

## SHORT REPORT

## Pure progressive aphemia

L Cohen, N Benoit, P Van Eeckhout, B Ducarne, P Brunet

**Abstract**

**Aphemia, also called anarthria or severe apraxia of speech, is a rare disorder of speech production usually resulting from vascular lesions affecting the inferior premotor cortex of the left hemisphere. A patient presenting with aphemia as the sole manifestation of primary progressive aphasia (PPA) is reported.**

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**Case study**

A 51 year old right handed pharmacist presented with a three year history of progressive speech impairment which was still compatible with nearly normal work. There was no significant medical history. She complained of isolated articulation difficulties. General clinical and neurological examination was normal. There were no cerebellar, pyramidal nor extra-pyramidal signs. Usual biological screening, CSF and CT scan were normal. The voice was slow, low-pitched and considerably aprosodic. Attempts to sing only yielded an unidentifiable monotone. There were a few inversions of phonemes (for example, "sacheur" for "chasseur"), and frequent phonetic transformations, yielding poorly differentiated speech sounds. There was severe bucco-facial apraxia: the patient could not blow a kiss, whistle, cluck her tongue, etc. Except for the arthric impairment, oral expression and comprehension, reading and writing, were flawless on clinical evaluation. Spontaneous drawing and copy of the Rey-Osterrieth complex figure were satisfactory. There was no limb apraxia for symbolic, arbitrary and utilisation gestures, on command or by example. On a simple tapping task, however, the patient could not keep a regular pace for more than a few seconds, and when tested on alternating gestural sequences, she made some perseverative errors.

Language output gradually deteriorated over the next two years, and the patient had to take early retirement. Five years after onset, speech was virtually unintelligible. Spoken output was effortful and severely dysarthric. Phonetic transformations affected all types of phonemes, although vowels were often more easily identifiable. The patient could not adequately alternate voiced and unvoiced sounds, yielding nearly permanent

voicing. Speech rate was irregular, and poorly identified additional sounds were often inserted (for example, /mawerdi/ for "mardi"). When she was encouraged to emphasise syllables and to adopt a slow, controlled and regular pace, speech became less slurred and more easily intelligible. Speech was strictly aprosodic, and the patient could depart from her monotone neither to produce linguistic prosody (for example, final rising in questions), nor to express emotions or sing. Voluntary and automatic speech (songs, prayers, days of the week) were equally affected. There was still a major buccofacial apraxia, but no evidence of pseudobulbar syndrome, and no swallowing or breathing difficulties.

The impairment remained apparently confined to pure arthric difficulties, and the patient could communicate quite effectively by writing down messages. Writing was virtually free of misspellings. However, there were occasional omissions of short words such as the definite and indefinite article, (for example, [The] sink is full; the man stands on [a] chair).

The following neuropsychological investigations were conducted five years after clinical onset. The patient was submitted to those subtests of a French version of the Boston Diagnostic Aphasia Examination (BDAE) that require no verbal output. She scored at ceiling level in all tests of auditory comprehension (word discrimination: 72/72; body-part identification: 20/20; commands: 15/15; complex ideational material: 11/12) and reading comprehension (symbol discrimination: 10/10; word recognition: 8/8; word-picture matching: 10/10; sentences and paragraphs: 10/10). Her performance was good on the writing subtests (mechanics: 2/3; serial writing: 44/47; simple dictation 15/15; confrontation naming: 10/10; spelling to dictation: 9/10; dictation of sentences: 10/12; narrative writing: 2/4). On a written test of fluency on controlled association, she produced 13 names of animals in one minute. The patient was perfect on an additional written test of picture naming (40/40 correct).

On the Wisconsin Card Sort Test, the patient identified 5/6 sorting criteria, correctly classifying 36/48 cards. She scored 27/36 correct on the Coloured Progressive Matrices test, corresponding to the 62nd percentile. There were no errors on subtests A and B of the Trail-Making Test, although the

Service de Neurologie,  
Hôpital de la  
Salpêtrière, Paris,  
France  
L Cohen  
N Benoit  
P Van Eeckhout  
B Ducarne  
P Brunet

Correspondence to:  
Dr Cohen, Service de  
Neurologie, Hôpital de la  
Salpêtrière, 47 Bd de  
l'Hôpital, 75651 Paris  
CEDEX 13, France.

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performance was not quite as fast as could be expected (1 standard deviation below the mean). Some constructional difficulties were evidenced by her IQ of 87 on the performance subtests of the Wechsler Adult Intelligence Scale, and by a correct but piecemeal copy of the complex Rey-Osterrieth figure. The patient scored at normal levels in the delayed recognition subtests for sentences, abstract drawings and object pictures of the BEM 144 memory scale.<sup>1</sup>

Additional anatomical and functional imaging data were gathered four years after onset. MRI revealed a moderately enlarged left lateral sulcus. Single photon emission computed tomography (SPECT) using inhalation of <sup>133</sup>Xenon showed a reduced cerebral blood flow (CBF) in the frontal regions, mostly affecting the lower part of the left frontal lobe. There was an additional slight hypoperfusion in the right temporal lobe. Despite global CBF increase following administration of acetazolamide, there was no significant response in the frontal regions.

### Discussion

Our patient fulfilled the diagnostic criteria for PPA: a history of progressive language impairment extending over five years, with a relative preservation of other mental functions and complete independence in daily life.<sup>2</sup> She presented with a primarily articulatory disorder, sparing all other components of language production and comprehension, characteristically corresponding to the syndrome of aphemia. This condition has been described under a wide variety of terms, depending on whether it was considered an aphasic (for example, phonetic disintegration, pure motor aphasia), a dysarthric (for example, anarthria, cortical or subcortical dysarthria), or an apraxic disorder (for example, apraxia of speech, articulatory dyspraxia).<sup>3</sup>

Aphemia generally results from focal lesions of the left hemisphere, affecting the lower part of the primary motor cortex (precentral gyrus), the contiguous premotor cortex (particularly area 44 or pars opercularis), or the immediately underlying white matter.<sup>3</sup> The role of this region in the process of speech is confirmed by activation data obtained with normal subjects using PET.<sup>4</sup> This area falls within the hypometabolic inferior left frontal region that was shown by SPECT in our patient.

Lesions are often more extensive, and aphemia is therefore generally associated with properly linguistic deficits of the Broca type.<sup>5</sup> This was not the case in our patient, apart maybe from the few syntactic errors in

written language mentioned previously. In cases where the inability to perform skilled gestures extends beyond the production of speech sounds, aphemia may be associated with buccofacial apraxia.<sup>6</sup> In our patient, aprosody was probably part of this generalised motor programming deficit, affecting the larynx in addition to the pharynx and mouth. Although a left hemispheric lesion is considered sufficient to produce aphemia and buccofacial apraxia, the severity of these deficits and the absence of any compensation might point to the additional right hemispheric involvement.

A wide variety of clinical types of aphasia have been described in patients with PPA. However, reduced fluency and phonological disorders are seemingly the most characteristic features of PPA, as opposed to aphasias associated with dementia of the Alzheimer type.<sup>2</sup> Our case is similar to the one described by Delecluse *et al*,<sup>7</sup> who showed a far more extensive cerebral involvement on SPECT. In the other reported cases of PPA, aphemia was consistently associated with other linguistic deficits affecting word finding, syntax or comprehension.<sup>8,9</sup>

PPA has a purely clinical definition, and has been shown to be associated with several well defined cerebral diseases, such as Pick's disease, Alzheimer's disease, or Creutzfeldt-Jakob disease. The protracted evolution with almost exclusive speech impairment, and the unusual clinical presentation as pure aphemia, make these diagnoses implausible and are more suggestive of more recently identified lobar atrophies.<sup>10</sup>

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