Atypical presentations of non-fluent neurodegenerative Aphasia

N. MOMETTO[1], R. PODA[2], S. ABU RUMEILEH[1], A. BARTOLETTI-STELLA[1], M. STANZANI MASERATI[2], P. CORTELLI[1,2], R. LIGUORI[1,2] , S. CAPELLARI[1,2]

[1]Department of Biomedical and NeuroMotor Sciences (DIBINEM), Alma Mater Studiorum, University of Bologna, Bologna, Italy.
[2]IRCCS, Institute of Neurological Sciences, Bellaria Hospital, Bologna, Italy.

Introduction

The Primary Progressive aphasias are categorized according to the Gorno–Tempini classification (2011) that recognizes the semantic, non fluent/agrammatical and logopenic variants. However, 15-30% of aphasias remain unclassified (Vanderberge R, 2016). The aim of this work is to compare three cases of atypical presentation of non-fluent aphasia. We focused our attention on the applicability of the classical criteria of Primary Progressive Aphasia subtypes in these particular situations and the limits of the actual classification.

Case 1

Male, second-grade familiarity for bvFTD.
At 52 years old progressive isolation, depressed mood and progressive speech impairment, with difficulty in verbal expression, short, simple phrases without grammatic structure, rich in phonological and semantical paraphasias.
MRI: left fronto-temporal atrophy
CSF: t-tau 237 pg/mL, p-tau 27 pg/mL, β-Amyloid 787 pg/mL.

NPS: MMSEc 30/30, BBDM: +0,16. Severely compromised spontaneous speech with intact comprehension and repetition. Defects in verbal test of long-term memory and phonemic fluency compatible with transcortical motor aphasia.

Case 2

Male, no familiarity. From 70 years old, behavioral changes: apathy, depression, obsessions, poor empathy. From 72 years old, progressive slowdown of speech, denomination impairment, difficulty of word-finding, use of circumlocutions.

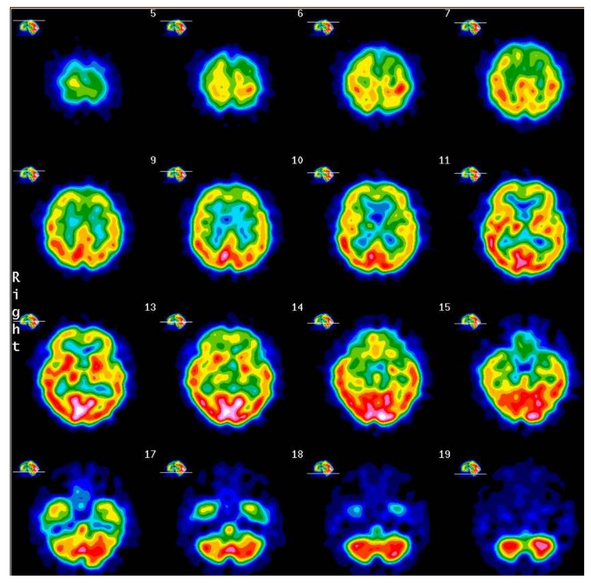
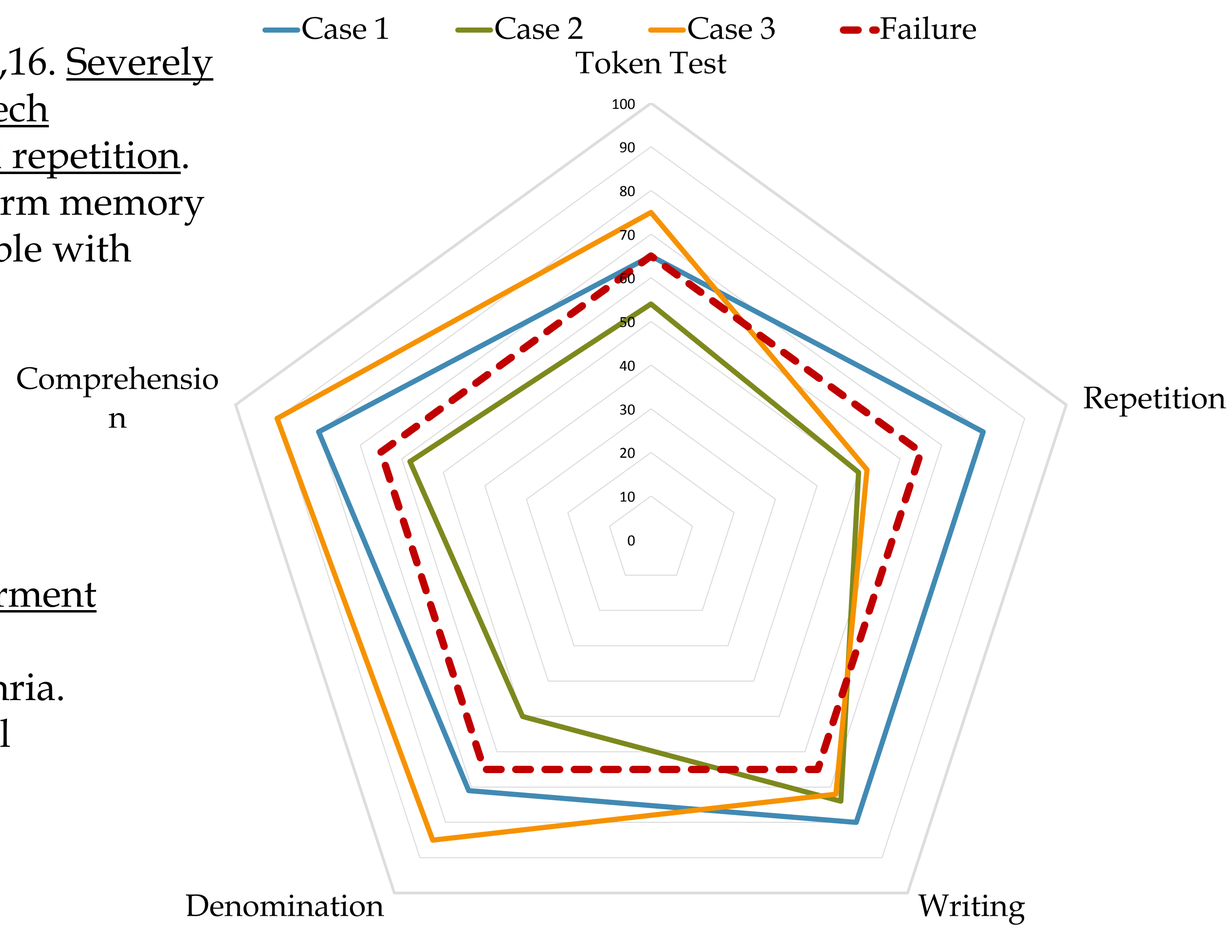


Figure 1: Perfusion SPECT of the Case 2

EON: Disprosodic, automatic language, rich in stereotypies, perseveration and automatisms.
MRI: Midbrain and left fronto-temporo-parietal atrophy.
Perfusion SPECT: Left frontal and temporo-parietal hypoperfusion. Modest right frontal lobe hypoperfusion.

NPS: MMSEc 25.7/30, BBDM +0.84. Long-term and short-term verbal memory impairment and low phonological fluency.



Graphic 1: Results of Aachener Aphasia Test in the three cases

Case 3

Male, starting from 68 years old progressive and selective impairment of articulation of speech.
EON: Speech hesitation, dysarthria.
CT (2 yrs): Left temporo-parietal atrophy
FDG-PET (2 yrs): Left temporo-parietal hypometabolism.

NPS (2 yrs): Pure anarthria, with slow, disprosodic, punctuated speech. Impaired sentence repetition and reading.

MRI: Dilatation of left temporo-insular subarachnoid spaces Lateral and third ventricles enlargement.
fMRI: Left frontal activation at the listening paradigm, absence of left temporal activation on noun-based verb generation.

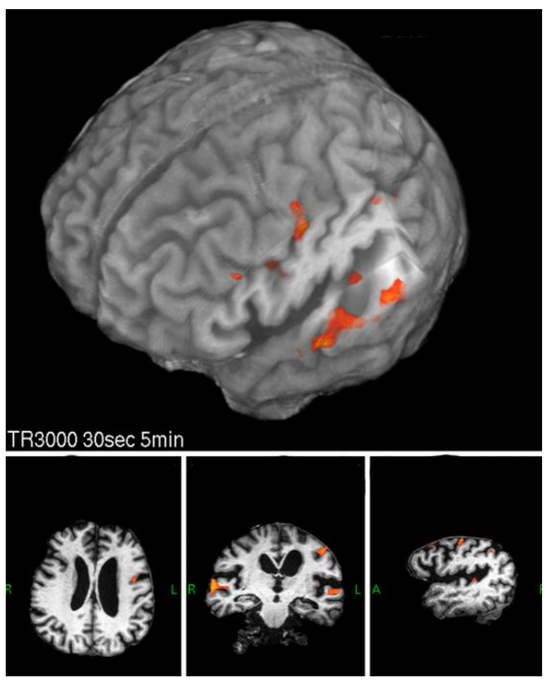


Figure 2: fMRI of the Case 3 in the noun-based verb generation paradigm

Nonfluent/Agrammatic Criteria	Case 1	Case 2	Case 3
Agrammatism	✓		
Apraxia of speech		✓	
Impaired comprehension complex sentences			
Spared single-word comprehension	✓	✓	✓
Spared object knowledge	✓	✓	✓
Imaging support	✓	✓	✓
Final diagnosis with criteria	PPA- NOS	PPA- NOS	PPA-NOS

Table 1: Application of the PPA-subtype criteria in the three cases (PPA NOS: PPA not otherwise specified)

Bibliography

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Conclusions

The three reported cases demonstrated that neurodegenerative diseases presenting with aphasia as the prominent feature can be very difficult to frame into the classification of PPA-subtypes. Because, in their extreme complexity, they can present with atypical language deficit, not fitting with any of the criteria.

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