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A CASE OF POSTERIOR CORTICAL ATROPHY WITH COMPLEX SET OF SYMPTOMS AND RAPID COURSE

Tondo G, De Marchi F, Sacchetti M, Terazzi E, Cantello R.

Department of Neurology – University of Piemonte Orientale – Novara.

INTRODUCTION

Posterior Cortical Atrophy (PCA) is a neurodegenerative syndrome characterised by progressive occipital and parietal dysfunction, including visual symptoms (in the absence of ocular diseases), apraxia, alexia, acalculia, language deficit; myoclonus and extrapyramidal signs can be associated. Neuroimaging and functional imaging studies show characteristic posterior brain abnormalities. PCA is often related to Alzheimer Disease; less times other etiologies are responsible, such as Corticobasal Degeneration, Lewy Bodies Disease, and the Heidenhain variant of Creutzfeldt-Jakob disease.

CASE REPORT

A 62-year-old female came to our Department because of neurological disturbance since 3 month, presenting as visual disorientation; she was an elementary school teacher and she was temporarily away from work for severe anxious-depressive syndrome, because of her symptoms. Neurological examination point out a pure *Balint's Syndrome*, with optic ataxia, oculomotor apraxia, simultanagnosia.
After 8 month, in consequence of a marked clinical worsening, she was hospitalized in Neurological Clinic. Than we observed a complex symptomatology characterized by pyramidal (diffusely brisk reflex, bilateral Babinski sign) and extrapyramidal (resting and postural tremor, wrist trochlea) signs, visual disturbances, grasping reflex, myoclonus.

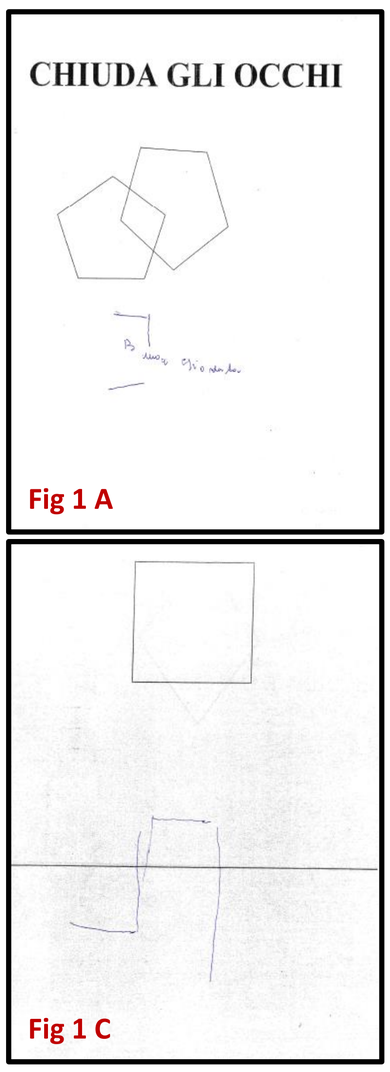

Fig 1 A

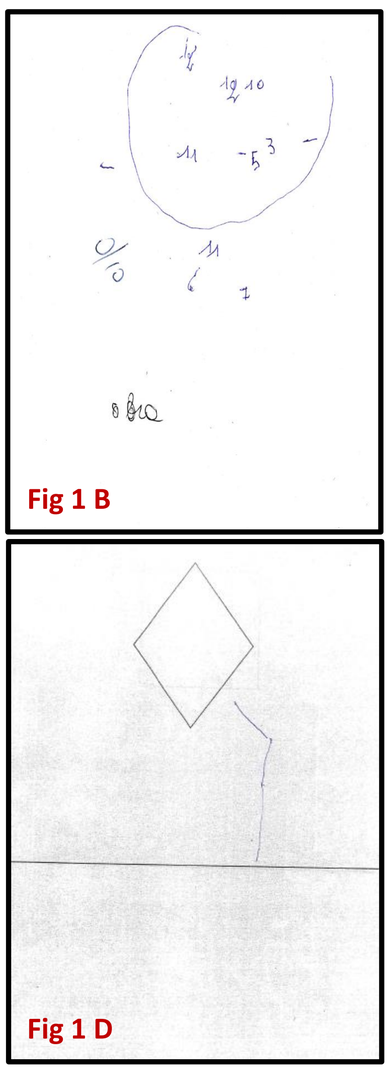
Fig 1 B

Fig 1 C

Fig 1 D

Fig 1 A MMSE showed a substantial integrity of spatial and temporal orientation and working memory, with falls in language and praxia. **Fig 1 B** Clock Drawing Test underlined a disorder of spatial knowledge, planning, executive function, and visuoconstructive skill. **Fig 1 C-D** By Test for constructional apraxia an early involvement of parietal and occipital cortex has been demonstrated.

Neuropsychological assessment evidenced moderate cognitive impairment (MMSE: 20/30 – **Fig 1 A**) and performance below normal in all tests administered, including tests to assess memory, attention and executive functions, with major compromising of visual, praxic and abstract reasoning functions (**Fig 1 B-C-D**).
CSF analysis remarked normal levels of 14.3.3 protein, tau and P-tau, low level of β-amyloid (317 pg/ml).
EEG was nonspecific, showing a diffuse slow activity, with occasional epileptic abnormalities in anterior regions.
Neuroimaging and functional imaging showed peculiar anomaly of posterior regions (**Fig 2 A-B**).

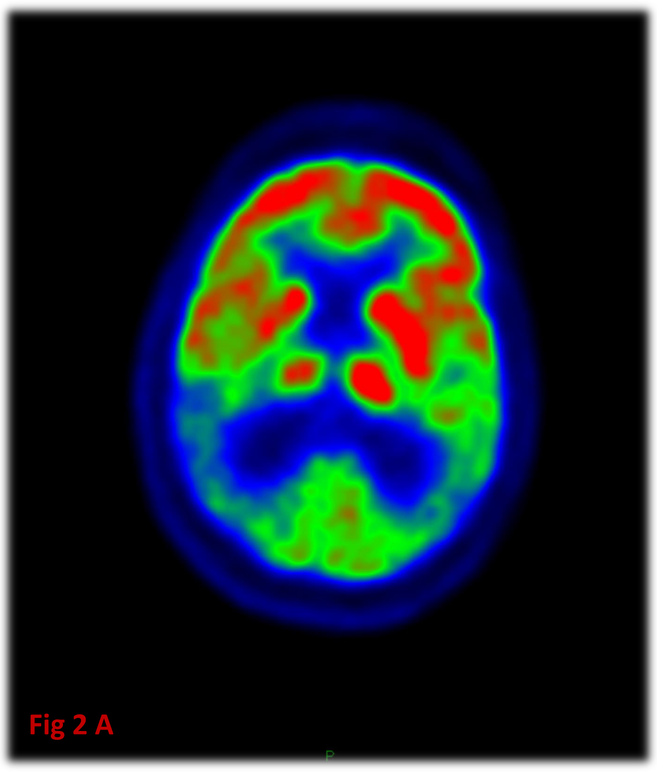
Fig 2 A

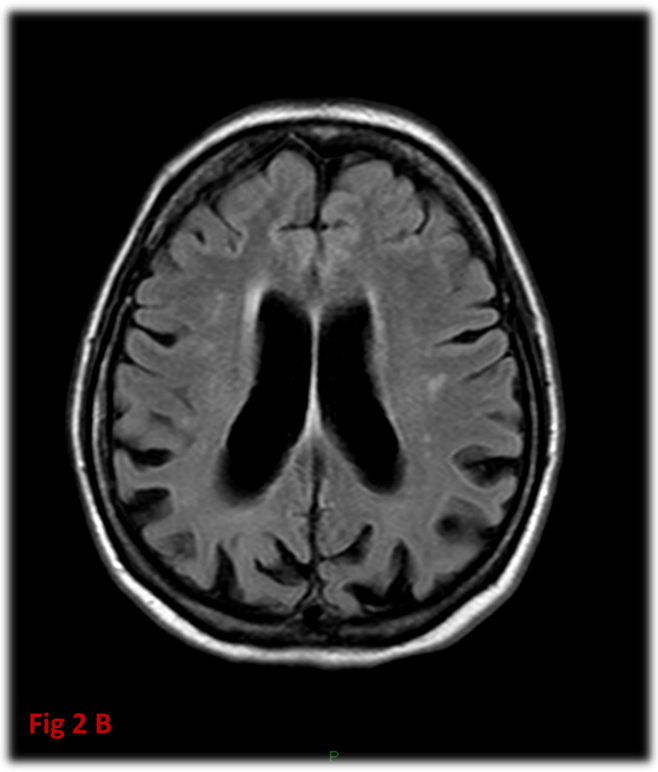
Fig 2 B

Fig 2 A Brain MRI (FLAIR sequences) showed bilateral posterior cortical atrophy. **Fig 2 B** Brain FDG-PET confirmed primary posterior hypometabolism.

On the basis of the clinical and neuropsychological features, supported by neuroimaging, according to diagnostic criteria, we diagnosed a PCA with unusual, very rapid, course.

CONCLUSION

PCA is a neurodegenerative disease pathologically related, in the majority of cases, to Alzheimer's Disease, as a visual variant; sometimes, PCA is associated with CorticoBasal Degeneration, Lewy Body's Disease, or Creutzfeldt-Jakob disease. This histopathological heterogeneity can reflected in a complex phenotype, that makes diagnosis difficult.
We describe a case of PCA, with typical visual symptoms, neuroimaging and CSF findings, unusual for the presence of less frequent (myoclonus) or even rare (tremor and grasping reflex) signs , as well as for a tumultuous course with a rapidly worsening dementia.

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