



ATYPICAL FRONTAL VARIANT OF ALZHEIMER'S DISEASE: A CASE REPORT

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Introduction

Atypical Alzheimer's Disease represents a heterogeneous spectrum within the AD pathology, which consists of three clinico-pathological syndromes: Logopenic Progressive Aphasia (LPA), Posterior Cortical Atrophy (CPA), and Frontal Variant of AD (fvAD) (Dubois et al, 2014). Such syndromes show cognitive symptoms that are not typical for AD, but all rest upon AD-related pathophysiology (Wolk 2013). FvAD is a particular phenotype that may mask the behavioral variant of Frontotemporal Lobar Degeneration (bvFTD) (Koedam et al, 2013). Although FTLD onsets typically over 60 years, tardive forms are not uncommon (de Rino et al, 2012).

Case report

A 80-year old graduated lady was evaluated at our Neurology Unit in February 2015 for the diagnostic assessment of progressive behavioral changes with aggressiveness, hostility to her daughter, reduction in personal care started almost 3 years before and gradually worsened. The most significant feature was a dramatic trend in hoarding, so that her home was found inaccessible by her daughter. In the past, she was a usually very elegant lady, teaching at high schools. She presented with sufficient temporal and topographic orientation, collaborative, showing reduction in speech and motor initiative. Medical history was positive for systemic hypertension and hypothyroidism in stable treatment, and two surgery interventions for uterine and breast cancer years before. The patient underwent a neuropsychological evaluation, which showed mild cognitive deficits mainly involving executive functions (planning, inhibitory control and divided attention, categorical fluency) and behavioral disturbances (delusions, hallucinations and depressed mood). Video-EEG showed regular, symmetric EEG pattern. Brain MRI scan did not show significant cortical atrophy. FDG-PET showed marked hypometabolism in the left mesial and lateral temporal lobe, and in the frontal lobes; posterior cingulate gyrus was spared. CSF classical biomarkers (A β 1-42, t-tau and p-tau) - measured by means of ELISA method (Fujirebio) - revealed A β 1-42 levels within the lower limit of reference (608 pg/mL, n.v.>550) and increased levels in total tau (571 pg/mL, n.v.< 300) and hyperphosphorylated tau (76 pg/mL, n.v.<60). A diagnosis of "atypical behavioral variant of AD" was finally formulated.

Follow-up visit

On July 2015, the neuropsychological and behavioral assessment did not show any progression.

Conclusions

As reported in IWG-2 criteria (Dubois et al, 2014) an estimated 11% of cases of AD present with an atypical phenotype sparing episodic memory accompanied by topographical evidence of brain damage in regions non typical for AD pathology. Differential diagnosis between fvAD and bvFTD is very challenging, needing the combination of pathophysiological and topographical biomarkers (Padovani et al, 2013).

References

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