Μη ρέουσα αφασία και λογοπενική μορφή της πρωτοπαθούς προϊούσας αφασίας: Είναι δυνατή η διάκρισή τους στην καθημερινή κλινική πράξη;

Παντελής Μαϊόβης, **MSc, PhD**
Νευρολόγος
Πανεπιστημιακός Υπότροφος
Β' Νευρολογικής Κλινικής ΑΧΕΠΑ

Μη ρέουσα/Αγραμματική παραλλαγή Πρωτοπαθούς Προϊούσας Αφασίας (αΠΠΑ)

Table 2 Diagnostic features for the nonfluent/agrammatic variant PPA

I. Clinical diagnosis of nonfluent/agrammatic variant PPA

At least one of the following core features must be present:

1. Agrammatism in language production
2. Effortful, halting speech with inconsistent speech sound errors and distortions (apraxia of speech)

At least 2 of 3 of the following other features must be present:

1. Impaired comprehension of syntactically complex sentences
2. Spared single-word comprehension
3. Spared object knowledge

2. Imaging must show one or more of the following results:

a. Predominant left posterior fronto-insular atrophy on MRI or

b. Predominant left posterior fronto-insular hypoperfusion or hypometabolism on SPECT or PET

Λογοπενική παραλλαγή Πρωτοπαθούς Προϊούσας Αφασίας (λΠΠΑ)

Table 4 Diagnostic criteria for logopenic variant PPA

I. Clinical diagnosis of logopenic variant PPA

Both of the following core features must be present:

1. Impaired single-word retrieval in spontaneous speech and naming
2. Impaired repetition of sentences and phrases

At least 3 of the following other features must be present:

1. Speech (phonologic) errors in spontaneous speech and naming
2. Spared single-word comprehension and object knowledge
3. Spared motor speech
4. Absence of frank agrammatism

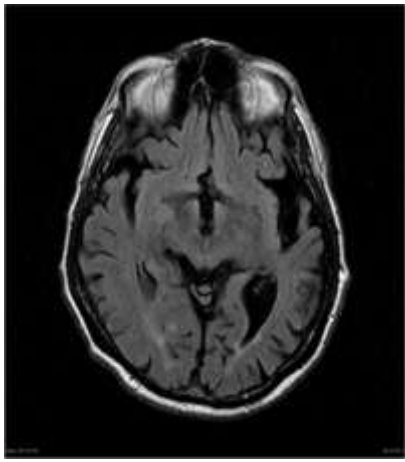
II. Imaging-supported logopenic variant diagnosis

Both criteria must be present:

1. Clinical diagnosis of logopenic variant PPA
2. Imaging must show at least one of the following results:

a. Predominant left posterior perisylvian or parietal atrophy on MRI

b. Predominant left posterior perisylvian or parietal hypoperfusion or hypometabolism on SPECT or PET



- Μη ρέουσα αφασία και λογοπενική μορφή της πρωτοπαθούς προϊούσας αφασίας: Είναι δυνατή η διάκρισή τους στην καθημερινή κλινική πράξη

  ΟΧΙ

  (τουλάχιστον όχι τόσο εύκολα…)

Τόσο διαφορετικές κλινικά;

λΠΠΑ	αΠΠΑ
• Λόγος αργός με συχνές παύσεις • Παραφασίες (φωνημικές) • Κατανόηση λέξεων άθικτη • Αγραμματισμοί (στις μακριές προτάσεις λόγω διαταραχής στην ακουστική λεκτική πρόσφατη μνήμη)	• Λόγος μη ρέων, με μακριές παύσεις • Παραφασίες (φωνημικές-AOS) • Κατανόηση λέξεων άθικτη • Αγραμματισμοί (frank agrammatism)

‘Σκληρές’ διαφορές-agrammatism

- Αγραμματισμοί στην αΠΠΑ: απαραίτητο στοιχείο για την κλινική διάγνωση ΑΛΛΑ
- Graham et al 2016 :Lack of Frank Agrammatism in the Nonfluent Agrammatic Variant of Primary Progressive Aphasia Dement Geriatr Cogn Dis Extra
- Πρώιμα στάδια αΠΠΑ;
- Σύγχυση με λΠΠΑ

‘Σκληρές’ διαφορές-AOS

- Ash et al.: Speech errors in progressive non-fluent aphasia. Brain Lang 2010, 113:13–20.
- speech errors in navPPA mainly arise within the language system and are not caused by a motor impairment
- AOS περίπου 20%

‘Σκληρές’ διαφορές-επανάληψη

- Σε πρώιμα στάδια λΠΠΑ μπορεί να μην υπάρχει τόσο σοβαρή δ/χη
- Δ/χες στην επανάληψη προτάσεων υπάρχουν και στην αΠΠΑ (για διαφορετικό λόγο)

And now (3·6 s) he's yoloding or yaw-yawning
And then here (2·4 s) the jar (3·4 s) window up went (3·5 s) yelling out

And it (10·8 s) he falling out of the window

And uh (3·8 s) this page here the (4·1 s) the boy
Is (3·5 s) int- col the j- jar broken
And (6·1 s) d- doadge... dog is have licking
And (2·8 s) he's happay.

Examiner: Tell me about what you do each day.

Patient: All right, uh, I get up (laughs), and uh, I keep a tidy house. My children can't believe why, how, but I, it doesn't get dirty, it doesn't do anything (laughs). So that's fine. It doesn't, it's not too much work. But I read a lot, and uh, I look at television here and there, and uh (1) then, I do go out. I like to go and walk once a day, and uh I've just started again this week uh because I didn't uh walk. I don't like walking in the hot hot weather, and this is the nice kind of weather to walk. So I go out for my forty-five minute walk and (2), and I'll (1)… I don't do very much, really, uh, I go visiting and have an odd person in to have a cup of tea and uh, then on the weekends I go out with my family and (3) (patient nods and drinks some tea, indicating she has finished her response).

λΠΠΑ: υπάρχει;

- Sajjadi et al Primary progressive aphasia. A tale of two syndromes and the rest. Neurology 2012

- 28.3%, 26.1%, and 4.3% fitted semantic, nonfluent/agrammatic, and logopenic categories respectively, and 41.3% did not fulfill the diagnostic recommendations for any of the 3 proposed variants.

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Short communication

The frontotemporal dementias in a tertiary referral center: Classification and demographic characteristics in a series of 232 cases

Panagiotis Ioannidis, Elina Konstantinopoulou, Pantelis Maiovis *, Dimitris Karacostas

B' Department of Neurology, AHEPA University Hospital, Thessaloniki, Greece

Results: From the 232 patients, 111 (47.8%) were diagnosed with bvFTD, 56 (24.1%) with naPPA, 21 (9.1%) with svPPA, 6 (2.6%) with lvPPA, 20 (8.6%) with CBD or PSP and 18 (7.8%) with rvFTD. 44% of the patients were under 65 years old at onset of symptoms, while only 4.3% reported family history of dementia. FTLD subgroups did not differ with respect to demographic characteristics, but early onset cases had higher educational level.

Συμπερασματικά…

- λΠΠΑ: παραλλαγή της ΠΠΑ με διακριτά κλινικά κι απεικονιστικά κριτήρια
- κλινική πράξη;
- Ηλικιωμένος, χαμηλού μορφωτικού επιπέδου ασθενής
- Πρώιμα στάδια
- Εξειδικευμένα εργαλεία