Relapse in Stuttering with the Onset of Primary Progressive Aphasia

Sir,

Developmental stuttering (childhood fluency disorder) manifests in approximately 5–10% of children between 2 and 7 years.[1] A total of 65–85% of these children recover by 16 years of age, making the prevalence less than 1% in adults.[2] Although reports on reappearance of developmental stuttering as a result of neurological conditions are sparse, there are a few case vignettes demonstrating this phenomenon in Parkinson's disease,[3] stroke[4] and Alzheimer's dementia.[5] Here, we report of a patient who experienced reappearance of stuttering with the onset of primary progressive aphasia (PPA). PPAs, often referred to as 'language-led-dementias', are a group of neurodegenerative disorders that present initially with a focused slow progressive deterioration of the brain systems that govern language.[6] There are three main criteria for a root diagnosis of PPA: (a) gradually developing language impairment which presents as frequent word-finding pauses and is often masked with filler words, grammatically incorrect sentences, subtle naming difficulties, impaired repetition of phrases, and sentences along with emerging comprehension and spelling difficulty, (b) language impairment that is caused by neurodegenerative etiology, (c) aphasia is the primary impairment affecting daily living while other cognitive skills are comparatively preserved during the onset period.[7] Most frequent subtypes of PPA are agrammatic (relatively preserved language comprehension (LC) and significantly affected syntax, fluency, repetition), logopenic (relatively preserved LC, syntax, fluency and significant word retrieval deficits, circumlocutions, phonemic paraphasias), and semantic (relatively preserved syntax, repetition and significant LC deficits, naming deficits, semantic paraphasias, circumlocutions).[7] Diagnosis is made through detailed review of medical records, informant and caregiver reported history, clinical examinations and is correlated with image related tests and/or confirmed pathological findings. The site of lesion is often not easily identified during initial stages and diagnosis continues to remain a challenge even for experienced clinicians.
A 73-year-old right-handed male, accompanied by his wife, reported to the neurology outpatient clinic. He complained of stuttering since 7 months and occasional difficulty in recalling names of people and objects since a year. He spoke Konkani, Kannada and English. He was diagnosed with logogenic-PPA (initial stages) by a multi-disciplinary team that comprised neurologists, neuro-radiologists, neuro-psychologists and speech-language pathologists (SLPs). MRI showed asymmetric atrophy of the left perisylvian frontal lobe [Figure 1]. SLPs decided to probe into his dysfluencies, since this was the primary area of concern for the patient and his wife.

Speech-language evaluation began with a detailed clinical history which revealed developmental stuttering as early as 10 years of age. The patient did not receive formal speech-language therapy (SLT). He coped with his dysfluencies using a self-found strategy of ‘talking slowly’ (i.e. reducing rate of speech). He recalled becoming fluent by 25 years of age with rare instances of hard contacts. However, at the time of interview, he reported a decline in speech fluency (i.e. 48 years later). He experienced effortful speech which later progressed to stuttering like behaviors. He described that dysfluencies he experienced were similar to those when he was growing up; however, severity was almost two to three times more than before. A strong familial history of stuttering was reported. Patient’s mother, sister, brother and maternal uncle exhibited childhood dysfluencies.

At the time of consultation, the patient’s dysfluencies comprised initial syllable repetitions, part word repetitions and filled pauses. No individual or situational variations were reported. Dysfluencies occurred in a similar fashion in all spoken languages. Secondary stuttering behaviors (nostril flaring and subtle facial grimaces) were noted. Stuttering severity instrument 4[9] [Table 1] denoted moderate stuttering. He also demonstrated an adaptation effect during reading task (decrease in stuttering with increase in reading passage familiarity). Considering all observations mentioned above, a diagnosis of relapse in stuttering was made.

Patient’s reading-writing skills were informally assessed and appeared adequate. He used short phrases and demonstrated appropriate syntax during conversations. Although his fluency, auditory verbal comprehension, repetition and confrontational naming was intact based on informal language evaluation, he presented with word finding difficulties during picture description and word fluency tasks. He also had an increased latency for naming objects. While he had no difficulty repeating words and short phrases, he exhibited difficulties in complex sentences (e.g. effortful recall, phonemic substitutions, omission of words). He required help to recall lyrics of rhymes, sequence complex events, identify semantic and syntactic anomalies, and summarize events when assessed on cognitive linguistic tasks. He also had subtle decrease in performance of working memory tasks (word list, delayed sentence, letter and number recall). Reasoning, problem solving and judgment related task performances were adequate with occasional delay in responses. This demonstrated initial stages of cognitive-linguistic (C-L) impairment.

Patient was enrolled for SLT. He could attend only one session since he had to travel. Based on primary concern of patient and his wife, SLPs observations and, limited availability of time, intervention for stuttering alone was provided. Fluency shaping therapy[9] was introduced and breathing exercises were demonstrated. An immediate improvement in speech fluency was seen. Patient was asked to follow these techniques until follow-up which was 45 days later. His speech fluency had considerably improved [see Video 1], stuttering severity was mild [Table 1]. However, he continued to show word finding and word fluency deficits confirming the underlying C-L decline. In fact, C-L deficits were more evident during follow-up than before. It is plausible that C-L deficits were overshadowed by the presence of dysfluencies. Therapy was continued with a focus on C-L skills (word fluency, semantic and working memory, complex reasoning, problem solving) for 6 months after which patient discontinued due to personal reasons.

Through this case report, we postulate that the patient’s initial awareness of speech and language difficulties may have led to increased anxiety,[10] which could have attributed to relapse in childhood stuttering. We recognize we are relying on self-report and caregiver report to arrive at the diagnosis of relapse in developmental stuttering. That said, this case report highlights the possibility of re-appearance of stuttering with onset of PPA which is not a common presentation. It also highlights the importance of a SLP in aiding in differential diagnosis and providing intervention for both, the prominent condition (stuttering) and the more subtle, gradually progressing C-L deficits (PPA).

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published.
and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest
There are no conflicts of interest.

| Test                          | Sub-section               | Initial assessment | Follow-up assessment |
|------------------------------|---------------------------|--------------------|---------------------|
| Stuttering severity          | Frequency                 | 7                  | 5                   |
| Instrument-4                 | Speaking task             | 6                  | 4                   |
|                              | Reading task              | 10                 | 8                   |
|                              | Physical concomitants     | 3                  | 2                   |
|                              | Total overall score       | 26                 | 19                  |
|                              | Impression                | Moderate stuttering| Mild stuttering     |

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