Chapter 9
Primary Progressive Aphasia in Northeast Brazil

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ABSTRACT

All 29 patients underwent structural neuroimaging examination. With the advent of analysis cerebrospinal fluid biomarkers, the authors were able to perform this in 38% of cases, with 45% having Alzheimer's pathology, 36% tau protein pathology, 9% having normal biomarkers, and 9% having increased β-amyloid protein alone. With technological developments, 55% of cases underwent brain single photon perfusion scintigraphy (SPECT) studies (81%) and brain PET/SCAN (19%). The clinical results were 43% compatible with the logopenic variant, 36% with the semantic variant, and 21% with the non-fluent variant. Around 3% of cases occurred due to a mutation in exon 5 of the VCP gene. Around 10% of cases underwent post-mortem studies, with Alzheimer's disease (logopenic variant) confirmed in 33%, Pick's disease (non-fluent variant) in 33%, and cortico-basal degeneration (variant not fluent).

Primary progressive aphasia (PPA) is a disorder mainly of language, neurodegenerative in nature, with a progressive course and a lethal evolution, differing widely in evolution both in each type of variant and in the time of its course (Ulugut et al. 2022). Until now, we believe that PPA, as a syndrome, has an open definition, especially regarding its variants and etiological correlation. In some cases, complaints of memory lapses occur before the appearance of language symptoms, or perhaps some patients do not know how to differentiate memory deficit from anomia; memory deficit is forgetfulness and anomie is the inability to say the name of an object (Brito-Marques, 2018). Language symptoms generally take around 2 years before

DOI: 10.4018/979-8-3693-0851-6.ch009

the syndrome begins to express symptoms other than those of language, usually detected by objective assessment (Mesulam, 1982).

Acute and chronic aphasic syndromes differ in terms of their topography (Binder, 2015). When comparing the topography between both aphasias, it was observed that PPA presents more involvement with the language neural networks in the left temporal cortex, especially in the temporal pole, while in aphasias due to acute lesions of vascular etiology, they generally spare the temporal poles (Binder, 2015). The mechanism between both aphasias may be different but with similar results. Although the semiology of PPA is mainly characterized by language alterations, early deficits in social cognition, emotion recognition and recent memory also stand out. There are also changes in the theory of mind and empathy, mainly in the semantic and agrammatic variants (Fittipaldi et al. 2019).

Depending on the variant, PPA can evolve into a more generalized state of dementia after 7 years of diagnosis, with the logopenic variant being the most aggressive form of PPA (Grossman & Irwin, 2018), and functional neuroimaging is more sensitive in pointing out early hypoperfusion in the same region (Brito-Marques-Brito-Marques, 2023). However, the literature does not provide a more comprehensive explanation as to whether PPA is an independent pathological condition or more of an atypical syndrome found in neurodegenerative diseases (Rahul & Ponniah, 2019). We observed that PPA presents itself in Northeast Brazil with the same clinical characteristics as anywhere else, including cases with pathological confirmation. The biggest difficulties are little support for research even within universities and the delay for patients to reach university services. Cultural comparative studies with other centers must be carried out.

BRIEF HISTORY

In 1892, Arnold Pick first described a progressive language disorder. In this disorder, there was atrophy in the frontal and temporal regions of the left cerebral hemisphere which he called progressive non-fluent or agrammatic aphasia (Hopper et al. 2023). A century later, Mesulam carefully studied six right-handed patients with slowly progressive aphasia, without intellectual and behavioral disorders. In most of these cases, symptoms began in the pre-senile phase due to anomic aphasia in 5 patients, and speech deafness in 1 of them. In the 90s, Neary et al. published the criteria for frontotemporal lobar degeneration. Based on these criteria, it was observed that lesions affecting the right side of the brain had different clinical expressions than those in the left hemisphere. On the right, behavioral variant frontotemporal dementia (bv-FTD) stood out, while on the left, PPA. An impairment in the left frontotemporal region can give rise to two distinct forms of PPA: the non-fluent