

Benson's syndrome or Posterior Cortical Atrophy

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Abstract

Benson's syndrome or Posterior Cortical Atrophy (PCA) is a rare degenerative disorder clinically distinct from [Alzheimer's disease](#) (AD). Its frequency is unknown. PCA refers to a clinical syndrome in which higher order visual processing is disrupted owing to a neurodegenerative disorder. The patients present with progressive and severe visual agnosia (inability to recognize and identify familiar objects or persons) and apraxia (loss in the ability to execute or perform skilled familiar movements). Manifestations include simultanagnosia, optic ataxia, oculomotor apraxia, prosopagnosia, alexia, and environmental disorientation. Patients have also constructional and dressing apraxia, ideomotor apraxia, agraphia, acalculia. Memory, language, insight, and judgment are relatively preserved until late in the disease course. Brain Magnetic Resonance Imagery (MRI) shows bilateral atrophy in the parieto- and temporo-occipital areas that is more severe in right hemisphere. Brain single photon emission computed tomography (SPECT) or positron emission tomography (PET) show hypometabolism of the brain posterior areas. The most commonly associated neuropathology is that of AD. Deposition of neuritic plaques and neurofibrillary tangles are specifically revealed in posterior cerebral areas and sometimes in primary visual areas. Management of PCA is based on visual aids and antidepressant treatments.

Key-words

Benson's syndrome – Posterior Cortical Atrophy– Alzheimer's disease – agnosia – apraxia – dementia

Disease name and synonyms

- Posterior Cortical Atrophy (PCA)
- Benson's syndrome,
- Visual variant of Alzheimer's disease (AD),
- Progressive agnosia and apraxia.

Definition / diagnostic criteria

The term PCA refers to a clinical syndrome in which complex visual processing is progressively disrupted owing to a neurodegenerative disorder. PCA was first described by Franck Benson in 1988: his original report included 5 patients with progressive dementia presenting with early onset of complex visual difficulties (Benson *et al*, 1988). The most commonly

associated pathology being that of AD, some authors also use the term "visual variant of Alzheimer's disease" to describe PCA. It is however an easily recognizable clinical syndrome very distinct from the well-known and more common amnesic AD.

This degenerative disorder involves the posterior parts of the brain used in visual and gestural activities. Thus, the predominant symptoms are visual agnosia and apraxia. For many years, the patients have significantly better language, less memory difficulty, more depression and greater insight into their illness than in a typical AD. The mean age of onset observed in 15 PCA patients was 58 years (range 51-64) (Mendez *et al*, 2002).

Epidemiology

PCA is a rare disease and its frequency is unknown as no epidemiological study has been published.

In the Memory Clinic in Lyons, the percentage of new cases of PCA in 2001 was 4% (i.e., 6 cases out of 154 new dementing cases). The other cases corresponded to: AD (55%), Frontotemporal dementia (11%), Lewy Body Disease (5%), Primary Progressive Aphasia (5%), Semantic Dementia (3%), Vascular Dementia (3%), subcortical dementia (5%), Mild Cognitive Impairment (10%). Since the Memory Clinic is a reference center for rare degenerative disorders, the percentage of PCA cases is surely over-rated.

Genetic

To date, there are no genetic studies regarding PCA.

Etiology

Autopsied brains of PCA patients have shown the neuropathology of AD in most cases (Victoroff *et al*, 1994). Deposition of neuritic plaques and neurofibrillary tangles are specifically revealed in posterior cerebral areas and sometimes in primary visual areas. There are relatively fewer lesions in temporal and prefrontal cortex than typically observed in AD.

Damage to the dorsal occipito-parietal pathway (the spatial or "Where" pathway) gives rise to Balint's syndrome, apraxia and spatial disorientation, whereas damage to the ventral occipito-temporal pathway (the object identification or "What" pathway) gives rise to agnosia.

Whether PCA can be considered as the visual variant of AD or as an entity distinct from classical AD depends on the definition of AD. If AD is a progressive clinico-pathological disease beginning with memory disorders and associated to neuritic plaques and neurofibrillary tangles, PCA is not just AD with prominent visual deficits

but a distinct clinical syndrome. But, if AD is only a pathological disorder characterized by the presence of neuritic plaques and neurofibrillary tangles, wherever the cerebral localization and whatever the clinical symptoms, PCA is the visual variant of AD.

Nonspecific gliosis was reported in patients with PCA as well as in Creutzfeldt-Jakob disease (Victoroff *et al*, 1994; Tom *et al*, 1998). However, the symptoms and the course of the diseases are very different.

Clinical description

(Benson *et al*, 1988 ; Croisile *et al*, 1991; Aharon-Peretz *et al*, 1999 ; Didic *et al*, 1999 ; Croisile, 2001 ; Mendez *et al*, 2002)

The disease course is progressive over several years. The initial manifestations are outstanding visual agnosia and apraxia, which are predominant throughout the evolution of the disease.

Agnosia is a disorder characterized by an inability to recognize and identify objects or persons despite having knowledge of their characteristics. Patients with apperceptive visual agnosia have difficulty in recognizing the features of an object and consequently they can neither name it, nor use it. Visual agnosia typically results from damage to specific areas in the posterior lobes of the brain.

Apraxia is an impairment in the ability to execute or perform skilled learned (familiar) movements despite having the desire and the physical ability to perform the movements. Apraxia typically results from damage to specific areas in the parietal lobes of the brain.

Complex visual disorders seen in PCA patients are: elements of Balint's syndrome (optic ataxia, oculomotor apraxia), apperceptive visual agnosia, alexia, environmental disorientation (they are lost while driving or walking in familiar places), simultanagnosia (difficulty in global analysis of a picture), prosopagnosia (inability to recognize familiar faces), left hemineglect. Patients describe strange manifestations: they can more easily read small letters than big ones; they can more easily see remote objects than close ones. They describe a blurry vision when they are trying to read or observe an object. Sometimes, objects or letters suddenly move or disappear or pop-up when they are looking at them. Misreaching for object is frequent. All these visual manifestations are characteristic of visual agnosia.

PCA patients have constructional apraxia (inability to draw or construct simple configurations), dressing apraxia, ideomotor apraxia (inability to carry out symbolic movements), ideational apraxia (inability to create a plan for specific complex movements usually the use of objects and tools), elements of

Gerstmann's syndrome (agraphia, acalculia, right-left disorientation, finger agnosia). They have difficulties in sitting in a chair or in a car. While intending to drive, they climb into the back seat of the car. It can be impossible for them to use a key, a pencil, a razor. They cannot imitate movements.

Mild memory impairment is present early in the course of the disease although it is very different from the amnesic syndrome usually observed in AD. Thus, verbal and visual memories are compromised in memory tests, whereas memory in daily living is preserved for many years. Reasoning, judgment, and insight are all well preserved. In spite of naming difficulties, conversational speech is intact.

Progression of the disease

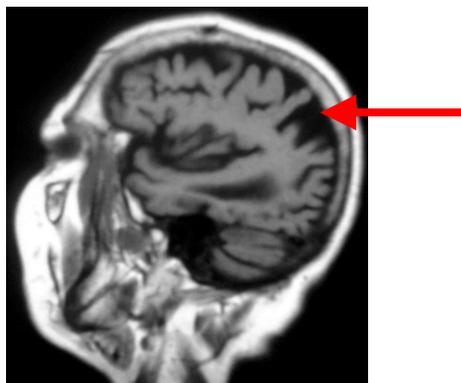
With time, there is a disabling progression of agnosia and apraxia. Visual agnosia and apraxia prevent PCA patients to recognize and to act upon the world, respectively. The behavior of the patient is that of a blind person. Then, there is an extension of the degenerative disorder in the anterior parts of the brain. Language and memory become more affected as well as reasoning. The course of the illness is typically from 8 to 12 years from the onset of symptoms to death.

Diagnostic methods

Neuropsychological examination is fundamental for the diagnosis. There is no biological marker of PCA. Examination of visual field sometimes shows visual field extinction or homonym lateral hemianopsia (more often right than left).

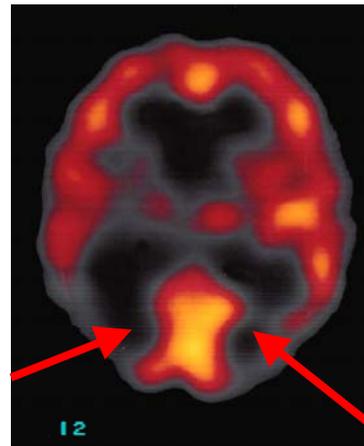
Computed tomography scan and Magnetic Resonance Imaging (MRI) demonstrate predominant bilateral parieto-occipital atrophy, more frequently in the right hemisphere (**Figure 1**). There is no detectable mesio-temporal atrophy as seen in typical amnesic AD.

Figure 1. Cerebral MRI in a case of Posterior Cortical Atrophy (sagittal slice): atrophy of the left parietal lobe.



Single-photon emission tomography (SPECT) and fluorodeoxyglucose-positron emission tomography (FDG-PET) show marked hypoperfusion or hypometabolism affecting the posterior cerebral hemispheres (Croisile *et al*, 1991; Aharon-Peretz *et al*, 1999; Nestor *et al*, 2003) (**Figure 2**). Occipito-parietal hypometabolism are usually worse in right than in left hemisphere.

Figure 2. HMPAO-SPECT in a case of Posterior Cortical Atrophy (horizontal slice): hypoperfusion of the parieto-occipital cortices.



Differential diagnosis

Visual variant of Creutzfeldt-Jakob disease (CJD): the course of CJD is more rapid than that of PCA; EEG and 14.3.3 protein are helpful in the diagnosis.

Parietal or occipital tumor: the symptoms are progressive but CT-scan and MRI are confirmative of a tumor in the posterior brain,

Occipito-temporal or occipito-parietal strokes: the abrupt onset is indicative of a vascular problem; CT-scan and MRI are confirmative of a stroke in the posterior brain.

AD (in its usual amnesic form) is clinically very different from PCA.

Management

The diagnosis of PCA has specific implications for management:

Visual and cognitive aids: referral to services for blind people, improvement of visual impairment through intervention of orthoptists, rehabilitation programs with speech therapists or neuropsychologists.

Treatment: Given the neuropathological similarities between AD and PCA, anticholinesterase drugs can be proposed, however, at least in France, there is no official authorization (specifically mentioning PCA) unless PCA is considered as the visual variant of AD.

Early treatment of depression is strongly recommended.

Unresolved questions

The exact prevalence and incidence of PCA remain to be estimated

The specific links with AD are yet unknown

The efficacy of anticholinesterasic therapies has to be investigated

Why is there a specific deposition of neuritic plaques and neurofibrillary tangles in posterior cerebral areas and sometimes in primary visual areas?

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