Introduction

Nonfluent Progressive Aphasia is a linguistic variant of the Frontotemporal Lobe Degeneration. It is reported a very rare association of PNFA with motoneuron disease. When this rare association occurs primary lateral sclerosis is the most frequent syndrome associated to PNFA.

Case discussion

G., 74-year old Caucasian right-handed male presented complaining a two-year history of progressive language deterioration, mainly characterized by word finding problems and reading difficulties. No family history for neurological diseases was reported. Neurological examination was normal. Neuropsychological evaluation revealed a non fluent aphasia, with agrammatism and speech apraxia. Phonological errors were present both in oral production and in transcoding tasks such as reading, repetition and writing to dictation. An MRI scan was normal. By contrast a SPECT scan revealed abnormal perfusion in the left inferior frontal gyrus. Qualitative features of the G.’s clinical picture did not change in the following examinations performed every six months.

Three years later he turned up complaining slurred articulation, and mild dysphagia. This time neurological examination revealed a pyramidal syndrome with severe dysarthria, Babinsky sign and spasticity in his four limbs. Swallowing examination revealed mild dysphagia. His language problem was worsened with more frequent agrammatism and phonological errors. Language comprehension was spared for single words while showed some problems in grammatically complex sentences. A dysexecutive syndrome was present. Electromyography-electroneurography did not reveal sign in keeping with lower motoneuron disease. Repeated neuroimaging did not reveal new findings. MAPT, progranulin and C9orf72 known mutations were absent.

A further two-year follow-up was qualitatively similar. Both neuropsychological and neurological signs were worsened. The patient needed a wheelchair because he was unable to walk alone cause of spasticity. Dysphagia was worsened. Repeated electromyography-electroneurography excluded lower motoneuron disease.

Conclusion

The present case combines the association of PNFA and Primary Lateral Sclerosis. Amiotrophic Lateral Sclerosis and Frontotemporal Dementia are closely related especially in patients with mutation of the C9ORF72 gene. The association of PNFA with PLS is unusual and represents a further phenotypic variant of Frontotemporal Lobe Degeneration.

References