ATYPICAL VARIANT OF PRIMARY PROGRESSIVE APHASIA: A CASE REPORT

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BACKGROUND

Primary progressive aphasia (PPA) is a clinical syndrome characterized by progressive language impairment that interferes with word production or comprehension. PPA is now typically classified into three variants: non-fluent variant (nfvPPA) with agrammatism or apraxia of speech (effortful and halting speech), semantic variant (svPPA) with a progressive impairment knowledge of word meanings, and logopenic variant (lvPPA) with frequent word-finding pauses and poor repetition. Whereas the first two variants are usually reported as clinical phenotypes of the Frontotemporal Lobe Degeneration (FTLD), the logopenic variant has been primarily related to Alzheimer's disease. We describe a case of FTLD presenting with an atypical variant of PPA.

CASE REPORT

XX, a 57-year-old, right-handed woman, came to our attention complaining a two year history of language disorder characterized by significant word-finding difficulty, trouble forming complete sentences and impaired writing and reading. In the last year she also presented mild deficits in attention and in autobiographical memory. There were no behavioural changes. Family history was negative for dementia. Neurological examination was entirely normal. Neuropsychological examination showed disturbances in oral and written language, with loss of syntax, production of incomplete sentences, severe anomic disturbance, dysgraphia and dyslexia characterized by letter or syllables insertions, substitutions, transpositions and omission. Rare semantic paraphasias were noted in naming task in absence of frank impairment of cross modal semantic association tasks. Repetition of words and sentences was preserved. In addition the patient presented dysexecutive syndrome with deficits in selective attention and inattention. Brain MRI showed mild frontal atrophy; FDG-PET revealed left frontal cortex hypometabolism. Genetical analysis, including PGRN, MAPT and C9ORF72, were negative.

CONCLUSION

The present case presents with the clinical picture of clinical and neuropsychological features of PPA. The imaging revealed a selective frontal damage and let us infer this case is likely a clinical variant of the Frontotemporal Lobe Degeneration. Anyway, the neuropsychological features of the language disturbance did not allowed to classify the present case neither as a semantic variant nor a non-fluent variant. A logopenic variant was excluded as well. The present case is of interest because is likely an atypical presentation of the frontotemporal degeneration. This case of "unclassified form" of PPA should support consideration for an alternative classification of patients presenting with primary progressive speech and language disturbance.

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