Asian Journal of Psychiatry xxx (xxxx) xxx–xxx

Contents lists available at ScienceDirect

Asian Journal of Psychiatry

journal homepage: www.elsevier.com/locate/ajp

Letter to the Editor

Posterior cortical atrophy variant of Alzheimer's dementia—A case report

ARTICLE INFO

Keywords:
Posterior cortical atrophy
Alzheimer' disease
Dementia
Atypical

ABSTRACT

Alzheimer's dementia (AD) is the commonest type of dementia presenting with initial episodic memory decline followed by involvement of other cognitive domains. Posterior cortical atrophy (PCA) is one of the variants of Alzheimer's dementia (AD) characterized by the atypical presentation of relatively persevered memory in the initial stage. PCA is an uncommon early onset dementia affecting adults between 50 and 65 years. It presents predominantly with visuo-spatial and visuo-perceptual deficits. PCA is a phenotype with varied etiology most common being Alzheimer's disease. The complex and atypical presentation with preserved memory and insight in patients with PCA poses challenge to clinicians in diagnosing at initial stages. There is also paucity of research on prevalence, course, prognosis and management of PCA. In this article we describe a middle aged gentlemen presenting with clinical features suggestive of PCA. We also discussed relevant literature.

1. Introduction

Alzheimer's disease is the most common neurodegenerative condition known to cause dementia (Ferri et al., 2005). Alzheimer's dementia typically presents with initial recent episodic memory deficits, word finding difficulty and later with apraxia, anomia, agnosia and visuo-spatial deficits (Burns and Iliffe, 2009; Förstl and Kurz, 1999). Alzheimer's dementia (AD) generally has onset after 65 years, generally progresses slowing for many years without any plateaus. Patients with AD usually have well preserved social cognition and insight about their cognitive deficits in the intitial stage (Ihara, 2016). Apart from the classical presentation of AD there are also atypical presentations of Alzheimer's dementia. Three atypical forms of AD are described in literature. These are frontal variant of AD, Logogenic variant of AD and posterior cortical atrophy variant of AD (Galton et al., 2000; Kawakatsu et al., 2017). Posterior cortical atrophy (PCA) is one of the important and less studied presentation of AD (Benson et al., 1988). Patients presenting with PCA are generally younger compared to those with typical AD. The clinical presentation initially would include visual symptoms without ocular disease, visuospatial deficits and visuo-perceptual deficits and later have memory deficits and language involvement (Mendez et al., 2002; Tang-Wai et al., 2004). These patients sometimes present with well-known syndromes such as Gerstman syndrome and Balint's syndrome (Charles and Hillis, 2005; Midorikawa, 2010). Patient presenting with PCA features will generally have delay in diagnosis due to atypical presentation. Few patients present to ophthalmologists due to visual symptoms. There is paucity of literature on the prevalence, course. Prognosis and management of PCA. In this article we describe about a middle aged gentleman presenting with PCA phenotype. We have also done relevant literature review.

2. Case history

A 50 year married gentleman with 10 years of formal education hailing from semi-urban background was referred by a physician to our facility for diagnostic opinion. Patient's younger brother who accompanied him reported complaints of calculation difficulties, unable to do regular work since 2 years and 6 months and forgetfulness since 1 year and 6 months. Patient was admitted for further evaluation and management. Patient premorbidly was well adjusted. Family history and past history were nil significant. Personal history revealed alcohol use since 20 years with quantity of 120 ml twice a week and was abstinent since 2 years. Nicotine use since 25 years in the form of smoking cigarettes in dependence pattern and abstinent since 1.5 years. On clarification of history, patient had insidious onset, gradually progressive cognitive decline since 2 years and 6 months. He initially started to have difficulties in calculations, handling finances and few months later developed difficulty in dressing. He later developed difficulty in judging the distances while riding car, when trying to grab an object made multiple tentative attempts before reaching. Subsequently, he developed difficulty in tracking sentences in newspaper and difficulty in navigating with visuospatial confusion, wearing foot wear wrongly and later progressive decline in reading and writing ability. From the past 1 year and 6 months, patient has developed recent memory deficits in the form of forgetting recent events, conversations, scheduled activities, misplacing his belonging, difficulty in recalling names of familiar people. For the last 1 year patient has reduced spontaneous speech, speaking only brief sentences and has difficulty in following complex commands. Since 1 year patient has given up driving car due to difficulty in handling multiple things required for driving and also due to navigational difficulties. On Physical examination patient's vital were stable. Nervous system examination showed nil focal deficits, no Parkinson's signs and cerebellar signs. On mental status examination patient had acalculia, dysgraphia, alexia, left to right confusion, dressing apraxia, poor constructional ability, visuo-perceptual deficits and simultognosia. Patient was also noted to have mild recent memory deficits, decreased fluency of speech. On standard cognitive assessments patients scored 14/31 on Hindi Mental Status Examination (HMSE) and 34/100 on Addenbrooke's cognitive examination (ACE-III). Patient's laboratory investigations were within normal limits as shown in Table 1. Fused F18 FDG PET CT images showed a. Asymmetric decrease in tracer uptake involving the parietal lobes involving the

https://doi.org/10.1016/j.ajp.2018.05.011
Received 8 May 2018

Table 1
Laboratory investigations.

S.No	Laboratory test	Results (reference range)
1	Haemogram	Within normal limits
2	Thyroid profile	
	T3	86.01 (87–178 ng/dl)
	T4	8.01 (87–178 ng/dl)
	TSH	3.70 (0.34–5.60 uIU/ml)
3	Renal Function test	
	Sr. Creatinine-	1 (0.81–1.44 mg/dl)
	Sr. urea -	19 (17–43 mg/dl)
4	Liver function test	Within normal limits
5	Vitamin B12	330 (180–914 pg/ml)
6	VDRL (blood)	Non-reactive
7	HIV	Non-reactive
8	CSF- analysis	Within normal limits
9	C-Reactive Protein	negative
10	Autoimmune encephalitis panel	negative

inferior parietal, supramarginal and angular gyrus (left > > right) b. posterior cingulate gyrus c, d. bilateral temporal lobes (left > > right) involving the parahippocampal gyrus, hippocampus and amgydala, e and f. precuneus (left > > right), cuneus, also involving middle and inferior occipital gyrus on the left side (Figs. 1 and 2). The diagnosis of Posterior cortical atrophy variant of Alzheimer's dementia was made. Patient was started on Donepezil 5 mg (p.o). Psychoeducation about illness to family members and home based cognitive retraining tasks were given to patient.

3. Discussion

The patient presented initially only with parietal symptoms in the form of acalculia, visuo-spatial deficits, and visuo-perceptual deficits and dressing apraxia. He also had simultognosia as early symptoms. Later when the disease progressed he had recent memory deficits and language involvement. Based on the pattern of cognitive deficits described above and the presence of severe hypometabolism of bilateral parietal lobe finding on PET-CT brain, a diagnosis of Posterior cortical atrophy was made as per the consensus classification by Crutch et al. (2017).

The typical age of onset of PCA is between 50 and 65 years which is younger compared to the common presentation of AD (McMonagle et al., 2006; Mendez et al., 2002). There are no population based prevalence studies of PCA. This might be due to lack of consistent criteria for PCA and also due to the rarity of this condition in elderly population. A study by Snowden et al. reported a prevalence of 5% among 523 consecutive AD patients presenting to their facility (Snowden et al., 2007). There is conflicting evidence for specific gender preference in PCA (Mendez et al., 2002; Renner et al., 2004).

PCA has distinct clinical features compared to AD and other dementia syndromes. In case series of 15 PCA patients described by Charles et al., 60% had visual symptoms and 46% had alexia. Nearly 53% of PCA patients also had memory decline (Charles and Hillis, 2005). In another study by Tang Wai et al. of 40 PCA patients, nearly 87.5% patients have one or more features of Balint's syndrome. Balint's syndrome features are simultognosia, ocular apraxia and optic ataxia. All the three symptoms of Balint's syndrome were present in only three patients. In the same study 61% of patients had one or more feature of Gerstmann syndrome that includes agraphia, acalculia, left to right confusion and finger anomia (Tang-Wai et al., 2004).

In PCA compared to typical AD the structural imaging there is predominant atrophy of parietal lobes and occipital lobes compared to medial temporal lobes. Studies reported asymmetrical atrophy of parietal lobes with right being more atrophied than left (Lehmann et al., 2011; Whitwell et al., 2007). Functional imaging (SPECT and PET) studies also reported similar areas of involvement as with structural imaging showing decreased metabolism of parietal and occipital lobes. In addition FDG-PET studies reported decreased metabolism in frontal eye fields and this is secondary to reduction in input from occipital lobes (Gardini et al., 2011; Kas et al., 2011; Nestor et al., 2003).In our case there is significant reduction in metabolism of bilateral parietal lobes.

PCA is a syndrome presenting with cluster of specific cognitive and unique radiological finding. The underlying pathogenesis for PCA differs from case to case. Majority of neuropathological studies done so far

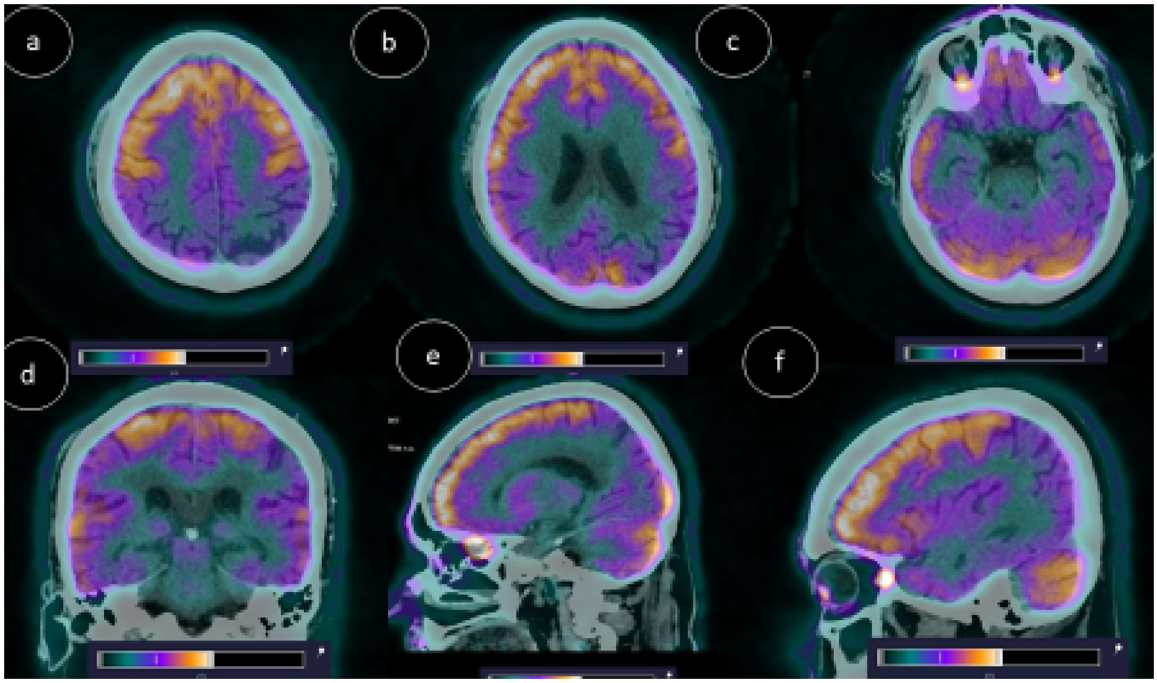

Fig. 1. Fused F18 FDG PET CT images show a. Asymmetric decrease in tracer uptake involving the parietal lobes involving the inferior parietal, supramarginal and angular gyrus (left > > right) b. posterior cingulate gyrus c, d. bilateral temporal lobes (left > > right) involving the parahippocampal gyrus, hippocampus and amgydala, e and f, precuneus (left > > right), cuneus, also involving middle and inferior occipital gyrus on the left side.