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Neuroimaging, neuropsychological assessment and CSF biomarkers in posterior cortical atrophy: a case report.

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Background: Posterior Cortical Atrophy (PCA) is an insidiously progressive disorder that presents with deficits in visuospatial and visuoperceptual processing. Neurodegeneration can involve the occipito-parietal stream (dorsal "where" pathway) or the occipito-temporal stream (ventral "what" pathway). PCA is included as a clinical variant of Alzheimer disease (AD) in new research criteria (NIA-AA, 2011), with a similar CSF pattern, supporting the hypothesis that in most cases PCA can be considered a variant of AD. Also at post mortem analysis, most of the cases appear to be due to AD pathology (Alladi, 2007).

Objective:
We report the case of a PCA patient presenting with normal CSF biomarkers and atypical neuropsychological and MRI findings.

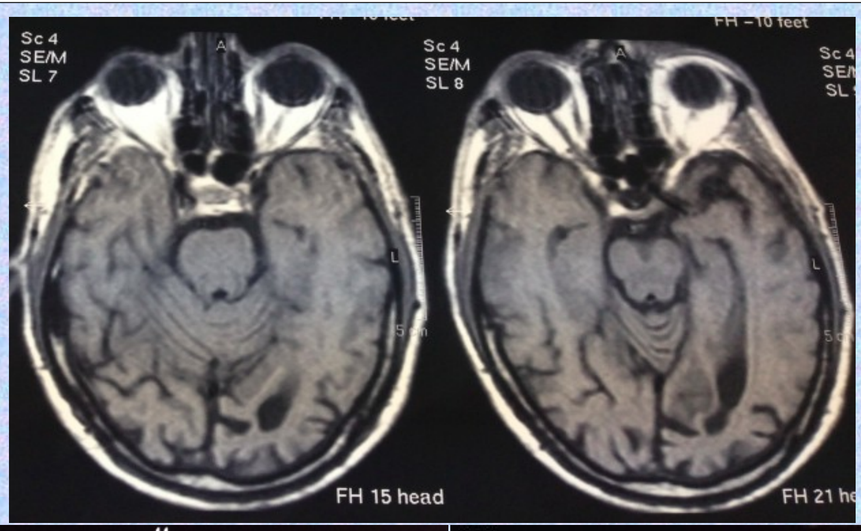
Subject and methods: A 73-year-old right-handed male with 8 years of formal education was referred to our Neurology Clinic because of progressive visual difficulties. He underwent neurological and neuropsychological evaluations, neuroimaging, lumbar puncture and genetic analysis.

Results:

- Neuropsychological evaluation: alexia, agraphia, visual agnosia, simultagnosia, prosopagnosia, with good insight and no other memory deficits; impairment of frontal abilities such as abstract categorization and reduced phonemic fluency.
- MRI (2012): left parieto-occipital atrophy.
- 3T MRI with spectroscopy (2014): prominent left parieto-occipital atrophy and a severe decrease in NAA levels in parieto-occipital regions.
- Positron emission tomography with FDG (2014): severe hypometabolism in occipito-parietal and posterior temporal areas bilaterally.
- Lumbar puncture: all CSF biomarkers, measured with Innogenetics kits, resulted in normal range (T-tau 83 pg/ml -nv <500 pg/ml; P-tau 36 pg/ml -nv <61 pg/ml; Aβ42 815 pg/ml -nv >500 pg/ml).
- ApoE genotype: ε3 ε3.

Conclusion: While most PCA case series report a clinical and anatomic predilection for atrophy in right over left hemisphere in MRI (Whitwell, 2007) and low liquoral Aβ42 levels, our case showed an atypical prominent left parieto-occipital atrophy and a CSF profile not typical of AD. In addition, our patient had ε3ε3 ApoE aplotype and signs of frontal lobe dysfunction at neuropsychological evaluation.
Further research is needed to identify which portion of PCA patients could present a pathologic substrate different from AD.

Fig. 1: Transverse, coronal and sagittal 1.5T brain MRI scan showing posterior cortical atrophy with no hyppocampal atrophy (2012).



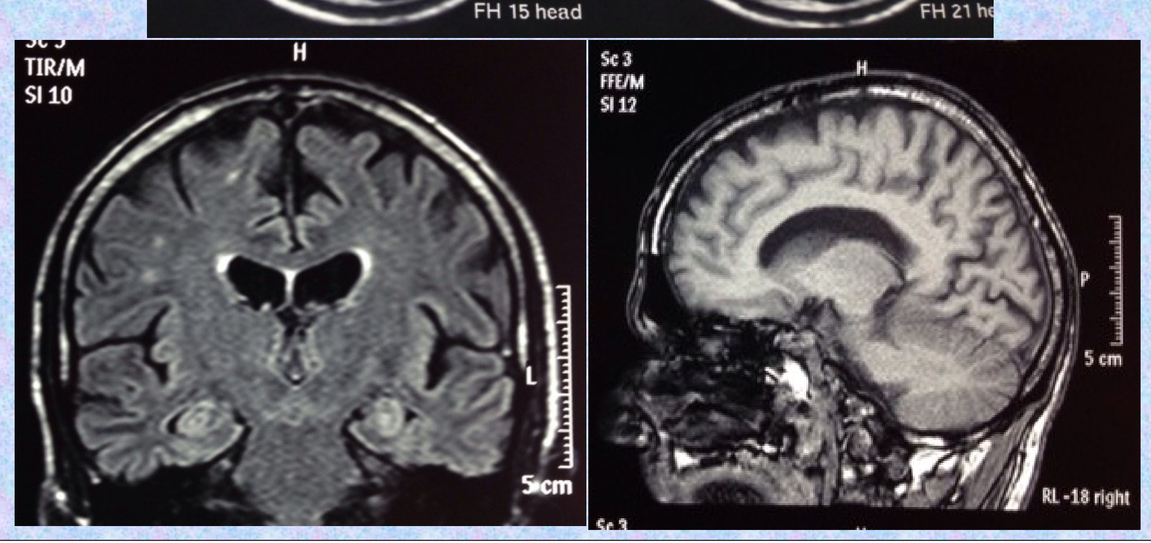
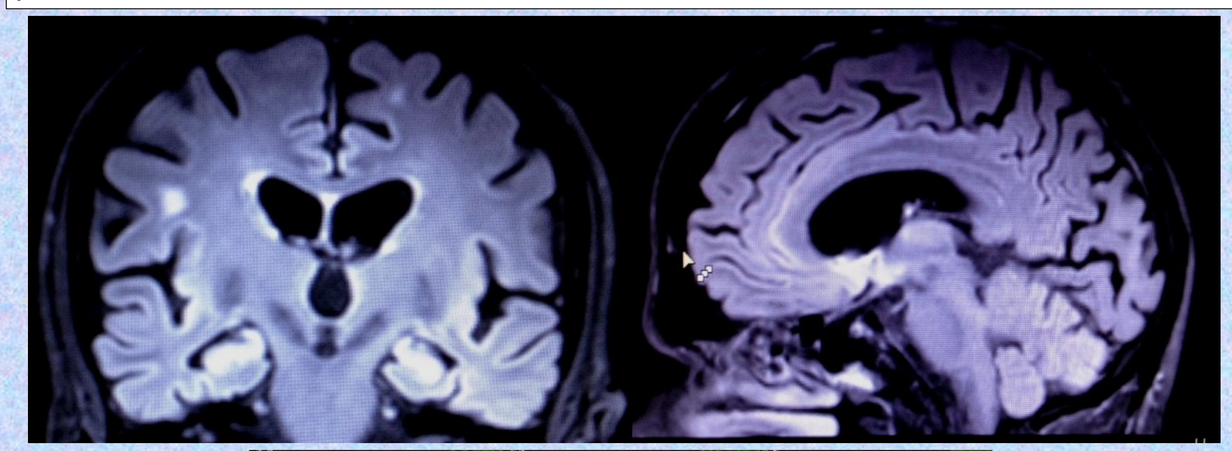
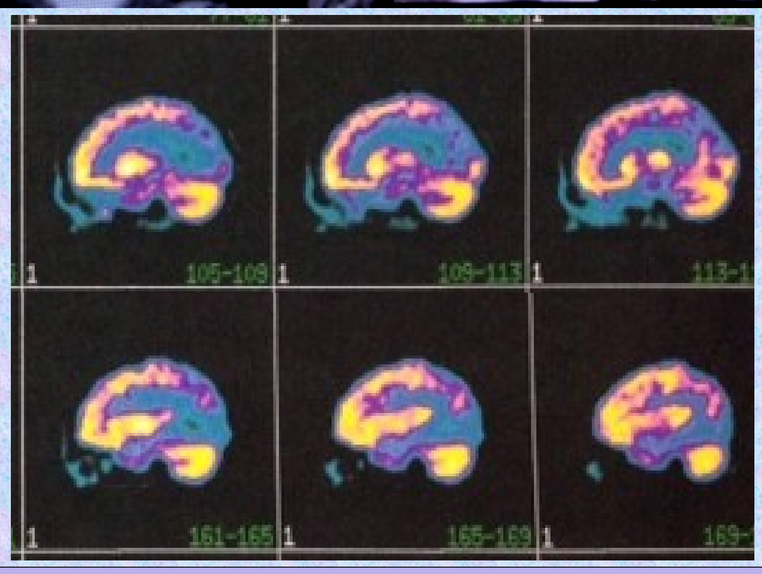


Fig.2: 3T brain MRI scan (2014) and FDG-PET (2014) showing an evolving pattern of atrophy (parieto-occipital and medial temporal lobes) and severe hypometabolism in posterior areas.





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