinetic dysarthrias[12].

Throughout the therapy sessions, we observed improvement in oral production, with upturn in the articulatory precision and speed of speech. Although the positive outcomes the verbal fluency and the speech articulatory pattern were not totally reestablished.

Regarding the orofacial motricity, initially she presented incapacity of voluntary mobilization of lips, tongue, jaw and facial mimic. The stimulation to the voluntary mobilization of the phonoarticulatory organs was carried through ever since the beginning of the speech-language intervention. When reestablishing the execution of the orofacial movements, they were very slow and inexact. The tongue movements were affected

by tremor. Oromyofunctional therapy was carried through, with exercises directed to the enhancing of amplitude, precision and movements speed, besides the strength gain of orofacial muscles. We observed improvement in the muscle mobility of lips, tongue and jaw with positive impacts in speech and swallowing.

After hospital discharge, the speech-language treatment was carried through in the institution clinic, following the previous conducts initiated during the admission, focusing on speech alterations, oral motricity, and swallowing, still partially reverted due to the extrapontine myelinolysis.

The brain MRI carried through at about two months after the first exam disclosed almost total regression of hyperintense signal in T2 and FLAIR cortic-subcortical, showing contrast-enhancing and diffusion restriction scanning some gyrus of frontal, parietal, occipital and temporal regions. We also observed regression in images with hyperintense signal in T2 and FLAIR and contrast-enhancing in the basal ganglia bilaterally, with areas in the present study of bilateral and symmetrical laminar encephalomalacia. This aspect could correspond to the regression of the previous extrapontine myelinolysis.

The total scores of quality of life in the questionnaires PedsQL™ 4.0 and PedsQL™ 3.0 was diverse as shown in table 1. It is considered total maximum score of 100 in these tests, and bigger the scores better is the quality of life[9,10].

Comparing the scores from the quality of life assessment before EPM and during oncology treatment to the scores of five months after EPM – at a time when deficit had been partially reverted - all scores presented considerably worsening, demonstrating the impact of the alterations due to EPM in patient's quality of life.

Table 1. Pre and post extrapontine myelinolysis scores in the quality of life

Scores of Quality of Life	PedsQLTM 4.0 Teenager report	PedsQLTM 4.0 Parents report	PedsQLTM3.0 Teenager report	PedsQLTM3.0 Parents report
Pre extrapontine myelinolysis	68,47	69,56	82,69	77,88
Post extrapontine myelinolysis	40,27	38,89	62,04	66,67

DISCUSSION

This paper describes a case report of extrapontine myelinolysis related to rapid hyponatremia correction in a teenager suffering from a central nervous system germ cell tumor, who presented also panhypopituitarism and diabetes insipidus.

In 1959, Adams et al.[1] first described central pontine myelinolysis as a disease related to alcoholism and malnourishment. Subsequently, several cases were described and the concept was amplified due to the acknowledgement that the lesion could take other areas from the central nervous system as thalamus, mesencephalon, basal ganglia, cerebellum, central and temporal white matter, external and extreme capsules, lateral geniculate body, cerebral cortex deep layers, hippocampus and corpus callosum, therefore called extrapontine myelinolysis[2,4,6].

The CPM and EPM are reunited under the title of osmotic demyelination syndrome and are frequently related to rapid hyponatremia correction[2-4]. The CPM is the most common disorder being followed by the EPM in 10-30% of the cases[13]. The EPM can be isolated – without involving the central pontis region - and is rare in pediatrics population[13,14].

The hormone disorder more frequently related to osmotic myelinolysis is the syndrome of inappropriate secretion of the antidiuretic hormone. Such hormone alteration often occurs in the panhypopituitarism context secondary to sellar and suprasellar tumors[15].

In the treatment of these regions tumors, electrolic fluctuation occurs easily in the context of hormone alteration. The diabetes insipidus is commonly observed in patients with suprasellar germ cell tumors and tend to be worse with the administration of steroids, which increase the chances of occurring osmotic fluctuation and demyelination diseases. The excessive compensation of antidiuretic hormone may cause hyponatremia in these patients[6].

During the hyponatremia, electrolytes are transported from the intracellular environment to the extracellular to prevent the entrance of water in the cell prevenient from this external hypotonic environment.

From this rapid hyponatremia correction, the incapacity of maintaining the intracellular hypertonicity due to the damaged regeneration of the intracellular osmolality results in reduction and death of the cell. Oligodendrocytes are particularly vulnerable to death by cell volume shrinkage, resulting in myelin loss[14].

The referred patient suffered an excessively rapid correction of the serum sodium level, which went from 103 mEq/l to 128 mEq/l in less than 24 hours, exceeding the limits considered safe for correction that should not be over 0.5 mEq/l/h[4].

The clinical manifestations of the osmotic demyelination syndrome has a biphasic course with a picture of encephalopathy and seizures due to hyponatremia, which is reverted after corrected, only to deteriorate in a few days, when can be observed general muscular weakness, mutism, dysarthria, dysphagia, catatonia and the locked-in syndrome[2-4].

In this paper case report, the teenager presented degradation of the conscience level associated to hyponatremia. After the sodium levels correction, she was alert and without neurological deficits. Only five days after later, she presented movement and behavioral (catatonia and emotional lability) alterations, mutism and dysphagia.

Literature has some reports of EPM in children. Very similar features to the observed in this paper case report were described by Choe et al.[6], who reported two cases of children with germ cell tumors that presented EPM associated to rapid hyponatremia correction. Areas of the basal ganglia and adjacent cerebral cortex were affected. Neurological deficits included pseudobulbar palsy symptoms, involving dysarthria and dysphagia, emotional lability and spastic quadriparesis. Patients gradually improved, but neurological deficits were not completely reverted.

Zhao et al.[5] described a case of EPM in the postoperative of a suprasellar arachnoid cyst resection. The images of magnetic resonance disclosed damages in the basal ganglia. The patient, a 3-year old boy, presented generalized dystonia, dysarthria and dysphagia. After three 3-month rehabilitation, the neurological alterations were partially reverted.

Srimanee et al.[15] reported two cases of EPM in adults with sellar tumors, who presented hypopituitarism and secondary adrenal insufficiency. The authors alert for the risk of the OD occurrence in this group of patients and recommend careful therapy of corticosteroids and hyponatremia correction, as well as proper monitoring of the adjusted serum sodium levels for the prevention of the ODS.

Some authors describe full neurological recovery[16], while others partially recovery presenting neurological sequelae[5,6,13,14], as in the case reported in this present study, which dysarthria remained in the patient.

The EPM-related repercussions go beyond the physical damages caused by the disease with impacts in the patients' quality of life, as observed from the reduction scores described in Table 1.

Considering the ODS an iatrogenic disease, although rare, which causes severe neurological deficit with significant impacts in the quality of life or even death is essential to recognize the risk patients and the proper hyponatremia correction. The strategies of OSD prevention are mandatory[17].

FINAL CONSIDERATIONS

Extrapontine myelinolysis causes severe neurological deficits as speech and swallowing disorders, with impacts on verbal communication, feeding and patients' quality of life.

Mutism, dysphagia and dysarthria are disorders described in EPM cases, as the presented in this paper. The speech therapists as part of a multidisciplinary health team are of great importance in the functional rehabilitation of patients affected by this disease.

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