Speech disorders in Multiple System Atrophy of Parkinson Type

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Abstract: Multiple system atrophy (MSA) is characterized clinically by the combination of Parkinsonian, pyramidal, cerebellar, and autonomic symptoms. If parkinsonism predominates in the clinical picture of the disease, the condition is named striatonigral degeneration or MSA type P (P stands for parkinsonism).

A 67 years' old woman with a previous clinical diagnosis of striatonigral degeneration and a five year history of the symptoms had initially been misdiagnosed as suffering from Parkinson's disease. However, she did not respond well to levodopa and soon developed severe dysarthria. The neurological features, helpful in differentiating MSA type P from other extrapiramidal disorders, included falling, dysarthria and dysphonia, respiratory stridor, hyperreflexia and ataxia. Cerebellar signs were well manifested, whereas autonomic symptoms were less severe.

Besides neurological and imaging study, a specially designed Speech Efficiency Test for dysarthria was performed. It revealed numerous speech deficiencies: a flat Fo contour, slow speech pace and a tendency to divide words into syllables. Vowels were centralized and reduced, obstruents articulated with impeded precision, and nasals produced with irregular soft palate timing. Glottal activity was characterized by lowered Fo and breathy phonation.

Key words: dysarthria, multiple system atrophy

INTRODUCTION

First coined by Graham and Oppenheimer [1], multiple system atrophy (MSA) describes a syndrome with features overlapping with Shy-Drager syndrome, striatonigral degeneration, and olivopontocerebellar atrophy. The less specific term «multiple system degeneration» refers to any and all of the primary neuronal degenerations [2]. Recently, MSA is divided into tree major groups: MSA type P (where parkinsonism predominates in the clinical picture), MSA type A (autonomic dysfunction predominates) and MSA type C (cerebellar dysfunction predominates).

MSA is characterized clinically by the combination of parkinsonian, pyramidal, cerebellar, and autonomic symptoms. Quinn et al. [2] described 188 pathologically proved cases of MSA, but only in 28% of patients all four systems were involved; 18% patients had a combination of parkinsonism, pyramidal, and autonomic disorders; 11% had parkinsonian, cerebellar, and autonomic symptoms; another 11% had parkinsonism and dysautonomomia; 10% had only parkinsonism, and parkinsonism was absent in 11% of all patients.

The onset of MSA takes place typically between 40 and 70 years of age. The spectrum of pathologic changes includes: cell loss and gliosis in the striatum (caudate and putamen), substantia nigra, locus ceruleus, inferior olives, pontine nuclei, dorsal vagal nuclei, Purkinje cells of the cerebellum, and the intermediolateral cell columns of the spinal cord. Involvement of at least three of these areas, including putamen and substantia nigra, is required for the neuropathologic diagnosis of MSA [2]. The presence of glial cytoplasmic inclusions, particularly in the oligodendrocytes, was ascertained in all autopsied brains of patients with Shy-Drager syndrome (SDS), striatonigral degeneration (SND), and olivopontocerebellar atrophy (OPCA), but not in the brains in the control group. This finding is a strong argument in support of the notion that these three disorders should be regarded as variants of the same disease entity, namely MSA [3].

The variable clinical and pathologic expression, however, suggests that MSA is not necessarily a single etiologic entity. Therefore, until a disease-specific marker is identified, the apparent distinction between the different disorders will continue to be blurred and spurious, and any attempts to separate these disorders must rely on traditional descriptions.

Although it is difficult to clearly differentiate the three types of MSA by clinical, if not also pathologic, criteria, it is widely accepted that MSA represents a disorder or a group of disorders distinct from Parkinson's disease. In contrast to the latter, which is inherited in at least 15% of cases, MSA occurs sporadically. Therefore, non-genetic etiology is most likely responsible for MSA and its three major subcategories.

STRIATONIGRAL DEGENERATION (SND) OR MULTIPLE SYSTEM ATROPHY PARKINSON TYPE (MSA - P)

According to Fearnley et al. [3], features helpful in differentiating SND from other parkinsonian disorders included an early onset, severe dysarthria and dysphonia, excessive snoring and sleep apnea, respiratory stridor, hyperreflexia and extensor plantar responses. Cerebellar or piramidal signs may occur as well, whereas autonomic symptoms are rare. Duration of illness ranged from 3 to 8 years, and no difference in survival was seen in levodopa responders as compared with non-responders.

At autopsy, the putamen is most prominently affected with neuronal cell loss and deposition of iron, producing brownish pigmentation [4]. There is also degeneration of the substantia nigra, and putamenal degeneration correlates with substanlia nigra cell drop-out. Lewy bodies or neurofibrillary tangles are not common. Goto et al. [5] noted selective degeneration of the metenkephalin-containing neurons in the putamen and globus pallidus, with relative preservation of the caudate nucleus. Previous studies report low levels of dopamine and increased dopamine beta-hydroxylase activity in the midbrain. Recently, vasomotor impairment in patients with SND was attributed to selective loss of tyrosine hydroxylase-immunoreactive neurons in the Al and A2 regions of the medulla oblongata [6].

Neuroimaging usually does not help to differentiate SND, not only from different form of MSA, but also from PD. Neuroimaging, specifically designed to assess putamenal integrity, may be helpful in differentiating this disease from Parkinson's disease and in predicting levodopa response [7]. PET scanning revealed decreased striatal and frontal lobe metabolism [8].

Striatonigral Degeneration is difficult to diagnose mostly because of the similarity of symptoms characteristic of Parkinson disease. In Quin's review of 10 patients, ranging in age from 47 to 50 years, with autopsy-proved SND, five were misdiagnosed as Parkinson disease, largely because of good response to levodopa. Dysarthria is believed to be a symptom characteristic for SND when its onset is early in the clinical picture of the disease.

THE GOAL OF THE STUDY

The diagnostic difficulties described above constituted the motivation to determine the dysarthria profile and an acoustic description of its features in the speech of a patient with clinically diagnosed SND. The goal of our study is to provide precise quantitative description of speech disorders present in the clinical picture of SND. These speech disorders are important because they appear relatively early in the natural history of the disease, and can be crucial in formulating a discriminative/differential diagnosis. The application of an original speech efficiency test can allow a better and more precise differentiation between SNA and PD. We hope that the method and procedure we suggest will be helpful in establishing the diagnosis with a view that in future it may develop to a comprehensive diagnostic tool.

PATIENT

In a recent case report we described a 67 years old woman with a clinical diagnosis of striatonigral degeneration and a five year history of the symptoms. She had initially been misdiagnosed as having Parkinson's disease, largely because of rigidity, bradykinesia and speech disorders. She did not, however, respond well to levodopa and, within a few months, developed severe dysarthria. The features helpful in differentiating SND from other extrapiramidal disorders included episodes of falling early in the history, severe dysarthria and dysphonia, respiratory stridor, hyperreflexia and ataxia. Cerebellar signs were present as well, whereas autonomic symptoms were less severe. As it is difficult to establish diagnosis during patient's life time and, in effect, the diagnosis is tentative, we decided to employ an extension to cover a study of speech disorders which are regarded as characteristic of SND and have been noted in the patient.

MATERIAL AND METHOD

Samples of the patient's speech were recorded on a digital video camera while being interviewed by a physician. The interview was carried out according to a scenario determined by the Speech Efficiency Test (SET) that had been worked out by a linguist-phonetician in cooperation with a clinical neurologist.

SET consisted of six sections, four of which concerned with assessing the accuracy of articulation of sound segments, and the others, with the correct rendering of suprasegmentals (stress and rhythm in words and sentences, intonation). Taking into account the complex structures of possible syllable onsets in Polish, the articulation efficiency in the production of segments was tested in word initial position in the stressed penultimate syllables of bisyllabic words. The word lists contained word-initial vowels (5 items), and voiced/voiceless consonants that differed in place of articulation (28 items). The lists of clusters included all frequently used combinations of place and manner of articulation and voicing, with 31 items of CC-clusters, 26 items of and CCC-clusters. The rendering of accent and rhythm was studied at word level on 15 four, five and six-syllabic words. As further extension of the study of the realization of suprasementals, SET contained three subsets of sentences. Subset A, consisting of five sentences of gradually increasing length, was used to assess the patient's ability to maintain articulation effort and produce spontaneous sentence accentuation. It also tested respiratory speech efficiency. In Subset B, a short conversation was included; it consisted in 4 turns of questions meant to assess the speaker's control of questioning intonation. Ability to superpose contrastive stress was examined in three focal variants of an 8-word sentence contained in Subset C.

RESULTS

At the suprasegmental level, speech was monotonous, with a flat F0 contour and without normal pitch variability; in comparison to normal articulation, the pitch range was 2,7 times narrower (Control: mean pitch 232, pitch range: 168-314; Patient: mean pitch 149, pitch range: 115-169). Glottal activity was implemented in an untypical way: F0 was lowered
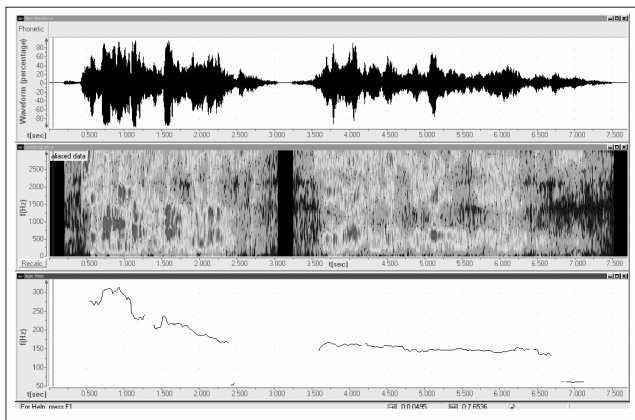
Figure 1 Pitch-tracking in control (left) and MSA-attected (right) utterances
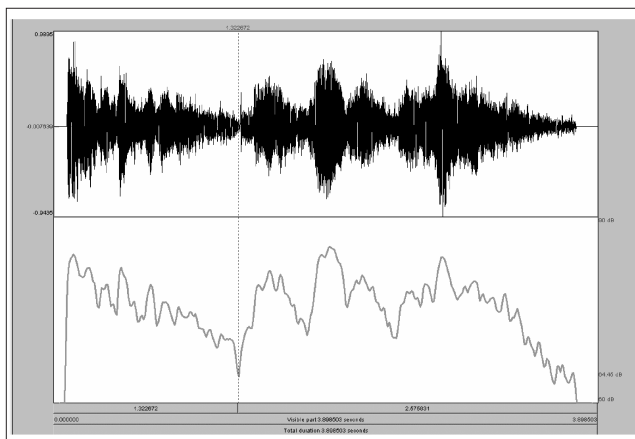*Przychodzę do poradni neurologicznej*

Figure 2 Duration and intensity in the control and pathological utterances
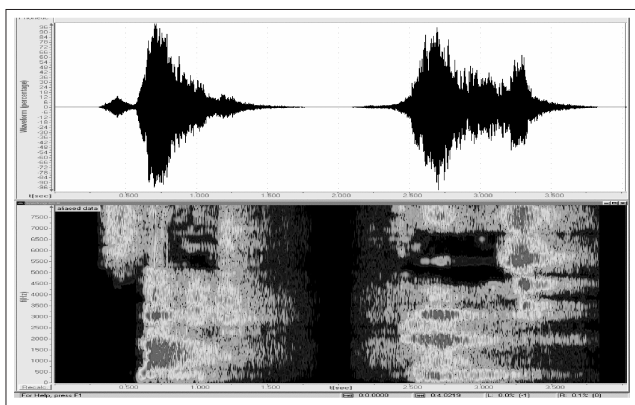*antydepresyjny*

Figure 3 Presence (left) vs. absence (right) of /s/ in a cluster in, respectively, in the control and pathological utterance *splamić*

and the phonation was breathy. Consider the pitch tracking shown in Fig. 1:

As was measured and calculated on the display, in the pathological utterance, the speech pace was 67% slower (7,6 sounds/sec) than in the control recording (11,3 sounds/sec.). Moreover, there was a tendency to divide words into syllables of relatively invariable prominence. The patient distorted the intensity pattern of word accentuation, compensating it by increasing the duration of the accented vowel; this is illustrated in Fig. 2:

Consonants, especially the apical obstruents, were articulated with impeded precision; in clusters, the initial /s/ was deleted; this is shown in Fig. 3:

Additionally, the nasal consonants were produced with irregular soft palate timing.

Word-initial stressed vowels did not display significant pathological variability, while the peripherally articulated /i/, /u/ and /a/ were centralized; in numerous cases word final non-front vowels were reduced to a *schwa*.

DISCUSSION

Dysarthria and dysphagia are known to occur in parkinsonian syndromes such as Parkinson disease (PD), dementia with Lewy bodies (DLB), corticobasal degeneration (CBD), multiple system atrophy (MSA), and progressive supranuclear palsy (PSP). Differences in the clinical and phonological picture of dysarthria in the diseases mentioned above have been noticed in professional literature. Clear-cut differentiation however, based on phonological and neurological methodology is still missing. To study the differences in the evolution of dysarthria and dysphagia in postmortem-confirmed parkinsonian disorders, Muller et al. [9] analysed eighty-three pathologically confirmed cases (PD, n = 17; MSA, n = 15; DLB, n = 14; PSP, n =24; and CBD, n = 13). His research was designed as a multicenter clinicopathological study and organized by the National Institute of Neurological Disorders and Stroke, USA. Cases with sufficient clinicopathological documentation were, for the purpose of the study, selected from research and neuropathological files of 7 medical centers in 4 countries (Austria, France, England, and the United States). Median dysarthria latencies were short in MSA (24months each) and long in PD (84 months). Similar median dysphagia latencies were intermediate in MSA (67 months) and long in PD (130 months). Authors suggested that dysarthria is relatively early symptom among a cluster of symptoms in fully developed MSA.

Dysarthria or dysphagia within 1 year of the onset of the disease was a distinguishing feature for atypical parkinsonian disorders, as compared to typical Parkinson disease but it failed to distinguish further the different forms of parkinsonism. Evaluation and adequate treatment of patients with parkinsonism who complain of dysarthria and dysphagia even without a complete clinical symptomathology might prevent or delay complications such as aspiration pneumonia, which in turn may improve the quality of life and increase the survival time [9].

Knopp et al. [10] focused in a more detailed way on MSA speech disturbances. They studied five MSA patients with a mean age of 51.2 years. Each patient was subjected to a neurological and a specific speech and voice assessment. The latter consisted of the following: clinical interview, myofunctional examination, and perceptual speech evaluation. Speech and voice complaints occurred at an average time of 1.1 year after the onset of the motor symptomatology. All MSA patients had the mixed type of dysarthria, where the hypokinetic, ataxic and spastic components were seen in each of the patients, although the hypokinetic component predominated among the others.

The authors stressed the fact that their results were different from what is commonly seen in Parkinson's disease in which the hypokinetic component is the only abnormal finding. The

authors suggested that specific speech and voice assessment is important to establish the diagnosis and to choose the best management of MSA patients.

Similar to the above mentioned study, Klunin et al. [11] characterized the dysarthria in patients with MSA. Their study, however, was more informative and easier comparable for the present authors from the methodological point of view. All patients examined by Klunin et al. [11] had dysarthria with combinations of hypokinesia, ataxia, or spasticity. Thirty-two patients had all 3 components, 13 had 2 components, and 1 had only 1 component. In most patients, the hypokinetic components were the most severe; they predominated in 22 patients (48%), whereas the ataxic components, in 16 (35%); the spastic components predominated in 5 (11%). In 1 patient (2%), the hypokinetic and spastic components were equal and greater than the ataxic components, and in 1 patient (2%), the hypokinetic and ataxic components were equal and greater than the spastic components. One patient (2%) had only the ataxic dysarthria. The predominant type of dysarthria corresponded well to the subtype of MSA studied here. They concluded with the finding that the mixed type dysarthria with combinations of hypokinetic, ataxic, and spastic components is consistent with both the overall clinical and the neuropathologic changes in MSA. As far as the dysarthria profile is concerned, our findings were in general consistent with those in the Klunin et al. [11] study.

The aim of our study was to analyze one of the MSA examples of speech disorders referred previously to as dysarthria. In the profile of dysarthria, prosody problems dominated such as narrowing and leveling of pitch range. It was characteristic of hypertonic dysarthria. On top of it, word initial clusters of two or three consonants were considerably reduced in our patient. Moreover, phonation was breathy, with air leaking through glottis, which are problems characteristic of flaccid dysarthria. Thus, we concluded that mixed, hypertonic-flaccid dysarthria is an early and characteristic sign of striatonigral degeneration (MSA type P).

We can suggest as well that motor speech examination can provide helpful information in evaluating patients with possible MSA.

COMMENTS

The speech disorder in the patient concerned the realization of speech, commonly referred to as dysarthria. In the studied pronunciation profile, prosody problems dominated such as narrowing and leveling of pitch range. Therefore, the disorder resembled hypertonic dysarthria. Word initial clusters of two or three consonants were considerably reduced, with the first element elided. This articulation problem is not characteristic of hypertonic dysarthria. Phonation was breathy, with air leaking through not fully abducted glottis. The latter problems are symptoms of flaccid dysarthria. Thus, the case study concerned mixed dysarthria as characteristic of MSA type P.

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