A PROGRESSIVE SPEECH DISORDER: A CHALLENGING CASE

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INTRODUCTION

The case study we bring has been pondered for a long time. The patient was referred to our Unit claiming to have been suffering from a speech disorder for at least two years. The verbal impairment consisted of dysprosody, articulatory difficulties, conduits d'approche and occasional phonological paraphasias. On the other hand, no morpho-syntaxis or comprehension difficulties were observed. The speech disorder progressively worsened, leading the patient to resort to using only pantomime and written word in a year's time. Our objective is now to discuss a probable diagnosis on the basis of available data, symptom progression and current criteria in the literature.

CASE REPORT

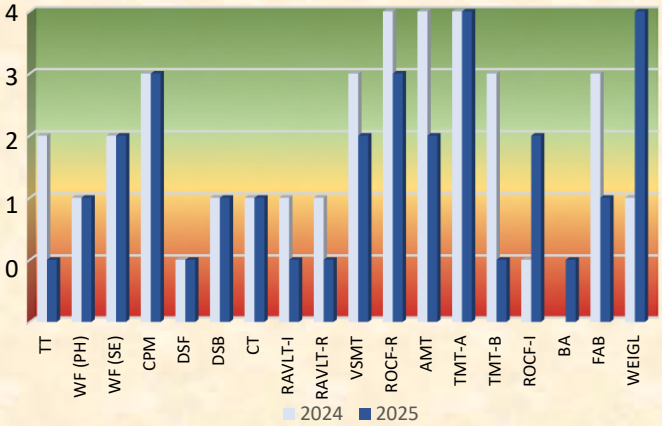
The patient is a 69-year-old man, is right-handed, still works as an architect and suffers from hypertensive cardiopathy and bilateral carotidal stenosis <30%. MRI imaging showed a moderate blood vessel disfunction and cortical and subcortical atrophy. PET scan showed a mesial temporal hypometabolism of the right hemisphere. Blood tests and biomarker analysis exclude AD-like neurodegeneration.

Neurological exams corroborated the diagnosis of a speech disorder, in the absence of other specific signs.

Baseline neuropsychological testing showed dysprosody, frequent phono-articulatory distortions, some phonemic paraphasias, occasional anomies and conduites d'approche. Despite this, the patient could still communicate effectively. Results showed borderline phonemic fluency and partially compromised verbal memory, alongside dubious behavioral issues (table 1).

One year since, a great worsening of the patient's speech capabilities was observed. The patient resorted to writing and pantomime as the only form of communication. The comprehension of the patient started to be questioned. MRI stayed the same, and neuropsychological testing confirmed a marked decline in the patient's ability to communicate verbally: all verbal tasks needed to be completed by writing. A significant comprehension deficit was now confirmed as well as some attentional difficulties and a slight worsening of long-term memory (table 1). A second PET scan showed another reduction of cortical activity in the frontal and right temporal-parietal regions.

PE



TT: Token Test (Spinnler e Tognoni, 1987); WF (PH): Word Fluency (M.D.B. 1996); WF (SE): Word Fluency (M.D.B. 1996); CPM: Colored Progressive Matrices (M.D.B. 1996); DSF: Digit Span Forward (Monaco et al., 2013); DSB: Digit Span Backward (Monaco et al., 2013); CT: Corsi Block Tapping Test (Monaco et al., 2013); RAVLT-I: Rey-Auditory Verbal Learning Test - Immediate (M.D.B. 1996); RAVLT-R: Rey-Auditory Verbal Learning Test - Recall (M.D.B. 1996); VSMT: Verbal Short Memory Test (Novelli et al., 1986); ROCF-R: Rey-Osterrieth Complex Figure - Recall (Caffarra et al., 2002); AMT: The Attentive Matrices Test (Spinnler e Tognoni, 1987); TMT-A: Trail Making Test - A (Giovagnoli et al., 1996); TMT-B: Trail Making Test - B (Giovagnoli et al., 1996); ROCF-I: Rey-Osterrieth Complex Figure - Immediate (Caffarra et al., 2002); BA: Buccofacial apraxia (De Renzi et al., 1966); FAB: Frontal Assessment Battery (Aiello et al., 2021); WEIGL: Weigl's Sorting Test (Spinnler e Tognoni, 1987).

Table 1

TEST	2024	2025
Naming	Borderline	Adequate
TT	29.25	21
WF (PH)	18.5	17.5
WF (SE)	35	33.91
CPM	29.4	30.4
DSF	3.99	3.99
DSB	2.87	2.87
CT	3.94	3.94
RAVLT-I	29.3	21.3
RAVLT-D	4.8	2.8
VSMT	15.5	11
ROCF-R	15.5	13
AMT	48.75	39.75
TMT-A	42	41
TMT-B	118	-
ROCF-I	27.5	30.5
BA		5
FAB (total)	15.47	13.47
WEIGL	5.5	15

DISCUSSION AND CONCLUSIONS

What led us to hypothesize a PPA diagnosis were the age of onset, the progression of the language deficit, the neuroimaging data and the preservation of everyday functionally and job productivity. We must take into consideration that, along with atrophy, both a chronic multi-infarct encephalopathy and significant leukoaraiosis have been found.

These conditions could have worsened the pre-existing neurodegeneration, leading to a faster decline in speech ability. By taking into account all APP subtypes, the logopenic variant was excluded: fistly, literature consensus confirms a link between logopenic PPA and AD, whereas biomarkers were negative; secondly, the patient's clinical profile does not fall in line with both core symptoms necessary for a diagnosis. On the other hand, the semantic variant was rejected too: while the speech disorder is advancing, the patient's naming ability is, in fact, maintained, even if almost exclusively in written form.

All the diagnostic criteria for the nonfluent agrammatic variant would seem to he satisfied, but there happens to be a large overlap with the criteria for primary progressive apraxia of speech: in fact, word finding, written language, grammar and comprehension were all maintained. The slight memory deficit would lead us to exclude both diagnoses but it does not present itself in all the administered tests.

We must also keep in mind that the patient suffers from cerebrovascular disease and does not report any difficulty in work or everyday life. Considering all available data, we could expect to find left hemisphere lesions, but neuroimaging shows instead damage on the right side of the brain. This does not fall in line with either diagnosis hypothesis.

Rare as it may be, we can't refuse with certainty the possibility of a right brain language lateralization, even though the patient is right-handed.

This case demonstrates the broad heterogeneity of progressive language disorders. The great overlap in clinical features makes subtype categorization quite challenging, and we must not forget the significant interference caused by vascular factors. The diagnosis of the condition will inevitably have to be based on observations of its development, in the hopes of identifying its trajectory.

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