Case Reports

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Posterior Cortical Atrophy With Interesting Presentation: Case Reports

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Citation Aghamollaii V, Fathollahzadeh Noor S, Bidaki R, Agah E. Posterior Cortical Atrophy With Interesting Presentation: Case Reports. Case Reports in Clinical Practice. 2021; 6(5):205-209.

Running Title Posterior Cortical Atrophy

Article info:
Received: 18 Sep 2021
Revised: 29 Sep 2021
Accepted: 19 Oct 2021

Keywords:
Agnosia; Atrophy; Alzheimer disease; Dementia

ABSTRACT

Posterior Cortical Atrophy (PCA) is a rare neuropsychiatric syndrome characterized by the dominancy of visuospatial impairments due to bilateral parieto-occipital divesting with the most common pathological changes presented in typical Alzheimer's Disease (AD). We reported 3 cases with similar diagnoses and interesting manifestations in this study. Gradual onset and progression of visual deficits without underlying ophthalmologic disease accompanied by intact memory and fluent speech and also the presence of simultagnosia, optic ataxia, oculomotor apraxia, dyscalculia, finger agnosia, and environmental disorientation, and the absence of any history of related diseases; all of them strongly suggesting posterior cortical atrophy.

Introduction

Posterior Cortical Atrophy (PCA) is a rare dementing syndrome characterized by the dominancy of visuospatial deficits due to bilateral parieto-occipital damage with the most common pathological changes seen in typical Alzheimer's Disease (AD).

Neurodegeneration in PCA tends to evolve earlier than typical AD, affecting people in their mid-fifties or early sixties. Usually, as the disease progresses, they will also experience symptoms of typical AD [1].

Inconsistent with the region of atrophy, this dementing illness is often heralded by impairment of higher visuospatial and visuoperceptual abilities. Patients eventually

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present components of Balint's or Gerstmann's syndromes, whereas in contrast to AD, insight, language, memory, and judgment are relatively persevered until late during dementia [1]. Psychiatric disorders, such as anxiety and depression, have been frequently reported, especially in the early stage of the disease; however, prominent psychotic features have been rarely reported except visual hallucination with about 25% prevalence [1, 2]. This paper aimed to highlight psychiatric features as a possible presentation of PCA. PCA is a posterior stream syndrome, which involves the occipitotemporal cortex over time [2]. Occasionally, patients may describe abnormalities with color vision, describing washes of color, or prolonged afterimages. This diagnosis is usually underestimated because of the loss of awareness. Its prevalence rate remains unknown [3].

Case Presentation

Case 1

A 51-year-old male with a history of cognitive impairment was referred to neurology service with a history of cognitive impairment. About 3-4 years ago, he presented nonspecific symptoms, such as anxiety and aggression, without other apparent symptoms. He was treated with Depakine 1000 mg and Risperidone 2 mg at night by referring to a psychiatrist; however, the signs were somewhat reduced.

About 2 years ago, he manifested symptoms, such as difficulty in reading, changes in the way he spoke and spoke little, and walked with short, slow, and uncertain steps, also dysfunction due to his inability to sew, which he was fired from work due to it. Since last year, the patient has been experiencing exacerbation in the symptoms of aggression, anxiety, and fear of dark environments and crowded spaces and has experienced severe agitation. His prayer was disturbed, and he forgot the sections of the prayer and did the wrong order of the steps of the prayer (sequence impairment). He has difficulty reading banknote numbers and often misses the buttons on his clothes.

He lost two fingers because of war trauma. He started using alcohol and opium some years ago.

The patient was then subjected to laboratory tests, neuroimaging, and neuropsychiatric evaluation. Laboratory studies were negative for inflammation or autoimmune diseases, thyroid diseases, metabolic disorders or deficiencies; serology for HIV/AIDS and syphilis, also cancer markers were negative.

His uncle was an AD patient with an onset age of 60 years.

Brain Imaging findings

Global cortical atrophy score was 0 due to sulci dilatation. The posterior-parietal atrophy rate was two due to parieto-occipital and posterior-cingulate sulcus dilatation and atrophy of precuneus lobule. The Medial Temporal Atrophy (MTA) score on both sides was equal to 1 due to choroid fissure dilatation. Midbrain diameter was 2.6 mm. Atrophy of the cerebellum with enlarged interfacial spaces was noted. White matter changes based on Fazekas rate was 1 (punctate high signal foci). No evidence of micro bleeding on lacunar infarction was detected in this study. Focal atrophy of bilateral parietal and occipital lobes cerebellar hemisphere was seen.

Neurological findings

Neurologic examination was remarkable with mild motor signs and myoclonus.

Case 2

A 45-year-old right-handed man with 12 years of academic education presented with 3 years of progressive visuospatial and neuropsychiatric symptoms. His initial complaint was difficulty seeing small complex writings, more prominent at night. After consulting with an ophthalmologist, he underwent cataract surgery; however, his symptoms remained progressing. In the following months, he started having trouble locating items, experienced spatial disorientation, and was lost several times, which led to extreme fear of going out alone. Subsequently, he demonstrated profound psychiatric symptoms, including depression with severe anxiety. He also experienced persecutory, paranoid, and somatic delusions, e.g. my brain is herniating out of my feet, and the delusion of reference; i.e., government and police control me. He also experienced visual hallucinations; he used to see his deceased mother in small size (just a few centimeters) walking around the food dish.

Furthermore, he used to see people passing by on the streets very tiny and scrawny, leading him to feel like they were starving and destitute and giving them his food and money. Auditory hallucinations were also present; he has claimed that he hears his host or police voices that threatened or ordered him. He lost his work at an installation service shop due to errors, difficulties in applying usual gadgetries, and psychiatric problems.

Accordingly, he was presented to a psychiatrist, and his psychiatric symptoms were reduced with an excellent

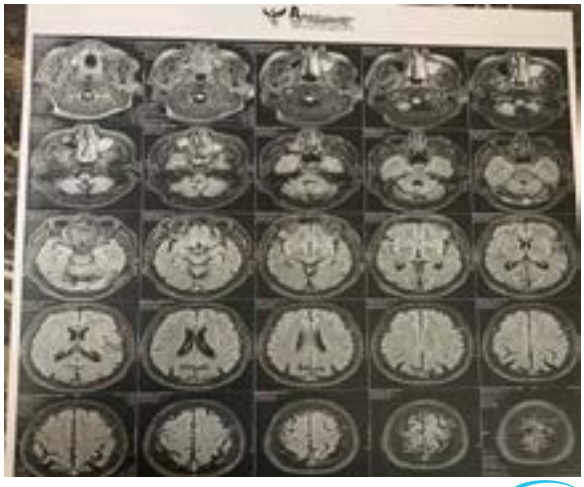

Figure 1. Obvious atrophy of bilateral posterior parietal and occipital lobes

response to antipsychotic drug treatment. He was referred to our neurologic clinic for further investigations.

On neurologic examinations, his Mini-Mental State Examination (MMSE) score was 24/30; he was fully oriented and had fluent speech and intact memory. His verbal memory was standard; however, his visual memory was significantly disturbed, and Brief Visuospatial Memory Test-Revised (BVMT-R) result was abnormal. He complained about the difficulty in estimating distances, and in the examination, simultanagnosia was noted, which was also reflected in his disability to distinguish between lines in a text. While watching TV, he mentioned that the images seemed to move indirectly and complained of after images. He also noted difficulty finding floors (i.e., instead of going downstairs, he went upstairs). All features of Balint's syndrome, simultanagnosia; optic ataxia; and oculomotor apraxia were presented. Acalculia and finger agnosia as partial Gerstmann's syndrome were also revealed. His visuospatial skills were deteriorating, and the Judgment of Line Orientation (JLO) test finding was abnormal; he also had problems with copying figure and clock drawing tests. Rather than constructional dyspraxia, ideomotor and dressing apraxia were also noted. His handwriting also deteriorated in structure. He had difficulties with tandem gait due to errors in estimating distances. Except for symmetric hyperreflexia, other neurologic examinations were otherwise normal.

Brain MRI investigations presented significant atrophy on posterior regions of the brain, including posterior temporal, parietal, and occipital lobes without pronounced asymmetry (Figures 1 and 2).

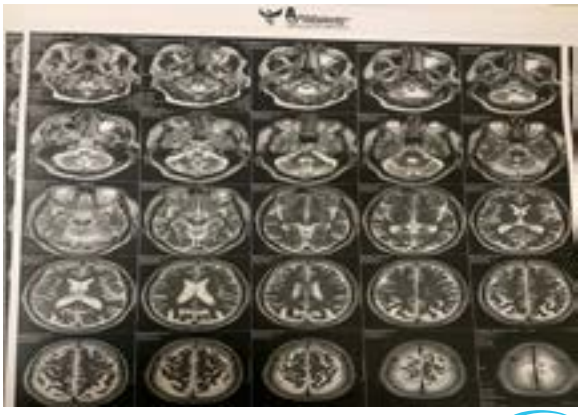

Figure 2. Obvious atrophy of bilateral posterior parietal lobes and cingulate

According to disturbing visuospatial skills, the patient has difficulty drawing the template and setting the clock at 11:10. (A) Cube shape template (B) Patient A's figure copying performance (Figures 3 and 4) (C) Clock drawing test (Figure 5).

Case 3

A 56-year-old woman with a car accident history was referred to our clinic. She had deteriorated course. She had a tension headache, agitation, word-finding difficulty, depression, and hopelessness. In the examination, we found dressing apraxia, slow motions, myoclonus, dysphagia, drooling, perseveration, visuospatial deficits, the decline in lexical resource, and right hemi parkinsonism as per the Hohen-Yahr Scale. Her sister was a bipolar patient. She had bruxism follow Quetiapine and Trazodone use. Drugs included Tab. Olanzapine 5 mg BID, Tab.Memantine 10 mg Daly, Tab. Depakene 500 mg BID, and Tab. Clonazepam 1 mg at night. Brain MRI data indicated Basal ganglia and posterior cortical atrophy. Hydrocephalus and aqua duct obstruction was not found (Figure 6).

Discussion

Gradual onset and progression of visual deficits without underlying ocular disease accompanied by intact memory and fluent speech and simultagnosia, optic ataxia, oculomotor apraxia, acalculia, finger agnosia, and environmental disorientation. The absence of any related previous disease, all of them strongly suggest Posterior Cortical Atrophy (PCA) in this patient according to proposed diagnostic criteria [1, 4, 5]; however, other underlying pathologies, such as Dementia with Lewy Bodies (DLB), prion disease, and corticobasal degeneration should be considered.

Figure 3. Visuospatial errors in copying and Clock Drawing Test (CDT)

The most prevalent psychiatric disorders or presentations reported in a patient with posterior cortical atrophy consisted of mood swings and especially anxiety disorders. Isella et al. demonstrated that the prevalence of behavioral and psychological symptoms is 95% in PCA patients, among which anxiety and apathy are the most common. They also declared that mood alteration is independent and separate from agitated behaviors [6]. It is suggested that these symptoms, i.e., more frequently observed in the early stage of the disease, are probably due to relative sparing of patients' insight into their unusual symptoms in this stage [1]. Our patient indeed experienced severe anxiety. After several environmental

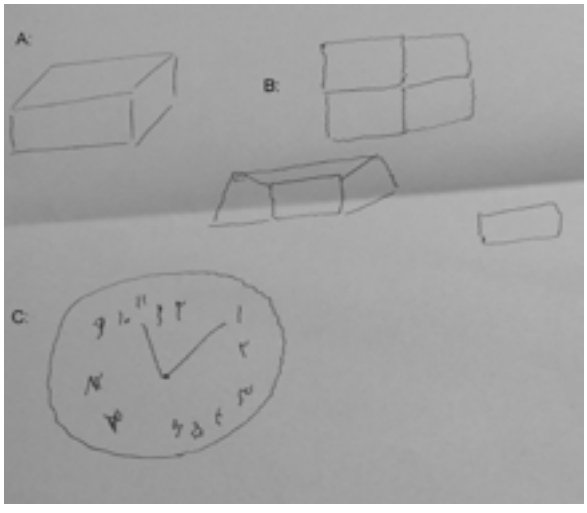

Figure 5. Copying figure and clock drawing test

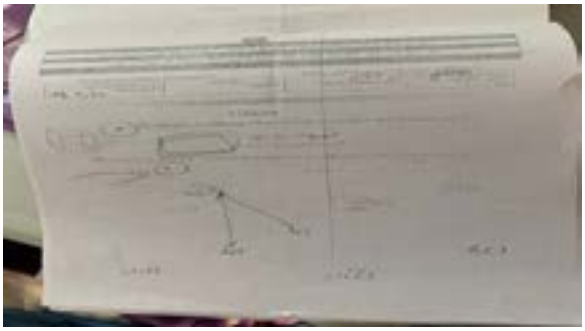

Figure 4. Severe abnormal cube copying test

disorientations, which led him to wander and ask for the police to help, his psychiatric symptoms exacerbated up to prominent psychosis, including complex visual and auditory hallucinations and deep persecutory, as well as somatic and referential delusions.

Great anxiety about his neurological signs could be an underlying basis for his delusions, especially paranoia and persecutory and even delusion of reference about control by police and government to the extent of having auditory hallucinations about hearing their voice.

Numerous patients with PCA present positive visuoperceptual symptoms, such as abnormally prolonged shape and color afterimages, reverse size phenomena, perception of the movement of static stimuli, and even 180° upside-down reversal of vision [1]. Thus, maybe seeing other people deformed long and thin and poor could be explained by unusual symptoms of an impaired visuospatial system; however, seeing his dead mother in the absence of an external stimulus is a visual hallucination which has been reported up to 25% of PCA patients [2, 5].

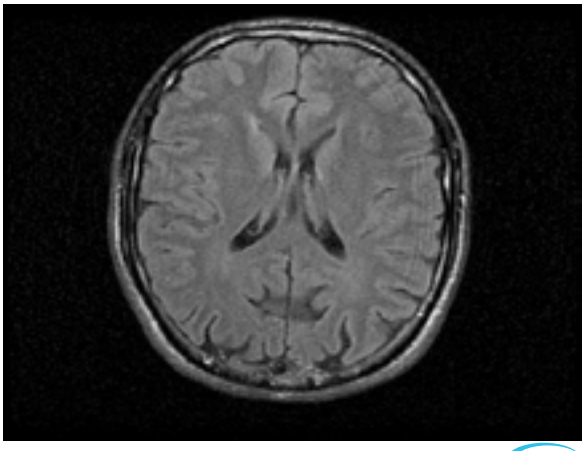

Figure 6. A brain MRI

Psychological signs other than visual hallucination have rarely been reported in PCA patients. We could find only a case with Frank psychosis, which had a significant response to the antipsychotic drug, like our patient [7].

As per Josephs et al., the clinical and imaging features of PCA patients, according to have visual hallucinations or not, were compared [8]. The results strongly suggest DLB as an underlying cause considering the higher incidence of Parkinsonism, rapid eye movement, sleep behavior disorder, and myoclonic jerks. In the present study, psychosis and cognitive decline can be attributed to Lewy body dementia. Still, lack of Parkinsonism, REM sleep behavior disorder, presence of typical Balint's and Gerstmann's syndromes and other PCA clinical and imaging criteria, and positive theatrical response to specific antipsychotic drugs are firmly against this diagnosis.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this study. The participants were informed of the aim of the research and its implementation stages. They were also assured about the confidentiality of their information and were free to leave the survey whenever they wished.

Funding

This research did not receive any grant from funding agencies in the public, commercial, or non-profit sectors.

Conflict of interest

The authors declared no conflict of interest.

Acknowledgments

The authors would like to thank all patients and their care givers for their participation in this survey.

Authors' contributions

Conceptualization and supervision: Vajihe Aghamollaii and Reza Bidaki and Setareh Fathollah Zade Noor; Methodology: Vajihe Aghamollaii and Reza Bidaki; Investigation, writing – original draft, and writing – review & editing, data collection, funding acquisition and resources: All authors; Data analysis: Vajihe Aghamollaii, Reza Bidaki and Elmira Agah.

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