

Posterior cortical atrophy: A rare variant of Alzheimer's disease

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Abstract

Posterior cortical atrophy is a rare condition first described in 1988 involving progressive degeneration and atrophy of the occipital cortex, often recognized after an unexplained homonymous hemianopsia may be discovered. We report a case in association with Alzheimer's disease in a 77-year-old female, who underwent brain single-photon emission computed tomography as well brain positron emission tomography using Florbetapir to further evaluate progressive cognitive decline. The patient had also been followed in Ophthalmology for glaucoma, where a progressive unexplained change in her visual field maps were noted over one year consistent with a progressive right homonymous hemianopsia. This rare combination of findings in association with her dementia led to a detailed review of all her imaging studies, concluding with the surprising recognition for a clear hemi-atrophy of the primary left occipital cortex was occurring, consistent with Alzheimer's disease affecting the primary visual cortex. Further awareness of this disease pattern is needed, as Alzheimer's disease typically does not affect the primary visual cortex; other conditions to consider in general include Lewy Body dementia, cortico-basal degeneration and prion disease.

Introduction

In 1988, Benson described 5 patients with a previously unknown condition that selectively affected the visual cortex, producing a progressive hemianopsia on a degenerative basis.¹ Since then, over 340 publications have been generated on the condition with a progressively greater recognition for the problem as representing a variant of Alzheimer's disease for most cases, termed posterior cortical atrophy (PCA) but Lewy Body disease remain in the differential as does cortico-basal degeneration and prion disease.

We present a very interesting case of

progressive dementia in a 77 year old female suspected of having Alzheimer's yet was found to have an unexplained hemianopsia that worsened over a one year period where the initial crescentic lateralized defect grew into a complete homonymous hemianopsia. Careful retrospective review of her imaging studies shown below confirm the diagnosis of PCA

Case Report

The patient is a pleasant 77 year old female who presented to the Neurology clinic to evaluate unexplained events of syncope. Incidentally, she reported cognitive difficulty over the one to two year period prior to first clinic visit; family history was positive for a sister with progressive dementia. Also, she reported an episodic right visual field event, with the first episode occurring 6 months prior that was relatively sudden when there was an alteration in vision that persisted raising concern for possible retinal ischemic event. Formal report by Radiology did not remark on the hemi-atrophy shown below in the occipital cortex and had no acute ischemic changes evident.

She had been followed closely by Ophthalmology, and was found to have right sided homonymous hemianopsia on exam which was confirmed by visual field test and had brain MRI which did not show any clear lesion other than small multiple nonspecific white matter lesions but also showed an asymmetrically larger posterior horn of left lateral ventricle with more prominent atrophy of left occipital lobe. She was not aware of her visual symptoms but later did admit that she cannot see well on her right side at times; this still did not compromise her daily activities and did not feel there was a significant visual field deficit. Her husband also confirmed that she had not had demonstrated any clear symptoms of right visual field deficit that was obvious to him. However, concern developed about her cognitive impairment on bedside cognitive function tests as well as clear right visual field symptoms with corresponding left occipital area atrophy, prompting further testing including a nuclear medicine brain SPECT perfusion scan as well as brain PET using the Florbetapir amyloid tracer (performed under informed signed consent as part of a research protocol approved by institutional IRB for the IDEAS trial).^{2,3}

Humphrey field plots are shown in Figure 1; hemiatrophy of the visual cortex is shown by CT and MRI in Figure 2; Figure 3 reveals the functional defect for the left occipital cortex; Figure 4 shows no differences in the localization of the florbe-

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tapir tracer.

Despite a major significant finding on exam, the patient still personally feels there is nothing obvious to her about a major defect in her vision. Clinical diagnosis remains as Alzheimer's disease and has been given Donepezil and Memantine; additional new diagnosis established as Posterior Cortical Atrophy (PCA).

Discussion

This case illustrates the need to screen carefully for visual field defects in Alzheimer's Disease. Traditionally this is thought not to affect primary motor or visual areas, but there are clearly exceptions, with visual cortex involvement shown here in a selective fashion for the left occipital pole. Although not an issue for the patient here who had given up driving long ago, detection of visual field involvement in Alzheimer cases may suggest the additional diagnosis of PCA and prompt the need for revocation of driving privileges.

Recently developed diagnostic criteria for PCA have been published by Crutch and colleagues,⁴ with three stages identified to the disease, with the first stage being pro-

dromal/suspected/possible PCA. Due to the presence of cognitive impairment in this individual, the classification for this case would be Advanced PCA using the newly published criteria.

With regards to the findings of this study, an excellent publication by Day and colleagues is noted where five individuals cases of posterior cortical atrophy were studied in detail by PET.⁵ The investigators used not only the amyloid tracer employed here (Florbetapir), but also used a new PET tracer that binds to tau within the brain (Flortaucipir). Their study detected higher levels of Flortaucipir binding in early stage symptomatic PCA than those with early stage or elderly controls, with binding greatest in the posterior occipital cortex.

Their study confirmed the view that the posterior occipital cortices are especially prone to accumulate tau in PCA, and that Flortaucipir a sensitive method to evaluate for the possibility of PCA.

Genetic risk factors have been recently identified as well by Schott and colleagues.⁶ By studying 302 PCA patients and their genotypes, their study identified three loci linked to risk for the disease, in/near *CRI*, *ABCA7*, and *BINI*. The protein encoded by the gene *BINI* is widely expressed in the brain (especially in oligodendroglia) and is thought to represent a major risk factor for Alzheimer's disease and also serves as an adaptor protein that can bind c-myc, clathrin, and dynamin. *ABCA7* is strongly expressed in the brain, especially in

microglia and is involved in transporter proteins that move lipids across membranes. *CRI* is found on human red cells and is the immune adherence receptor that binds complement fixing immune complexes

Conclusions

All cases of suspected Alzheimer's disease should be carefully screened by clinical and radiographic examination to exclude PCA, as this represents a risk to the patient with driving and other related activities requiring fully intact binocular vision. Although rare, it is suspected that increasingly more frequently will this condition be

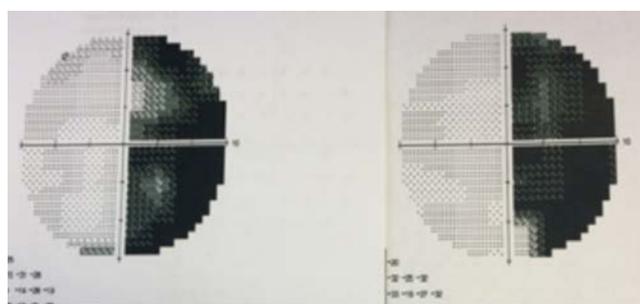


Figure 1. Visual field plots for the left eye (right hand column) and right eye (left hand column) for the baseline exam one year ago (bottom row), 6 months prior (middle row), and current exam taken 12 months after the baseline study, all confirming a progressive encroachment upon the right hemi-field, consistent with an evolving right homonymous hemianopsia over one year.

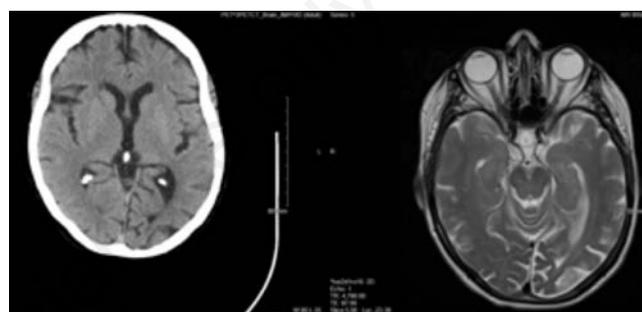


Figure 2. Left occipital cortex atrophy evident on computed tomography at left and magnetic resonance imaging at right.

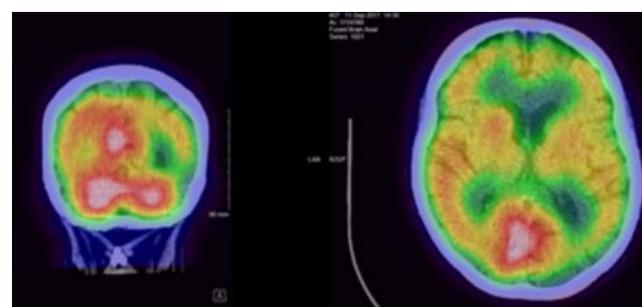


Figure 3. Left posterior parietal hypo-perfusion, characteristic to Alzheimer's disease with associated marked reduction in perfusion to the primary visual cortex as shown for the coronal view through the occipital pole at left and on the axial view at right.

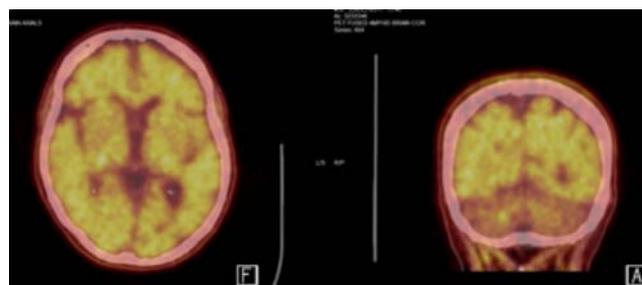


Figure 4. Positron emission tomography imaging with 18F-Flutetapir reveals no asymmetry in tracer localization.

identified with greater awareness to its existence and associated methods that may enhance detection including brain SPECT as shown here.

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