International Journal of Gerontology 15 (2021) 385–387
https://doi.org/10.6890/IJGE.202110_15(4).0018

International Journal of Gerontology

journal homepage: http://www.sgecm.org.tw/ijge/

Case Report

Posterior Cortical Atrophy, a Neurodegenerative Clinico-Radiological Syndrome: Neuropsychological, Electroencephalographic and Neuroimaging Study

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ARTICLE INFO

Accepted 2 March 2021

Keywords:
Alzheimer's disease,
dementia,
neuroimaging,
neuropsychology,
posterior cortical atrophy

SUMMARY

Posterior cortical atrophy (PCA) is a clinico-radiological syndrome that is initially characterized by visual disturbances and other posterior cognitive deficits, and late involvement of memory functions. This report details a case of a 58-year-old woman with 7 years of delay in the diagnosis and that began with alterations in reading and writing, later visuospatial alterations and visual agnosias were added and finally showed memory and language disturbances. Neurological, neuropsychological, structural neuroimaging and quantitative electroencephalography (qEEG) studies was performed. Neuropsychological evaluation showed severe reading and writing disorders, acalculia, right-left disorientation (Gertsmann syndrome) and ideomotor apraxia, but also showed attentional, mnemonic, language and memory alterations. Neuroimaging study showed generalized cortical atrophy with predominance in the posterior parietal regions, and qEEG showed marked decrease in the absolute power in all channels mainly in temporal and occipital derivations. PCA should be suspected in patients with unexplained progressive visual disturbances. Neuropsychological, neuroimaging and qEEG studies may be inexpensive and accessible tools to support the diagnosis.

1. Introduction

Worldwide it is estimated that there are over 24.2 million people with dementia, and that each year 4.6 million new cases are added.[1] The incidence of dementia cases continues to increase and this incidence is predicted to double every 20 years.[1] The posterior cortical atrophy (PCA) is a neurodegenerative clinico-radiological syndrome that was described in 1988 by Benson, and it is estimated that PCA syndrome occurs in 5% of Alzheimer's disease (AD) cases confirmed by autopsy, but other neuropathological etiologies have also been described in this syndrome, such as: Lewy body dementia, corticobasal degeneration and prion disease.[2,3] One of its main characteristics is visual impairment, which is a consequence of predominantly occipito-parietal and/or occipito-temporal atrophy rather than afferent visual deficit.[4]

The clinical diagnosis of the patient with dementia can be a challenge, especially when the patient have an early-onset dementia and when have an atypical presentation, which reflects in a significant diagnostic delay in some cases.[5] According to some studies, the frequency of AD patients with atypical clinical presentations is approximately 14%.[6] However, there is no reliable statistics on the matter and it is very important for the clinician to know and become familiarized with the uncommon clinical presentation forms of dementia to promote early diagnosis and treatment in these patients. In this sense, the neuropsychological assessment and the quantitative electroencephalogram (qEEG) can be inexpensive and useful tools to support the diagnosis.

2. Case report

A 58-year-old right-handed woman, graduated in social work, who worked as a special education teacher, with no family history of dementia or chronic diseases. Her condition started seven years ago with gradual progression of reading and writing problems: her hand-writing began to distort until it became incomprehensible, at the same time reading began to become slow, jumping letters, words or lines, losing the row or page, etc. During these first 3 years of evolution the patient did not present memory, behavior or language alterations. As these alterations in literacy increased affecting her work performance, so that she decided to retire. During this period of time, it was seen by different ophthalmologists and optometrists, and even psychiatric assessment without any diagnostic conclusion. Later, problems to locate and identify objects with her eyes and with the left hand were added. Recently (last year), she started to present temporal and spatial disorientation and recent-memory alterations. In the last months, her visual impairment worsened, requiring family support for most activities of daily living, and these activities are carried out only with the right hand; in addition, recently she has displayed periods of agitation and aggression, and even sporadic visual hallucinations.

On neurological examination, visual acuity could not be assessed, she presented no visual fixation or eye tracking; inconsistently she identified some shapes, objects or colors; the ocular fundus and pupillary reflexes were normal, no ophthalmoplegia or

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atavic signs were present. The results of the complete blood count, blood chemistry, serum electrolytes, thyroid profile, liver enzymes, treponemic test, lipid profile, levels of vitamin B12, and folates were normal.

For neuropsychological assessment, the *brief neuropsychological evaluation in Spanish* (NEUROPSI)[7] and *Barcelona's Test* were used. In the NEUROPSI test, the patient obtained a total score of 11 points (maximum of 130 points), which places her 3 standard deviations below that expected for her age and education, for which it was classified as severe alteration. Due to the evolution time of the patient (7 years), multiple cognitive deficits were observed in different cognitive domains, however, the predominant deficits and suggestive of PCA were: reading and writing disorders, acalculia, right-left disorientation (Gertsmann syndrome) and ideomotor apraxia (dressing apraxia), but also a severe deterioration in attentional, mnemonic and language functions was found. Particularly, changes were observed in the posterior attentional system, cortical blindness, signs of agraphesthesia, amorphognosis, asterognosis, and alterations in basic somesthetic functions (light touch and proprioception exploration strategies). Furthermore, we observed a severe anterograde episodic memory.

qEEG showed marked global decrease in the absolute power (AP), which is reflected in decreased density of the power spectrum, consisting of several peaks (generalized disorganization) (Figure 1 top). The relative power analysis indicated greater proportion of delta and theta rhythms with some alpha and a relatively higher proportion of beta rhythm. All these alterations were more evident in the posterior leads (temporal and occipital) bilaterally (Figure 1 bottom).

Brain magnetic resonance imaging study (MRI) showed generalized cortical atrophy with predominance in the posterior parietal regions bilaterally. No micro or macrovascular lesions recent or ancient, or changes in the subcortical white matter were observed (Figure 2). The diagnostic conclusion was posterior cortical atrophy (PCA). The patient was managed with anticholinesterase drugs, antidepressants and neuroleptics, received a cognitive stimulation program as well as a psychoeducational program for the patient and her family. Despite this, the patient continued to progress towards deterioration, developing immobility, progressive difficulties in swallowing until she died of pneumonia two years later; their relatives did not authorize the autopsy study.

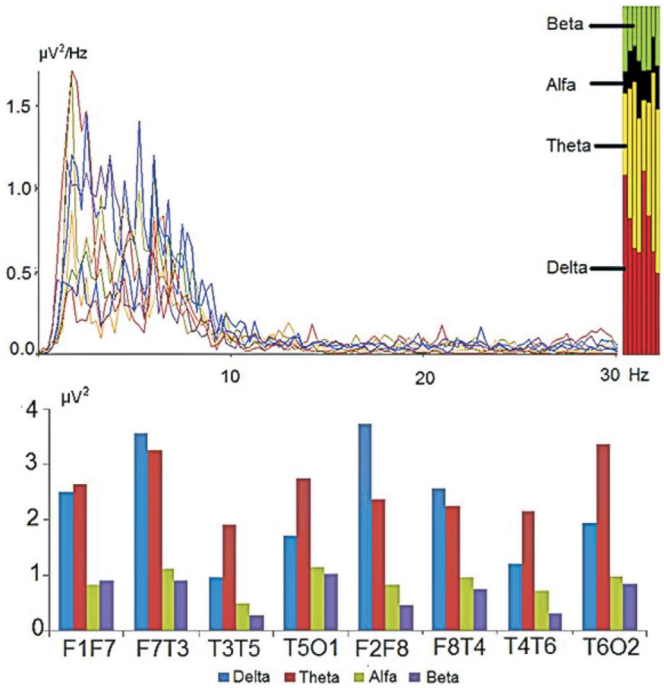

Figure 1. Shows the power spectral density and the relative power of four frequencies (top). The histogram (bottom) shows the absolute power of delta, theta, alpha and beta bands of frequency in the different electrode positions (F1F7, F7T3, T3T5, T5O1, F2F8, F8T4, T4T6, T6O2).

3. Discussion

The PCA is a clinico-radiological syndrome that was first described in 1988 by Benson of which diagnostic criteria and classifications have recently been proposed.[2] The core clinical features of PCA are: insidious onset, gradual progression and prominent early visual and other posterior cognitive functions. Among the most frequent cognitive characteristics are: space perception deficit, simultanagnosia, object perception deficit, constructional dyspraxia, oculomotor apraxia, alexia, acalculia, agraphia, etc.; for the diagnosis is required at least 3 of these cognitive characteristics in addition to a relative preservation of: memory, executive functions, behavior, personality and language functions (at the beginning of clinical picture).[2] In the case presented here, one of the initial symptoms was reading problems, which are considered one of the core clinical characteristics for the diagnosis of PCA, and which can be produced to either a true alexia or a combination of visual alterations present in this syndrome such as: simultagnosia, optic ataxia, ocular apraxia, macular crowding, and visual field alterations.[8] In this sense, recent research has shown that the spatial and perceptual components of reading such as the length of the words, the space between letters, their size and even the type of letter are decisive in the reading alterations that are observed in PCA.[9] However, although the pathology of AD is the most common finding in dementia syndrome, other pathological substrates have also been described: the Lewy body variant of Alzheimer's disease, Parkinson's disease with AD, dementia with Lewy bodies, corticobasal degeneration, and even prion diseases.[2] In the literature, descriptions of the electroencephalographic features of patients with PCA are scarce. However, within the few reported cases, alterations in brain activity have been described in posterior temporal regions, as well as bilateral temporoparietal dysfunction.[10,11] In this report, we present the case of a patient < 58-year-old with a predominance of involvement of the dorsal visual stream (biparietal) with a long delay in diagnosis (> 7 years) in which the qEEG showed severe functional impairment of cortical and subcortical neural assemblies especially at the parietal posterior area, which may represent the anatomical and functional substrate of her

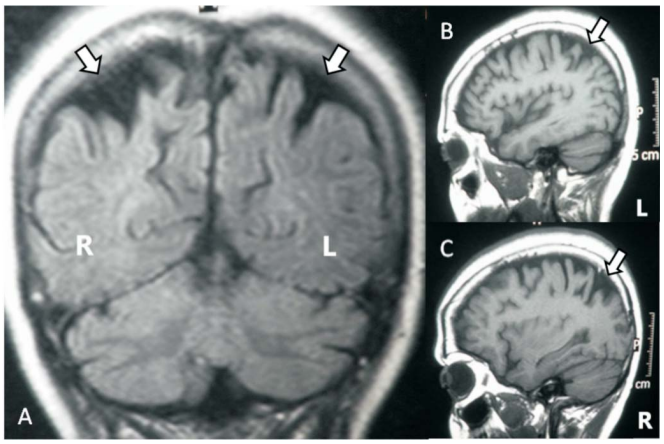

Figure 2. Structural MRI in coronal FLAIR sequence (A) and sagittal T1 sequence (B and C) showing bilateral atrophy, observed mainly in posterior parietal areas (arrows). L: left; R: right.

perceptual and cognitive disturbances as has been recently reported,[11] these EEG findings are important because the possibility of performing other types of cerebrospinal fluid (CSF) biomarkers or molecular neuroimaging studies may be very limited in developing countries, therefore the EEG analysis may be a very valuable tool to support the diagnosis of PCA, as has been previously demonstrated in other dementias such as Lewy body dementia.[12,13]

It is important to highlight some limitations of the present case, the most important one is that due to the long evolution time of the patient (7 years), the present clinical case are not those typically described in the diagnostic criteria of PCA, which are more focused on the prodromal or early diagnosis of the disease; another limitation is that in this case it was not possible to perform CSF biomarkers or positron emission tomography imaging due to the cost and lack of access to them in our country; on the other hand, it was also not possible to perform pathological confirmation by autopsy because the family not authorized, so the approach of the present case is done only at syndromic level.

Finally, it's important to mention, that the severe delay in the diagnosis in the case presented highlights the need for the clinician to be aware with the atypical manifestations of dementia in order to provide more timely treatment and thus improve the quality of life of these patients.

4. Conclusion

The PCA is an uncommon clinico-radiological syndrome in which qualitative visual disturbances are usually the initial symptoms, and in which may be observe a long delay in diagnosis like in present case; one of the diagnostic tools that have been used less in this entity is the qEEG, which has the advantage to be an accessible, economic, and non-invasive study who may be able to demonstrate anatomo-functional alterations in advanced stages of the disease as in the present case.

Ethics statement

The relatives of the patient presented here signed an informed consent to use and publish their information, maintaining their anonymity at all times.

Funding

None.

Conflict of interest

None.

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