Panel: Unusual Aphasias: Slowly Progressive Aphasia

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My task in this special session is to deal with the underground, subversive, sneaky side of aphasia, or what has been called slowly progressive aphasia. Like any subversive activity or insidious process, its very existence has been questioned, denied, overidentified, or viewed as just part of a larger all-out war on the central nervous system. Without wishing to instigate an aphasiologic McCarthysism, my purpose is to review evidence regarding the existence of slowly progressive aphasia, to present two cases I believe represent the condition, and to discuss the implications of such a disorder for our clinical practices.

Disorders that might be associated with a gradual decline of language ability extend beyond the common etiologies of aphasia and typically lead us to confront the debatable notion of "aphasia in dementia." Etiologies of dementia include several conditions which usually are diagnosed by history, clinical examination, and by default, and often are confirmed only at autopsy. Alzheimer's, Pick's, and Creutzfeldt-Jakob diseases, hereditary dysphasic dementia, and "unknown" probably represent the conditions in which a diagnosis of aphasia is most often debated.

Many neurologists talk with confidence about the "aphasia of dementia." For example, Kertesz (1985) and Cummings et al. (1985) report that it is a consistent manifestation of Alzheimer's disease. Bayles and Kaszniak (1987), on the other hand, point out that many speech pathologists are uncomfortable calling demented patients aphasic because it violates our concepts that aphasia should be disproportionate to other cognitive deficits, reflect a focal lesion, and be associated with sudden onset. Wertz (1982), referring to the tendency of demented patients to bring urine samples to their language evaluations, and to accuse us of making "silly goddamned requests" like "open your tongue," probably expressed the majority view when he said "because the language, the prognosis, and the appropriate management for aphasia and dementia differ, I do not believe that talking about aphasia in dementia is very useful" (p. 358).

That viewpoint makes a lot of sense. But does it include individuals who present with aphasia of insidious onset, gradual progression, and prolonged course, in the absence of generalized cognitive impairments (at least for a substantial period of time), due to a degenerative condition, presumably and predominantly involving the left perisylvian region of the brain? A weaker definition of slowly progressive aphasia would view it as the prominent early symptom of a dementing disease. Evidence documenting such a condition would challenge traditional concepts of aphasia's etiology and course, and perhaps "focality," or at least the required presence of an identifiable left hemisphere lesion.

THE LITERATURE

What does the literature tell us? Although very uneven in the quality of its documentation of aphasia, there is evidence that suggests the condition exists. There are several reports of the emergence of progressive aphasia as the first and prominent manifestation of Pick's disease. Wechsler (1977) described a 67-year-old with a two-year history of speech difficulty

in which personality and behavior changes did not emerge until the second year. Wechsler stressed the importance of recognizing the possibility that aphasia may be the presenting and most prominent symptom of degenerative dementia. He and his colleagues subsequently presented autopsy findings which confirmed the diagnosis of Pick's disease for their case (Wechsler et al., 1982).

Holland and colleagues (1985) documented a case with autopsy-confirmed Pick's disease in which its only apparent manifestation for the first 10 years was language decline. Although the authors studiously avoided calling the problem aphasia, and pointed out that the language difficulty was "atypical" of classic aphasia, it clearly represented a progressive deterioration of language in the presence of relatively preserved memory and cognition. (For other relevant cases of Pick's disease, see Cole et al. (1979) and Scully, Mark, and McNeely (1986)).

With regard to Alzheimer's disease, Popacar and Williams (1984) reported an autopsy-proven case in which a relatively isolated aphasia was the prominent symptom for the first two years. In contrast, Folstein and Breitner's (1981) work is of questionable relevance to issues regarding the existence of slowly progressive aphasia. Although they argued that aphasia was a distinguishing feature of familial Alzheimer's disease, they provided less than flimsy evidence that their subjects were aphasic and did not clearly establish its predominance over other cognitive deficits. Another example along this line is reflected in the work of Morris et al. (1984) who appear to have coined the term "hereditary dysphasic dementia." This term suggests to me that aphasia is/should be the prominent or only symptom of this type of dementia. In fact, however, their 10 patients' first symptoms typically included memory deficits, intellectual decline, and behavior and personality changes. While they did indicate that aphasia occasionally was the initial symptom, this does not seem to justify the inclusion of the term aphasia within the diagnostic label, at least as I've defined it above.

There also have been several case studies documenting the presence of slowly progressive aphasia of undetermined etiology. The most interesting and frequently referred to work in this regard is that of Mesulam (1982) who presented six cases whose aphasia was isolated or largely isolated for 5-11 years. The presenting complaint in all cases was related to speech and language, and examination confirmed its isolation or prominence. The EEG, CT scans, and/or auditory evoked potentials implicated the left perisylvian region in each patient. Those for whom WAIS scores were obtained showed a verbal-performance discrepancy in favor of performance. Reports by Heath et al. (1983), Kirshner et al. (1984), and Horner (1985) also document the occurrence of aphasia as the initial or prominent manifestation of slowly progressive degenerative disease. (The cases of Gordon and Selnes (1984) and Mingazzini (1913-14) are also of relevance.)

Taken together, it seems there are at least 25 cases in the literature with histories and complaints of slowly progressive language disability with absence or relative absence of other signs and symptoms typical of global dementia.

There is also some anatomic and physiologic evidence to support the potential for a progressive isolated impairment of language. For example, a number of the cases reviewed above have had EEG evidence of focal or predominant left hemisphere abnormalities. CT scans frequently demonstrated exclusive or greater left than right hemisphere perisylvian atrophy. The autopsy findings for patients with Pick's disease documented by Holland

et al. (1985) and Wechsler (1982) showed asymmetric atrophy of the left perisylvian area.

A PET scan study by Chawluk et al. (1986) demonstrated an overwhelmingly clearcut focal decrease in left temporal or parietal metabolism in two patients with slowly progressive aphasia. In addition, recent PET studies of patients with clinically diagnosed Alzheimer's disease (Foster et al., 1983; Haxby et al., 1985) show that asymmetric reduction of glucose metabolism correlates with disproportionate involvement of language or visuospatial functions.

That slowly progressive aphasia potentially can occur also receives indirect support from reports of isolated progression of other higher order deficits. For example, DeRenzi (1986) and Crystal et al. (1981) have presented cases of slowly progressive, relatively isolated visual agnosia, generalized apraxia, and "right parietal lobe syndrome." DeRenzi concluded in his study that "an isolated deficit of language, or visual recognition or praxis can for a long time and possibly forever stand out as the only sumptom of a progressive, degenerative disease" (p. 179).

ILLUSTRATIVE CASES

I'd like to present two cases now from my own clinical experience that are representative of 15 or 20 patients I've seen in the past four years with isolated or relatively isolated slowly progressive language decline. I believe they do not correspond to the histories or language behavior of "typical" demented patients.

Case 1. The first is that of a 47-year-old predominantly left handed pipe fitter who presented with a four-year history of progressive speech and language problems. Six weeks after onset, at another institution, he received a diagnosis of CVA solely based on his speech and language problem. He had therapy at that time for six weeks, perhaps with some improvement. From then on there was a slow but definite progression of his problem. Because of this, he stopped going out on jobs as a pipe fitter a year prior to our evaluation, but continued to work on an assembly line. His wife felt his memory might be impaired but could not give examples that could not also be attributed to his language deficits. His personality was reportedly unchanged. He was able to do all activities of daily living and drove as always. His wife had noted progressive clumsiness in his right hand since the onset of his speech difficulty.

His neurologic examination found a mild right central facial weakness, clumsiness of his right hand, and some right arm hyperreflexia. CT scan was negative but MRI showed dilation of the lateral ventricles, with the left slightly larger than the right. EEG showed a significant, nonspecific disturbance in both hemispheres, maximal frontally and at times of greater prominence in the left fronto-temporal-central area.

On measures of verbal comprehension and retention, performance was accurate but marginal for single words and simple commands and clearly deficient for complex commands and digit span. Verbal expression was nonfluent, with short phrases, some telegraphic responses, and numerous pauses. Content was somewhat empty. He made two corrected semantic errors on picture naming. Oral spelling was 4 of 6 words and slow. He supplemented his speech with some gestures that sometimes helped to augment meaning. He was unable to repeat more than 3-word sentences, with loss of information,

syntactic errors, and apraxia of speech characterizing errors. Reading aloud was very slow, with numerous verbal paraphasias. He comprehended 13 of 16 basic sentences. He wrote his name and drew simple objects adequately, but misspelled 3 of 10 dictated words.

There was no dysarthria but he did have a moderate to marked apraxia of speech which was apparent on imitation and spontaneous tasks. He had a sucking reflex, and nonverbal oral apraxia on command and imitation. I saw no evidence of confusion and nothing to suggest generalized cognitive impairment beyond his aphasia. My diagnosis was aphasia and apraxia of speech and I recommended speech and language therapy. In my opinion, he behaved like many patients with aphasia and apraxia of speech due to a focal left hemisphere CVA.

Psychometric assessment noted extreme difficulty on any task that was verbally dependent and verbal IQ could not be estimated. His Wechsler Memory Quotient, which is verbally dependent, was 49. Performance on the Bender suggested a mild constructional apraxia. He also had trouble on nonverbal portions of the WAIS, scoring 73. This led to the conclusion that he may have had cognitive compromise beyond that explainable by his aphasia.

He left with a neurologic diagnosis of slowly progressive cortical degenerative process, perhaps atypical early Alzheimer's disease. He was seen one year later and the neurologist felt his aphasia and writing difficulties had worsened. He had an apraxia of all motor movements and some myoclonic tremor of his right upper extremity. The neurologist questioned the presence of "silly affect." He didn't stay for other exams, although an EEG had been recommended to check for Jakob-Creutzfeld disease.

Case 2. This case is that of a 60-year-old right-handed dermatologist and member of MENSA. Three years prior to our assessment he developed word-finding difficulties and word substitutions. It was difficult to establish if the course was one of subacute onset with stabilization or very subtle progression, but we ultimately concluded it was progressive with good compensation. He was seen by many neurologists, some of whom raised questions about Alzheimer's disease, although most felt the problem was too restricted. Outside neuropsychological testing raised suspicions about cognitive decline but admitted that language impairment contributed to reduced scores.

Perhaps most telling was the comment of the examining neurologist. He said, "I spent at least 1.5 hours with Dr. ____ focusing on whether or not I could detect any cognitive dysfunction outside of language/motor speech . . . and I could not . . . His cognitive functioning seems to be better than mine! (which may not be saying much)." Outside CT and MRI scans showed predominant left sylvian atrophy. The initial impression was that he probably had had a small dominant hemisphere infarct but focal degenerative processes could not be ruled out.

Language exam revealed the following. He struggled a bit with complex commands and made 10 errors on Part V of the Token Test. He worked hard to repeat 6 digits forward and 5 reversed. Verbal expression was slow and obviously measured, with frequent pauses and pronoun for noun substitutions or failure to establish a referent. Paraphasias most often involved pronouns and prepositions. Circumlocution was apparent and his content bordered on being empty at times. He sometimes strayed from the topic but I wasn't sure if this was secondary to circumlocution versus true tangentiality. He named 18 of 18 pictures accurately but with delays. He scored only 26 on the Word Fluency Test. He repeated 6 of 10 sentences, omitting substantives and

making some semantic errors. His word definitions were concrete, I thought, beyond that explainable by his language deficit. For example, he defined bargain as "If you go to a regular store and there is a sale you can get 50% off." He perseverated once and misspelled 1 of 6 simple words.

Reading was slow but accurate and sentence and paragraph comprehension were adequate. He made one verbal paraphasia when writing to dictation but made no spelling errors. His own sentence generation was adequate.

In terms of motor speech, he made some sound substitutions and revisions on repetition and spontaneous tasks, particularly on multisyllabic words and complex sentences. There was no dysarthria or nonverbal oral apraxia.

There was no evidence of confusion or disorientation, but when I asked him to evaluate his performance following several tasks, he felt it was normal.

His wife admitted to me that, "Sometimes I get the feeling he's not here." She stated that he would sometimes walk away when being talked to and occasionally eat at a party before everyone was served.

It was my impression that he had a moderate aphasia and mild apraxia of speech. I also felt his relative lack of awareness of errors (perhaps a component of his tendency to be tangential), and what his wife had told me raised questions of more generalized cognitive compromise. I did, however, feel that the great majority of his difficulty on verbal tasks was consistent with focal language disturbance and apraxia of speech. I recommended speech and language therapy.

Psychometrics showed a verbal IQ of 103, performance IQ of 84. This represented a 10-point decline in verbal IQ from two years previous but no decline in performance score. His memory quotient was 101 but his recall after one hour was 100%! The neuropsychologist felt that although there was evidence for some generalized cognitive impairment it was atypical for dementia.

The final neurologic disposition was that the weight of evidence favored degenerative disease, possibly the "slowly progressive aphasia" described by Mesulam (1982).

One obvious weakness of these cases is that the patients were not followed serially by us. However, by history their problems were slowly progressive and unusually focal to speech and language. Another important difference between these two cases and the typical patient with dementia is that both had an apraxia of speech and one was quite obviously nonfluent. We have seen at least three additional patients with similar histories and speech and language deficits.

COMPOSITE OBSERVATIONS

Pulling all of this together, I think we can make the following observations. Those with so-called slowly progressive aphasia present with primary complaints of language difficulty and not complaints about memory, personality change, or inappropriate behavior. Their language, although not always, in many, most, or all ways can look and smell like the aphasia we all identify with acute onset focal etiologies. Contrary to that usually seen in dementia, the aphasia may be nonfluent and accompanied by apraxia of speech and unilateral motor deficits. There may or may not be other confirmatory evidence of left hemisphere abnormalities.

TENTATIVE CONCLUSIONS

I think the following conclusions are justified or at least legitimate subjects for debate:

1. The condition of "slowly progressive aphasia" probably does exist.
2. Its history and presentation are different from so-called "Alzheimer's aphasia" or other language problems which are embedded within more globally dementing conditions.
3. The aphasia may or may not be distinguishable from that associated with more typical etiology and identifiable locus.
4. Its underlying neuropathology may be heterogeneous, possibly reflecting a focal, lateralized version of Pick's, Alzheimer's or Creutzfeldt-Jakob diseases, or may be associated with an undefined or nonspecific atrophic process.
5. Even if it is simply a precursor to more global deterioration, its early prominence and relative isolation justify its distinction from other common dementing behavioral profiles.
6. Its existence justifies a modification of our concept of aphasia as requiring an acute, nonprogressive etiology with clearly identifiable focal anatomic and neurophysiologic manifestations.
7. The existence of slowly progressive aphasia highlights or raises some important issues and challenges. First, it challenges further our ability to distinguish the language of typical focal aphasia from aberrations in language that may reflect more global involvement. It should also push us to take a more careful look at some of the nonlinguistic behaviors of patients whose etiology is known focal impairment, such as CVA. We may accept too readily the normalcy of such functions in those with focal lesions and may only attend to them when the issue of focal versus diffuse involvement is in question. Comprehensive diagnosis requires that we look as closely at these behaviors in patients with known etiology as in those with unknown etiology.

Second, most patients with suspected slowly progressive aphasia will be evaluated neuropsychometrically. This should promote increased attention to the performance of any aphasic patient on verbal and nonverbal measures of intelligence and memory, for only if we know the range and patterns of performance on such measures of those with known focal aphasia can we state with confidence that the performance of those with suspected slowly progressive aphasia does or does not reflect more generalized impairment. It renews questions about the inferences regarding general intellectual abilities that justifiably can be drawn for anyone with prominent and predominant aphasia.

Finally, should slowly progressive aphasia be managed? It seems to me that it should, although I'm not at all sure it should be managed in the same way as nonprogressive aphasia. If we do manage this problem, how do we assess efficacy? This presents some interesting challenges to single-case designs in which evidence of efficacy may hinge on maintaining a plateau or slowing deterioration of specific abilities. (At least we wouldn't have to worry about the effects of spontaneous recovery.) Should we be able to reduce the rate of decline or maintain or improve communication we would know whether to treat the condition when it occurs and may have some potentially powerful additional data to support the efficacy of aphasia therapy in general.

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