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SHORT COMMUNICATION

COMMUNICATION DISORDERS AS AN EARLY SYMPTOM OF MOTOR NEURON DISEASE (MND)

ZABURZENIA KOMUNIKACJI JAKO WCZESNY OBJAW ZESPOŁU NEURONU RUCHOWEGO (MND)

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ABSTRACT

Introduction

Motor Neuron Disease (MND) is a term that concerns the whole group of disorders with degenerative nature within both upper and lower motoneurones. More often MND is considered to be a multisystemic disease with a special predilection to motoneurons. The most common symptom is asymmetric weakened strength in one limb, usually arm, manifesting in stiffness, slowing down and clumsiness of motion.

Aim

Description of diagnosing MND based on rare early manifestations, voice and speech disorders.

Material and methods

Two female and one male Patient, at the age of 62, 62, 78 were referred to the Department of Phoniatrics and Audiology due to voice and speech disorders. The Patients underwent complete phoniatric and neurological assessment. Phoniatric examination included perceptual voice evaluation, examination of the voice emission, evaluation of larynx phonological activity with videolaryngoscopy and acoustic voice analysis.

Results

Variety of the symptoms was connected with abnormalities of the nerve impulses transmission to the internal muscles of the larynx. Abnormalities in the evaluation of the phonation activity confirmed objective examination of acoustic voice analysis (MDVP), which revealed significant disturbances in all parameters defining frequency perturbation measures, most of all jitter.

Conclusions

Communication disorders are rarely the first symptoms of MND, but they can appear as the main complaint in the first period of the disease when no more symptoms occur. Motor neuron disease of the larynx may occur in various forms depends on damaged muscles: dyspnea, hypofunctional dysphonia or atrophy of the voice muscles.

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STRESZCZENIE

Wstęp

Choroba neuronu ruchowego (Motor Neuron Disease, MND) jest grupą schorzeń o nieznanej etiologii, charakterze zwyrodnieniowym w obrębie górnego oraz dolnego neuronu ruchowego. Choroba charakteryzuje się podstępnym przebiegiem. Najczęściej zauważalnym wczesnym objawem jest asymetryczne osłabienie siły mięśniowej jednej ręki, utrata sprawności manifestująca się sztywnością, spowolnieniem oraz niezdarnością wykonywanych ruchów.

Cel

Opis diagnozy choroby neuronu ruchowego na podstawie wczesnych objawów w postaci zaburzeń komunikatywnych.

Materiał i metody

Dwie kobiety oraz jednego mężczyznę w wieku kolejno 62, 62, 78 lat, hospitalizowano w Klinice Foniatrii i Audiologii z powodu zaburzeń głosu i mowy oraz okresowej duszności. U pacjentów przeprowadzono badanie foniatryczne oraz badanie neurologiczne. Badanie foniatryczne obejmowało ocenę percepcyjną głosu, badanie elementów emisji głosu, ocenę czynności fonacyjnej krtani w badaniu laryngoskopowym oraz analizę nagrań głosu.

Wyniki

W badaniu foniatrycznym zaburzenia głosu polegały na niestabilności głosu, chrypce, drżeniu głosu, dodatkowo u jednej z chorych występowała okresowa duszność. Różnorodność objawów związana była z nieprawidłowością przewodnictwa impulsów nerwowych do poszczególnych grup mięśni wewnętrznych krtani. Nieprawidłowości w ocenie czynności fonacyjnej krtani potwierdziło obiektywne badanie analizy tonu krtaniowego (MDVP), gdzie stwierdzono znaczne odchylenia w zakresie wszystkich parametrów określających względne zmiany częstotliwości, zwłaszcza jitter tj. względną zmianę częstotliwości z okresu na okres. W badaniu neurologicznym stwierdzono wygórowane odruchy ścięgniste, fascykulacje języka, zaniki mięśni kłębu kciuka i/lub palca małego.

Wnioski

Zaburzenia procesu komunikatywnego rzadko są pierwszym objawem MND, mogą być jednak główną albo jedyną skargą chorych w początkowym okresie choroby. MND w krtani może przebiegać pod różnymi postaciami w zależności od grupy zajętych mięśni: od duszności poprzez dysfonię hipofunkcjonalną po zmiany w postaci atrofii mięśni głosowych.

Słowa kluczowe: choroba neuronu ruchowego, zaburzenia głosu, dysfonia, chrypka

Introduction

Communication disorders, which are known in neurological diseases and are described by neurologists or psychiatrists mainly concern speech disorders of the nature of aphasia or dysarthria (*Roth et al. 1996, Beukelman et al. 2011*). Nevertheless, not as much attention is paid to voice disturbances, despite the fact that they may appear as an early symptom of neurological disorders (*Stepp et al. 2017*).

Definition of Motor Neuron Disease (MND) is used nowadays in respect of the group of diseases with unknown etiology, with prevalence of 2–3/100,000 people, with typical onset between 50 to 60 years (*Cleveland et al. 2004*). Degenerative changes, therefore, can concern both superior and inferior motor neuron, resulting in progressive muscle weakness, atrophy and spasticity (*Kierman et al. 2011*). Contrary to multiple sclerosis and polyneuropathy they do not lead to sensory loss (*Balendra et al. 2016*). Unlike myasthenia gravis, they do not concern motor nucleus of oculomotor nerve. (*Talbot et al. 2012, Wang et al. 2017*).

In 1896 Jean Martin Charcot described Amyotrophic Lateral Sclerosis (ALS) with lesion of superior and inferior motor neuron, which was known in french literature as Charcot disease, in Anglo-Saxon literature as a MND, in USA as a Lou Gehring disease (*Cleveland et al. 2004, Simon et al. 2019*). MND is not homogenous medical condition and include ALS, primary lateral sclerosis (PLS), progressive bulbar paresis, spinal-muscular-atrophy-like syndrome and other rare syndromes such as Kennedy syndrome (*Howard et al. 2002, Balendra et al. 2016, Hashizume et al. 2017*). There are three diagnostic categories: Clinically definite, probable and possible MND. The Revised El Escorial diagnostic criteria and Awaji electrodiagnostic criteria are useful in setting a diagnose and evaluate evidence for progression (*Balendra et al. 2016, Makkonen et al. 2016*).

Current outlook on etiopathogenesis concerns mainly low socioeconomic status, environmental chemicals, dietary deficiency of neurotrophic factors and glutamate metabolism disorders. One or more of those factors in a person with particular genetic predisposition may lead to lesion of the superior and inferior motor neuron cells (*Comba et al. 2007, Tylleskdr et al. 1991*). Commonly "the split hand" is seen, which is the muscle atrophy of the thenar and hypothenar (*Kierman et al. 2011*, *Simon et al. 2019*), though the first symptom of MND may be bulbar presentation. In treatment riluzole - a glutamate antagonist, is approved, but it prolongs survival with up to three months (*Howard 2002*). There is no effective treatment of MND, treatment remains focused on the symptoms (*Ball et al. 2004, Miśkiewicz et al. 2019, Simon et al. 2019*). The disease is usually fatal within 2 to 5 years after setting a diagnose (*Ucelli et al. 2007, Balendra et al. 2016*).

The aim of the work was the presentation of commonly occurring communication disorders as a main symptom of motor neuron disease, basing on 3 Patients' cases from the Clinic.

Aim

The aim of this study it is a decscription of diagnosing motor neuron disease based on rare early manifestations, voice and speech disorders.

Material and methods

In the Department and Clinic of Phoniatrics and Audiology three Patients were hospitalized with voice and speech disorders as one of the first symptoms of MND. Patients, 62 years-old man (Patient 1), 78 years-old woman (Patient 2) and 62 years-old woman (Patient 3) were admitted to the hospital with gradually progressing hoarseness, voice fatigue and effort dyspnoea. The Patient 1 had additionally movement disorders for several years – stiffness and slowness of movement, initially on the left side, now within four limbs. In the Patient 3 there were additionally difficulties in swallowing, disturbance of phoniatric-articulatory and respiratory coordination and emotional instability. Due to the increasing dyspnoea in a Patient 3, a tracheotomy was performed. It was followed by Kashima's procedure (glottis widening) on the left side. Even after that, the tracheostomic tube could not have been removed because of the persistent dyspnoea.

All of the Patients were previously hospitalised on neurological departments, where the following diagnoses were set: MND with symptoms of multilevel damage of the anterior

spinal cord motoneurons in Patient 1, Parabullbar palsy in Patient 2, Multiple System Atrophy (MSA) in Patient 3.

Results

Neurological examination

In all of the Patients, in neurological examination increased reflexes, muscle twitch of the tongue, shoulder girdle, thighs and calves, muscle atrophy of the thenar and hypothenar were found.

Laryngological examination

Out of the relevant abnormalities in laryngological examination lack of throat reflex, limited tongue mobility with vibrations on both sides in all of the Patients. In Patient 1 on both vocal folds along their free edge glottic sulcus, more distincted on the left fold, in Patient 2 on right fold along the free edge glottic sulcus, additionally dissociated paresis of soft palate was observed (no fonation movement, with preserved movement during swallowing), with food remaining in both piriform sinuses. In Patient 1 and Patient 2 in the breathing phase, the abduction of vocal folds was only to the intermedia position, absence of a complete glottalisation in intramembranous part of glottis vocalis or in a shape of hourglass. In Patient 3, right fold was wider than the left one, there was also a scar in the back on the left fold after the Kashima surgery, mobility of both folds in the area of the cricoarythenoid articulation retained, disappears after few seconds.

Phoniatric examination

In phoniatric examination the voice of the Patient 1 was assessed as matte, quiet, occasionally hoarse with tremor, formed with neck muscle hyperkinesia, with weak activation of supraglottic resonators, 20s phonation time, average speaking voice position varying from 180 to 220 Hz and speech range from 150 to 250 h. Expiration of the articulation drive was noticed. The voice of the Patient 2 was matte, quiet, sometimes hoarse with trembling, with a 4s phonation time, an average spoken voice position varying from 260 to 280 Hz and a voice range from 210 to 320 Hz, expiration of the phonation drive. Speech was slow, blurred, monotonous, devoid of rhythm and melodics, with a nasal tint. There were moments of repeating the final syllables and uncoordinated articulation movements. The articulation of many sounds was blurred. Hypernasal speech was observed. Voice of Patient 3 was resonant, created with neck muscle hyperkinesia, with low activation of the supraglottic resonators, 12s phonation time, average speaking voice position ranging from 180 to 220 Hz and a voice range from 150 to 250 Hz, and expiration of the articulation drive.

Throat laryngoscopy

In throat laryngostroboscopy in Patients 1 and 2 the oscillations of vocal folds were uneven, non-simultaneous, with increased vibration amplitude with no marginal shift and phonation shorting; while increasing the volume of the voice, there was a tendency to sphincteric contraction of the folds. Only in Patient 3 vibration of vocal folds were of the same, simultaneous, with no marginal shift on the left voice fold, full phonation shorting; While increasing the volume of the voice, there was a tendency to sphincteric contraction of the folds.

Acoustic analysis of the voice

Acoustic analysis of the laryngeal tone with the MDVP method showed the same changes in all the 3 Patients: significant deviations in the range of all parameters defining relative changes in frequency, especially jitter, i.e. the relative change in frequency from period to period. Slightly changed parameters specifying amplitude changes (Shimmer, vAm). Significant deviations in VTI, FTRI, ATRI and to a lesser degree DSH parameters.

The results in our Patients confirmed the changes in the sound of the voice, as well as the atrophic changes in the vocal folds, which were observed in the videostroboscopic picture.

As a treatment, physical therapy was implemented in the Patients, including calcium iontophoresis, electrifications, of the larynx, as well as voice, breathing and articulation exercises.

Discussion

ALS typically appears with spastic-flaccid dysarthia, including symptoms of slow speed rate and strained-strangled voice as a result of spasticity and hoarse, breathy vocal quality, consonant distortions, short phrases as a result of flaccidity. Hypernasality can be a feature of both of the components. These are the symptoms that appeared in all of our Patients. Roth *et al.* (1996) observed, that the first symptom of MND may be spasmodic dysphonia. They reported, that bulbar symptoms are the earliest in in 25% of the patients (*Roth et al. 1996, Traynor et al. 2000*) reported, that in the bulbar-onset population of ALS Patients, dysarthria was eight times more common than dysphagia as an initial symptom (*Travnor et al. 2000*). According to Balendra *et al.* approximately 30% of the Patients present with one of the bulbar symptoms including dysarthia, dysphagia and sialorrhea (*Balendra et al. 2016*). Bak *et al.* described six patients with MND, who had communication disorders as an early and main symptoms of the disease (*Bak et al. 2000*). Among 79 patients with MND, which Carrow *et al.* in 80% was described with harsh voice quality, 73% with hypernasality, 63% with voice tremor, 60% with strained – strangled voice, 38% elevated and 8% reduced average speaking voice position (*Carrow et al. 1974*). Silbergleit *et al.* investigated acoustic analysis of voice and discovered, that it can be sensitive indicator of early deterioration in ALS (*Silbergleit et al. 1997*). It is used nowadays to detect changed features of the voice, which can be confirmed even without clinically significant changes. The differences for jitter, significantly reduced MPFR, variability in shimmer and SNR scores may indicate limitation of the voice in ALS.

Ball *et al.* proposed introduction of the protocol to diagnose and monitor early bulbar paresis basing on the voice estimation, rate of the speech and effectiveness of communication with MND patients (*Ball et al. 2001*). ●In the paper "The Disablement Model" proposed by WHO there are proposed three levels of disorder: impairment, activity and participation, and it was used as a framework to monitor dysfunction. Their results show, that early communication disorders are evident in all of the parameters of "The Disablement Model". All of the estimated parameters were significantly changed, but their results showed, that changes in vocal quality were an early bulbar speech sign. Ten years later, Yanusova *et al.* presented a protocol for assessment of bulbar dysfunction and predicting progression of the disease based on the same four subsystems: respiratory, phonatory, resonatory and articulatory. Like other advanced protocols it is not used as a standard, because their responsiveness to bulbar dysfunction do not appear to be satisfactory. In 2013, Green, *et al.* (2013) was still looking for a standardized, usable diagnostic procedure for assessing bulbar dysfunction in ALS. It shall objectively assess bulbar deterioration, that leads to impairment of speech and swallowing functions. They tried to introduce the procedure based on assessing four speech subsystems: respiratory, phonatory, articulatory and resonatory. It shows, that MND, namely bulbar dysfunction, is not easy to be adequately and objectively assessed, even though bulbar symptoms significantly change the quality of patients' life, and what is more, shorten their lives (*Green et al. 2013*).

Conclusions

Voice and speech disorders can be predominant symptom in early stages of motor neuron disease. Changes can be of a different type and include dysphonia or bulbar symptoms such as dysarthria. Changes in acoustic analysis of the laryngeal tone with the MDVP method observed in our Patients show significant prevalence of functional character. Observed glottic sulcus, which occur as a result of atrophic changes in vocal muscle, is the laryngeal symptom of motor neuron disease.

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