Archives of Neurology and Neuro Disorders

ISSN: 2638-504X

Volume 1, Issue 2, 2018, PP: 04-06

Asymmetric Cortical Atrophy in the Primary Progressive Aphasias

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Abstract

We present two patients with variants of primary progressive aphasia (PPA), with differing patterns of atrophy on MRI. Clinical deficits in this condition depend on the location of focal atrophy. We present imaging findings in two patients with different variants of PPA, which serve to highlight the differentiation of this condition into distinct aphasic syndromes, as well as the utility of structural imaging in its diagnosis.

Keywords: *Primary progressive aphasia;semantic dementia; progressive non-fluent aphasia; magnetic resonance imaging; asymmetric cortical atrophy*

Introduction

Primary progressive aphasias (PPA) are a group of neurodegenerative dementias characterized by dissolution of language as the principal clinical abnormality and relative preservation of other cognitive domains for most of the disease course. [1]. They are usually associated with fronto-temporal lobar degeneration (FTLD) pathology but also on occasion with the pathological features of Alzheimer's disease. Up to 43% of patients with FTLD have been clinically classified as having PPA[2]. Brains of patients with PPA display asymmetric atrophy of the peri-sylvian language network in the left hemisphere. [3,4] Progressive language dysfunction can be of various types depending on the anatomical location involved. We present two patients with variants of PPA exhibiting focal atrophy in different locations on magnetic resonance imaging (MRI), serving to differentiate these variants.

Patient 1

A 76 year old woman presented with progressive reduction in verbal output for 3 years, and was nearly mute for the last one year. Examination revealed motor aphasia with severely impaired speech fluency with phonemic paraphasias and anomia with intact repetition and comprehension. She had preserved frontal executive functions, recent memory, behaviour and comportment. Visual-spatial function and praxis were normal. A diagnosis of progressive non-fluent aphasia (PNFA) was considered.

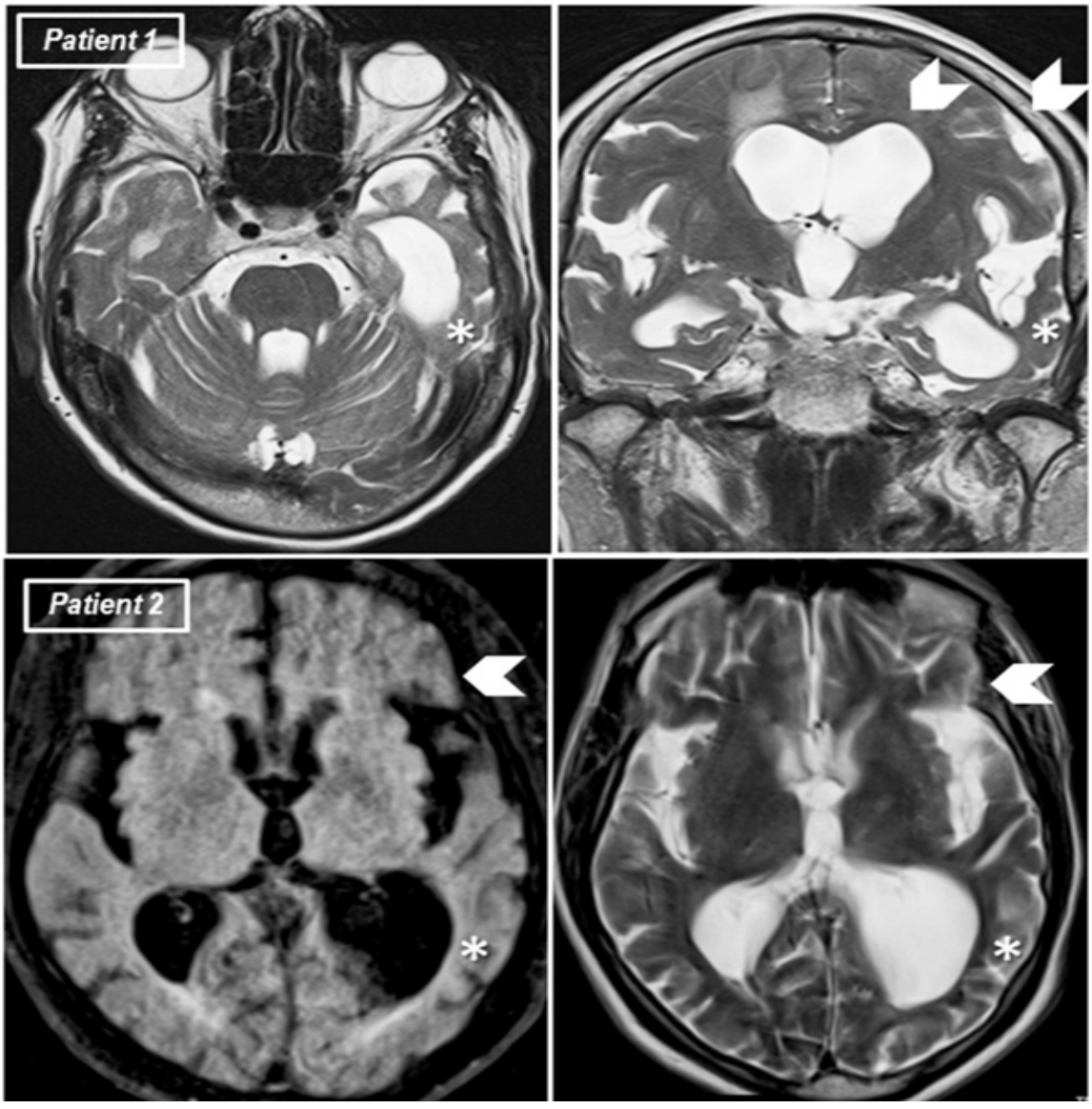
Patient 2

A 69 year old man presented with progressive language dysfunction for two years. This was characterised by circumlocutous speech with difficulties with confrontation naming, semantic paraphasias, and impaired category fluency and comprehension of single words, with relatively preserved verbal fluency, repetition and syntax. Associative visual agnosia was present. No other cognitive deficits could be detected, although memory testing was difficult because of the language impairment. A diagnosis of sematic dementia (SD) was made.

PNFA is a tauopathy presenting mainly as an expressive language disorder with severe problems in word retrieval. Patients present with agrammatism and changes in fluency, pronunciation, or difficulty in word location. Speech is aprosodic and effortful. Phonologic errors are usually obvious in conversation, but comprehension is relatively preserved, and behavioural changes are uncommon until later in the

disease.[5-7] . MRI [Figure, Patient 1] shows significant insular, anterior temporal and inferior frontal atrophy of the dominant hemisphereon axial and coronal T2-weighted imageswith *ex vacuo* dilatation of the left temporal horn (asterisks), asymmetric peri-sylvian atrophy with widening of the left Sylvian fissure, and dilatation of the left frontal horn (chevrons). The mesial temporal structures are spared. [5,6,8,9].

Semantic dementia (SD) is thought to cause a breakdown in conceptual knowledge rather than a specific problem with language.Although patients with SD maintain good articulation, repetition, and syntax, they tend to use substitute phrases and stereotypical questions. Some patients also show deficits on nonverbal tasks that use visual, auditory, and other modalities. Semantic dementia (SD) is characterised by hypometabolism in the left anterior temporal and parietal lobes, in contrast to predominant left frontal hypometabolism seen in PNFA. Functional imaging presents more obvious differences than structural MRI. [5,10] In contrast to patient 1, patient 2 with SD exhibits predominantly antero-inferior left-sided temporal lobe atrophy (asterisks) on axial T2 and FLAIR, while the inferior frontal lobe is relatively preserved (chevron).[6,10] The axial fluid attenuated inversion recovery (FLAIR) image demonstrates high signal intensity in the corresponding subcortical white matter, which is probably related to gliosis and demyelination [6]. [Figure, Patient 2] The entorhinal, parahippocampal, and fusiform gyri are often affected. The right temporal lobe can be involved to a less severe degree.[6]



In conclusion, clinical features of PPA depend on the anatomical location involved by the disease process. MRI serves to distinguish between variants of PPA depending on the location of focal atrophy.

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Citation: **Aaron de Souza, Varun R Pai Kakode, Zico D'Costa**. *Asymmetric Cortical Atrophy in the Primary Progressive Aphasias. Archives of Neurology and Neuro Disorders. 2018; 1(2): 04-06.*