PRIMARY PROGRESSIVE APHASIA

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PURPOSE

• Review current diagnostic criteria for PPA’s
• Review qualitative/behavioral observations that are helpful to the practicing neuropsychologist
• Review an abbreviated test battery that may help in the differential diagnosis of these disorders
DISCLOSURE

• I have no financial interest in any of the tests presented today.
• Lori and Anna and the BSWH tech team are cool cats.

GOOD READS


CORE DEFINING FEATURE:

All of 1-3
1. Most prominent clinical problem is language
2. Language impairment is primary ADL difficulty
3. Aphasia is the most prominent deficit at initial phase

THE ROLE OF OTHER INVESTIGATIONS

• Imaging
  – Presence of imaging findings correlating clinical findings is supportive, but not diagnostic in and of itself
• Pathology
  – Overlaps, but does not necessarily define the clinical condition
AGRAMMATIC PPA- DIAGNOSTIC CRITERIA

• Must have one of the following
  – Agrammatism in language production
  – Effortful, halting speech with inconsistent speech sound errors and distortions (apraxia of speech)

CORE FEATURE: DESCRIPTION

• Typically short, simple phrases and omissions of function words, inflection
• Slow, labored, speech
• Articulation= planning of speech sounds
• Speech sound errors: distortions, deletions, substitutions, insertions, transpositions and they are aware of these
• Prosody disrupted
• Written production can mirror produced speech, at least in terms of grammatical errors.
AGRAMMATIC PPA- DIAGNOSTIC CRITERIA

• At least ⅔ of the other features
  – Impaired comprehension of syntactically complex sentences
  – Spared single word comprehension
  – Spared object knowledge

MORE CORE FEATURES

• Comprehension deficits only in syntactically complex situations.
• “The car that the truck hit was green.”
CORE ABSENT FEATURES

- Not better accounted for by another disorder
  - Many turn into PSP/CBGD variants, so go by initial symptom
- Not better accounted for by psychiatric disorder
- Not prominent memory, visual memory, or visuoperceptual impairment
- Not prominent initial behavioral disturbance

AGRAMMATIC PPA-NEUROANATOMY/NEUROIMAGING

“Imaging supported diagnosis”

1. Clinical diagnosis +
   a. Left posterior fronto insular
      i. Atrophy on MRI
      ii. Hypoperfusion/metabolism on SPECT/PET

AGRAMMATIFIC PPA - NEUROPATHOLOGY

“... with definite pathology”
- Clinical diagnosis +
  - Histopathology of a degenerative condition
    - Most common FTD-tau
    - Less often, FTLD-TDP pathology
  - or/and presence of a known pathogenic mutation

AGRAMMATIFIC PPA - CLINICAL ASSESSMENT

- Traditionally, we have looked at fluency and phrase length in anterior language disturbance.
- How can you quantify these?
- Can we reliably differentiate these from other sources of speech difficulty?
- The (An) answer....
THE NORTHWESTERN ANAGRAM TEST (NAT)

- Order printed words to be syntactically consistent with the action in a target picture.
- Arrows and labels so comprehension less of an issue.
- Give them the first two words of the sentence
- 10 Item short form

![Diagram of a boy and girl with arrows and labels]


WHERE TO GET IT

http://flintbox.com/public/project/19927/

- Pro's
  - Brief
  - Evidence Based
- Con's
  - Lack of psychometric properties
  - Lack of extensive normative data
NAT IN A WELL SELECTED GROUP


BRAIN/BEHAVIOR RELATIONSHIP

SEMANTIC PPA - DIAGNOSTIC CRITERIA

Must have both
- Impaired confrontation naming
- Impaired single word comprehension

MORE ABOUT CORE FEATURES

- Naming can be impaired in all the PPA's, but severe here
- Single word comprehension is poor for less familiar objects
- Poor in all modalities
- Usually general semantic loss, but can be selective (i.e. worse for people/animals than objects)
AT LEAST 3 OF:

• Impaired object knowledge, particularly for low frequency or low familiarity items
• Surface dyslexia or dysgraphia
• Spared repetition
• Spared grammar/motor speech/production

MORE ABOUT ADDITIONAL FEATURES

• Regularization errors with “irregular” words
• Usually grammatically accurate, though “paragrammatic” errors such as closed class errors
  – “I know what they are doing but I can’t think the words what they’re doing.”
SEMANTIC PPA-NEUROANATOMY/NEUROIMAGING

Anterior temporal lobes
Usually worse L>R


SEMANTIC PPA-NEUROPATHOLOGY

Usually FTLD-TDP
SEMANTIC PPA - CLINICAL ASSESSMENT

- Recommended: Items 157-192 from PPVT-4
- Also know:
  - Pyramids and palm trees
  - Sydney language battery
  - Semantic Object Recognition Test

PPVT-4 157-192 IN A WELL SELECTED GROUP

LOGOPENIC PPA - DIAGNOSTIC CRITERIA

- Must have both
  - Impaired single word retrieval in spontaneous naming and speech
  - Impaired repetition of sentences and phrases

ADDITIONAL DESCRIPTION-LOGOPENIA

- Slow rate, frequent pauses due to significant word finding
- No frank agrammatism
  - Different than PNFA who are dysprosodic + motor errors
- Naming errors are frequently phonologic in nature

LOGOPENIC PPA MORE FEATURES

At least 3 of the following:
- Phonological errors in spontaneous speech and naming
- Spared single word comprehension
- Spared motor speech
- Absence of agrammatism
MORE LOGOPENIA...

- Phonological short term memory is poor
  - Can repeat short words, not longer, ones
- Phonological paraphasias in spontaneous speech and naming
  - Errors that are well articulated

LOGOPHENIC PPA - NEUROANATOMY

- Predominent left posterior perisylvian or parietal atrophy on MRI/hyperfusion on SPECT
- Can be left temporo-parietal (posterior temporal, supramarginal, angular gyri)

LOGOPENIC PPA- NEUROPATHOLOGY

- Presence of pathology and/or known mutation
- Perhaps most commonly AD

LOGOPENIC PPA- CLINICAL ASSESSMENT

- >60% Items 157-192 from PPVT-4
- >60% NAT
- Repetition is difficult….because it's not difficult...

WELL, IT’S NOT THAT...


BRAIN/BEHAVIOR RELATIONSHIP
PUTTING THE PIECES TOGETHER...


CAVEATS

- Very well educated sample
- Need to assess in lower education populations
- Some patients are mixed
- May depend on when you see the patient.
QUESTIONS?