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UNIMORE
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ANALYSIS OF A PECULIAR MOTOR SPEECH DISORDER IN A CASE OF PROBABLE PROGRESSIVE SUPRANUCLEAR PALSY.

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Background: Dysarthria is an early clinical manifestation of atypical parkinsonian syndromes (APS) such as progressive supranuclear palsy (PSP) and multiple system atrophy (MSA). In APS, the predominant dysarthric pattern is the hypokinetic type, followed by the ataxic and spastic patterns. The majority of patients show variable combinations of these components, outlining a condition of “mixed dysarthria”.

Case report: A 81 year-old man with a 4 years history of progressive speech slowing, developed during a year dysarthria, dysphagia, postural instability with frequent falls, pathological laughter and crying. Neurological examination showed vertical supranuclear gaze palsy, frontal release signs, a positive applause sign, pseudobulbar laughing and crying, retrocollis, symmetrical hypokinetic-rigid parkinsonism, diffuse hyperreflexia and bilateral positive Hoffmann's sign. Perceptual analysis of spontaneous speech and reading revealed severe intelligibility impairment, short rushes of speech, strained-strangled voice and imprecise articulation. Acoustic analysis of sustained vocal phonation and spontaneous speech was performed using PRAAT software showing high fundamental frequency and uncontrolled alterations in voice pitch with excessive pitch fluctuations. Brain MRI revealed mesencephalic and cortical atrophy. Genetic testing for C9orf72, TDP43 was negative. According to current diagnostic criteria, a diagnosis of probable PSP was made and a levodopa therapy was started, without significant improvement.

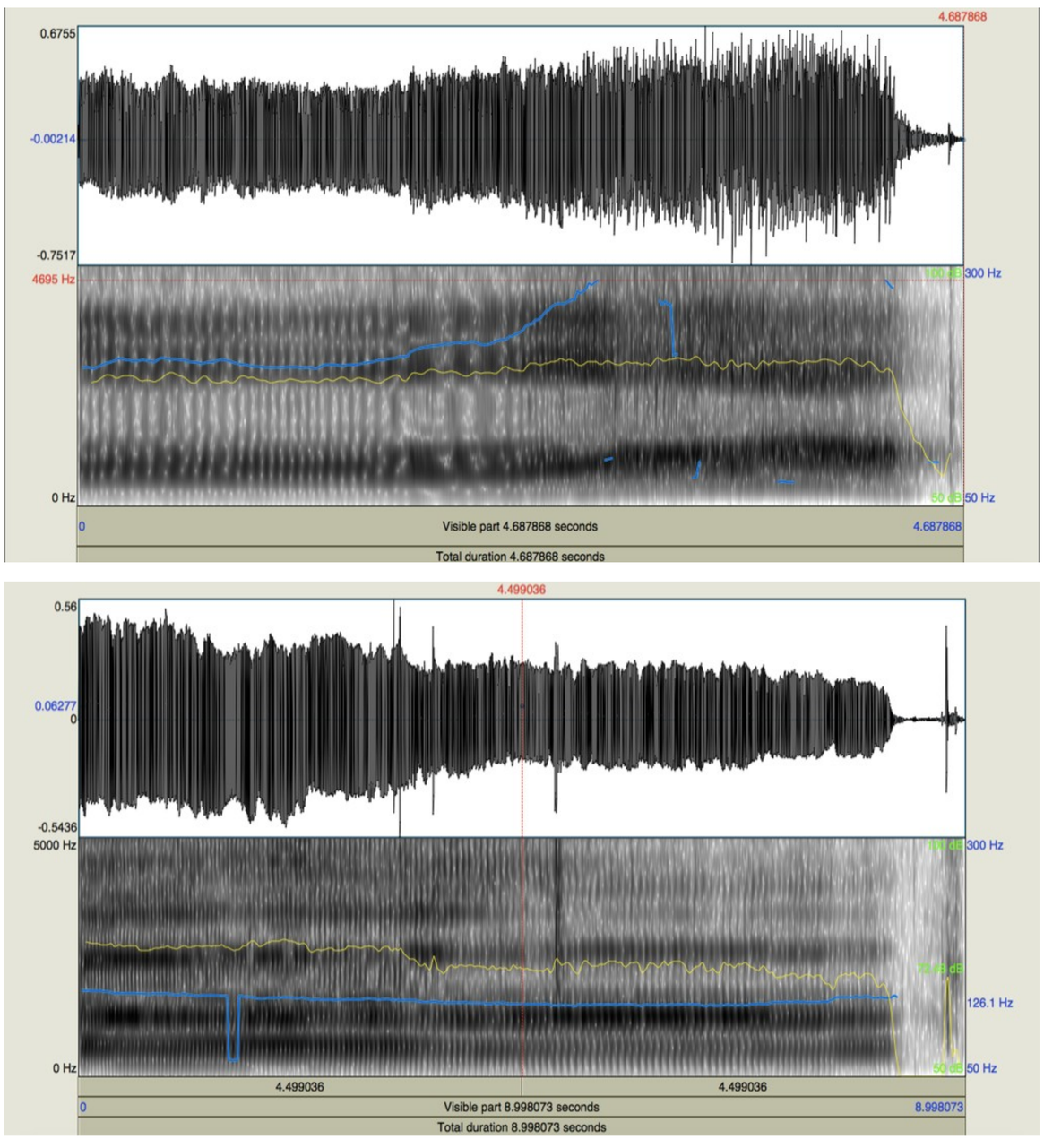

Fig 1: Acoustic analysis of sustained vocal phonation (vocal “A”) in our patient (top) and in classical PSP (bottom). The blue line shows the variations of voice pitch, the yellow one is related to the vocal intensity.

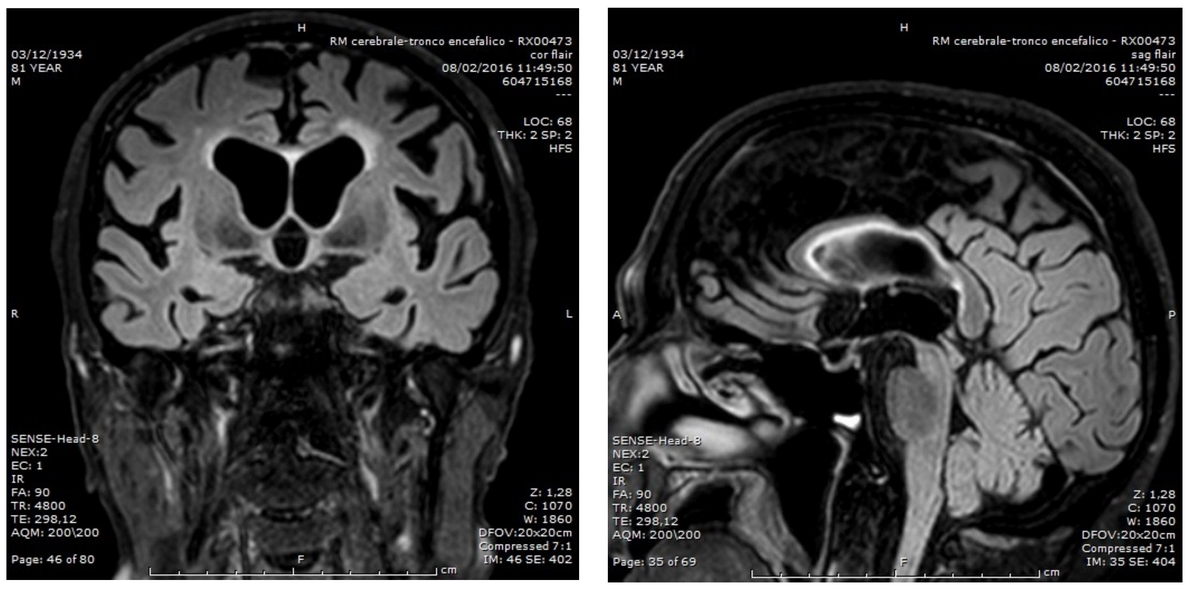

Fig 2: FLAIR brain MRI revealed cerebral atrophy and ventricular enlargement. Sagittal sequence shows mesencephalic atrophy.

Conclusion: Speech and language disorders in PSP could vary from “mixed dysarthria” to pure apraxia of speech or progressive non-fluent aphasia. Literature data consider excess pitch fluctuations as a distinctive feature of ataxic dysarthria, more common in MSA patients. In our case, the perceptual and acoustic speech alterations move towards a spastic-ataxic dysarthria that is rarely seen in PSP. This case could therefore represent an atypical and rare clinical pattern of dysarthria in PSP.

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