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Diagnosis & Treatment of Complex Cases in Speech-Language Pathology

This series is presented in partnership with Rush University Medical Center

Guest Editor: Richard Peach, PhD, CCC-SLP, BC-ANCDS
Primary Progressive Apraxia of Speech and Aphasia in a Complex Case of Neurodegenerative Disease

Presenter: Richard Peach, PhD, CCC-SLP, BC-ANCDS

Moderated by:
Amy Hansen, M.A., CCC-SLP, Managing Editor, SpeechPathology.com

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Primary Progressive Apraxia of Speech and Aphasia in a Complex Case of Neurodegenerative Disease

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Rush University Medical Center
Chicago, Illinois

Learning Objectives

• To describe the clinical presentation of progressive apraxia of speech and aphasia in a complex case of neurodegenerative disease
• To identify the speech and language characteristics of progressive apraxia of speech and aphasia
• To provide recommendations for the management of the disorder
Case Study: Pt. M

- 67 y.o. female
- Right-handed
- January, 2012: Acute onset of speech disturbance
  - Slowed speech but without articulatory difficulty
  - Reading/writing preserved
- PMHx:
  - Depression
  - Chronic hearing loss
  - Smoking (1 pack/day x 40 years)
- Social Hx:
  - 2 yrs of college
  - Retired office manager
  - Lives with daughter

Evaluations: 2012 – 2013 (OSH)

- November, 2012; May, 2013: Neurology
  - Stroke, myasthenia gravis workups negative
  - EEG normal
  - MRIs/MRAs essentially normal with exception of very mild chronic white matter vascular changes
  - ?s re: possible L > R temporal atrophy
- December, 2013: Speech-language
  - Mod-severe dysarthria
  - Poor intelligibility
  - ↓ labial, lingual strength, ROM, and coordination
  - Preserved auditory comprehension
  - Mild deficits in reading comprehension
  - Mild expressive aphasia (word-finding difficulty)
  - Mild cognitive deficit
  - No dysphagia
Evaluation: June, 2014 (RUMC)

- **Neurology consultation**
  - MMSE = 30/30
  - Significant speech fluctuation (variable slowing)
  - Normal prosody
  - Poor intelligibility
  - Paraphasic errors, esp. on confrontation naming
  - Difficulty with repetition
  - Speech for reading worse than spontaneous speech
  - Writes spontaneous sentence, unable to perform to dictation
  - No significant CN, motor, or sensory abnormalities other than dysarthria
  - Vibration minimally diminished in R toe

- **MRI Brain**
  - Minimal chronic small vessel ischemic changes within PVWM
  - No acute intracranial pathology

- **DDX: PPA versus AOS**

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**Progressive apraxia of speech as a sign of motor neuron disease (Duffy, Peach, & Strand, 2007)**

- **Level of Evidence**: Case series
- **Method**:
  - Retrospective study of 7 pts with dx of MND and AOS
    - Identified from among 80 patients with AOS due to various neurodegenerative diseases (Duffy, 2006)
    - Age: mean = 67 years; range = 48-84 years
    - History, complaints, neurological and speech-language findings documented
- **Findings**
  - Speech first symptom in 5 pts (prominent complaint in all 7)
    - 1 with personality changes
    - 1 with toe paresthesias
  - Non-verbal oral apraxia in 5 pts; later emerged in remaining 2 pts
  - Dysarthria present in all (slow rate, reduced intelligibility, monopitch & monoloudness)
    - 3 with spastic
    - 2 with mixed flaccid-spastic
    - 2 with undetermined type (later mixed)
  - Variable presence of aphasia
    - No aphasia in 3 pts; 2 with mild-mod “nonfluent” aphasia; 2 w/ abnormal performance unable to be directly attributed to aphasia
  - Short breath groups during speech, despite adequate respiratory support (4 of 7)
Case history

- Primary complaints:
  - Poor speech
  - Overall fatigue
- Both have worsened since May 2014
- Difficulty in “getting words out”
  - Per daughter: “Knows what she wants to say, but can’t say it.”
- No changes in mental status reported
Clinical Observations

• No difficulty observed swallowing small amount of liquid
  – Eats several small meals/day
  – Recent weight loss
  – GERD reported

• Mildly unsteady gait

• Fatigue (increasing throughout session)

• Uses marker board to aid in communication

Assessment - WAB-R

<table>
<thead>
<tr>
<th>Subtest</th>
<th>Score</th>
<th>Impressions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spontaneous Speech:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Information Content Fluency</td>
<td>9/10</td>
<td>Non-fluent production, reduced phrase length, incomplete sentences</td>
</tr>
<tr>
<td></td>
<td>4/10</td>
<td></td>
</tr>
<tr>
<td>Auditory Verbal Comprehension</td>
<td>7.95/10</td>
<td>Y/N responses preserved, moderately impaired sequential commands</td>
</tr>
<tr>
<td>Repetition</td>
<td>3.2/10</td>
<td>Hesitations, syllable segmentation, consonant and vowel substitutions, verbal substitutions</td>
</tr>
<tr>
<td>Naming &amp; Word finding</td>
<td>8.3/10</td>
<td>Mildly impaired object naming and word fluency Moderately reduced sentence completion</td>
</tr>
</tbody>
</table>
Assessment - WAB-R (cont.)

<table>
<thead>
<tr>
<th>Domain</th>
<th>Score</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reading</td>
<td>84/100</td>
<td>Mild-moderate impairment</td>
</tr>
<tr>
<td>Writing</td>
<td>78/100</td>
<td>Moderate impairment</td>
</tr>
<tr>
<td>Praxis</td>
<td>9.2/10</td>
<td>Mildly reduced</td>
</tr>
<tr>
<td>Raven’s Coloured Progressive Matrices</td>
<td>29/36</td>
<td>51st percentile</td>
</tr>
<tr>
<td>Aphasia Quotient (AQ)</td>
<td>64.9</td>
<td>Cutoff = 93.8</td>
</tr>
<tr>
<td>Language Quotient (LQ)</td>
<td>72.7</td>
<td></td>
</tr>
<tr>
<td>Cortical Quotient (CQ)</td>
<td>74.4</td>
<td>Cutoff = 90</td>
</tr>
</tbody>
</table>

Assessment (cont.)

- **Hearing screening**
  - Otoscopic exam
  - Pt report:
    - Difficulty in R ear
    - Uses phone on L only
  - Pure tone audiometry:
    - Did not pass 40 dB HL in either ear

- **Oral Motor Exam**
  - Gross facial symmetry at rest
  - **Mild flattening of R nasolabial fold**
  - Adequate labial, mandibular strength
  - Symmetrical tongue (at rest and upon protrusion)
  - **Mild R lingual weakness**
  - Adequate palatal symmetry and ROM
  - Gag present
  - Suck & snout absent
  - No lingual, chin fasciculations
Assessment - ABA-2

<table>
<thead>
<tr>
<th>Subtest</th>
<th>Cutoff</th>
<th>Score</th>
<th>Impressions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Increasing word length</td>
<td>1</td>
<td>8</td>
<td>Severe impairment</td>
</tr>
<tr>
<td>Repeated trials of complex words</td>
<td>28</td>
<td>7</td>
<td>Moderate impairment</td>
</tr>
<tr>
<td>Utterance time (sec) polysyllabic words</td>
<td>15</td>
<td>22</td>
<td>Mildly lengthened</td>
</tr>
<tr>
<td>Limb apraxia</td>
<td>44</td>
<td>46</td>
<td>None</td>
</tr>
<tr>
<td>Oral apraxia</td>
<td>44</td>
<td>49</td>
<td></td>
</tr>
<tr>
<td>Apraxic Speech Behaviors</td>
<td>4</td>
<td>14/15</td>
<td>Phonemic transposition errors</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Highly inconsistent errors</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Numerous and varied</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>off-topic attempts</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Abnormal prosodic features</td>
</tr>
</tbody>
</table>

Assessment – Real Time Pitch

<table>
<thead>
<tr>
<th>Measure</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>AMRs</td>
<td>/p^/=2.5/s, /t^/=2.3/s, /k^/=2.3/s</td>
</tr>
<tr>
<td></td>
<td>(Normal: /p^/=6.3/s, /t^/=6.2/s, /k^/=5.8)</td>
</tr>
<tr>
<td>SMRs</td>
<td>/p^+t^+k^/=1/sec</td>
</tr>
<tr>
<td></td>
<td>(Normal: /p^+t^+k^/=5.0/sec (SD 0.7))</td>
</tr>
<tr>
<td>Mean Fundamental Frequency</td>
<td>Automatic speech: 131.1 Hz (SD 27.1)</td>
</tr>
<tr>
<td>Mean Loudness</td>
<td>Reading: 122 Hz (SD 17.0)</td>
</tr>
<tr>
<td>Maximum Phonation Time</td>
<td>Average: 3.2 sec (Normal median= 14.4, SD 5.7)</td>
</tr>
</tbody>
</table>

- AMRs and SMRs
  - Slowed rate with irregular rhythm
  - Decreased coordination
- Maximum Phonation Time - Substantially reduced respiratory-phonatory support for speech production
Assessment – Speech

- Conversational Speech
  - Breath groups = 2-3 words
  - Short, visible inhalations
  - Marked vocal fatigue
  - Effortful, strained, hypernasal, imprecise, monotone/monopitch
- Sentence intelligibility: 53% (poor)
- Speaking rate = 35 WPM (normal = 190)
  - Rate of intelligible speech = 18.9 WPM
  - Rate of unintelligible speech = 16.9 WPM

Picture Description

Okay...father flying kite...kite...now daughters were (unintelligible)...fish her...her...and then a lot of stuff...The girl's uh...has her /s/ sting (sand) sting...and the man reading book and the woman's um...putting cup...pouring...drink...and the...and the...sailboat

Oral Reading
Impressions: Cognition

• Nonfluent aphasia:
  – Moderately reduced speech fluency, grammar, and phrase length
  – Mild impairments to auditory and reading comprehension
  – Mild naming deficits
  – Moderate writing impairment
• Nonverbal cognition: Low normal

Impressions: Speech

• Characteristics:
  – Slow rate
  – Frequent insertion of filled pauses w/in words
  – Syllable segmentation
  – Perceived sound substitutions
  – Reduced speech prosody (monotone & monopitch)
• Additional observations:
  – Hypernasality
  – Imprecise articulation
  – Strained vocal quality
  – Reduced respiratory-phonatory support
  – Poor speech intelligibility in both known and unknown contexts
Diagnosis

• Results are consistent with:
  – Primary progressive apraxia of speech & aphasia
• Co-occurring:
  – Spastic dysarthria
• Progressive AOS and aphasia can be first and only symptom of neurodegenerative disease for extended period of time
• Prognosis: guarded to poor
  – Based on suspected degenerative nature

Apraxia of speech in degenerative neurologic disease (Duffy, 2006)

• Level of Evidence: IV
• Method:
  – Retrospective review of 80 pts seen between 1985 and 2004 who had AOS not less severe than any aphasia present in which cause was degenerative
    • Average age= 69 years (range=36-86 years)
    • Speech-language difficulty was first symptom in 80%
      – Was only initial pt complaint in 56%
• Findings:
  • Dx of AOS, majority displayed: slow rate, distorted substitutions, segmentation of syllables or excess and equal stress, poorly sequenced SMRs, and ↑off-target artic. errors with ↑utterance length
  • Aphasia present in 49% (mild-mod severity, mostly non-fluent)
  • Dysarthria present in 50% (2/3 had spastic, hypokinetic, or mixed spastic-hypokinetic)
  • 77% had nonverbal oral apraxia
  • None had nonaphasic cognitive deficits worse than their AOS
  • Neurologic:
    – Grossly normal EEGs & CT scans; MRIs indicated atrophy in only L, or L>R; SPECT indicated L>R abnormalities in 48%
Apraxia of speech in degenerative neurologic disease (Duffy, 2006)

- Conclusions:
  - 44% of sample received neurological dx from the neurologist based exclusively on speech-language findings or were strongly influenced by them
    - 90% of patients with neurological dx primarily based on:
      - presence of AOS
      - diseases lateralised to L hemisphere OR
      - Conditions assoc. with prominent motor manifestations (CBD, IPD, ALS/MND)
    - AOS can be the predominant and sometimes ONLY symptom in pts for whom S/L symptoms are first manifestation of degenerative disease
      - Distinguish between PPA and primary progressive AOS and aphasia
        - Implications for management (is there a language component?)

Motor speech disorders associated with primary progressive aphasia (Duffy et al., 2014)

- AOS more strongly associated with PPA than dysarthria
- Among dysarthrias occurring with PPA, spastic and hypokinetic types occur most frequently
- AOS and dysarthria are uncommon in semantic and logopenic variants of PPA although features of AOS may be present in a minority of logopenic cases
- AOS is very common in nonfluent variant PPA; approximately one-third of patients with agrammatism and AOS have dysarthria
Recommendations

• Audiologic examination
• Neurologic examination to further describe nature of neurodegenerative disorder
  – PET
  – Electromyography
• Continued speech-language treatment
• Follow-up assessment in 6 months

Treatment Recommendations

• Train compensatory strategies for enhanced verbal communication and QOL
  – Provide semantic cues and topic areas for semantic context
  – Use gestures and orthographic cues
• Continue use of writing tablet/other AAC
• Consider use of TTY
• Increase vocal loudness to improve respiratory support and speech intelligibility
• Develop plan for weekly homework to maintain current best levels of function
• Discontinue isometric/isotonic oral exercises

Wambaugh et al., 2006b; Abbs & De Paul, 1989
Patient and family education

- Describe components of speech-language assessment
- Review findings
  - Progressive AOS and aphasia with co-occurring spastic dysarthria
  - Not the result of a single degenerative condition; tends to be associated with diagnoses that have prominent motor rather than cognitive deficits (CBD, PSP, MND)
- Discuss recommendations
  - Compensatory strategies vs. oral exercises
  - Importance of tracking the progression

Follow-up: January, 2015

- Contact with Neurology to prepare for speech-language pathology follow-up testing
  - August, 2014: Blood tests for paraneoplastic antibodies (negative), chest CT for complaints of cough, weight loss
  - Neurology follow-up scheduled for November, 2014; patient refused to attend
  - No attempts to schedule speech-language pathology follow-up
- No further medical care provided at RUMC
## Summary

### Progressive AOS

- Slow rate
- Insertion of pauses w/in words
- Syllable segmentation
- Perceived sound substitutions
- Reduced speech prosody

### Aphasia

- Reduced fluency
- Poor sentence grammar
- Reduced phrase length
- Mildly impaired auditory and reading comprehension.
- Mild naming deficits
- Writing impairment

### Spastic dysarthria

- Imprecise articulation
- Strained vocal quality
- Reduced respiratory-phonatory support
- Hypernasality
- Poor speech intelligibility

### Additional findings:

- toe paresthesias
- possible L or L > R atrophy
- overall weakness/fatigue
- NVIQ: low range of normal
Conclusion

• AOS can be the first and most prominent manifestation of neurodegenerative disease (Duffy, 2006)

• Speech-language assessment essential to identifying and describing the communication deficits found in neurodegenerative disorders as well as to track their progression over time

• SLP must be cognizant of the nature and progression of a neurodegenerative disorder when planning intervention

References


