ABSTRACT

Behavioural and psychological symptoms of dementia (BPSD) are common in dementia. These symptoms differ with the type of dementia. Dementia with Lewy bodies (DLB) is a subtype of dementia characterised by visual hallucinations and sleep disorders.

INTRODUCTION

Dementia is an acquired global impairment of intellect, memory and personality without impairment of consciousness. The World Alzheimer Report 2016 estimated that there are approximately 46.8 million people living with dementia worldwide (Prince, Comas-Herrera, Knapp, Guerchet, & Karagiannidou, 2016). Dementias are classically divided as cortical and sub-cortical depending on anatomical location. Another classification which is based on aetiology, classifies dementia as reversible and irreversible. But the most common clinically used guidelines delineates dementia into 4 sub types – 1) Alzheimer’s dementia 2) Dementia with Lewy Bodies (DLB) 3) Vascular Dementia 4) Fronto temporal dementia.

In ICD-10 (International Classification of Diseases – 10) for a primary diagnosis, requires decline in both memory and thinking which is sufficient to impair personal activities of daily living for a duration of 6 months. In DSM-5 (Diagnostic and Statistical manual – 5), the word dementia has been replaced with neurocognitive disorders – major and mild. 6 domains are considered, such as complex attention, executive function, learning and memory, language, perceptual motor function and finally social cognition, out of which impairment of one domain is required to make a diagnosis of dementia.

The epidemiology of the dementia subtypes varies worldwide. Dementia with Lewy bodies is the second most common subtype of dementia worldwide with an estimated proportion of 10-15% of all cases (Borroni, Agosti, & Padovani, 2008). But in India, the prevalence is relatively less and it is the 3rd most common type of dementia accounting for around 5% of proportion of cases (Shaji et al., n.d.).

The international psychogeriatric association defines the Behavioural and Psychological symptoms in dementia as “signs and symptoms of disturbed perception, thought content, mood or behaviour that frequently occurs in patients with dementia”. BPSD (Behavioural and Psychological symptoms) in dementia is categorized under organic psychosis in ICD-10 whereas in the DSM–5 it is coded as a specifier. The signs and symptoms of BPSD can be classified into three subtypes- perceptual, affective and personality. The perceptual subtype includes delusions (23%-75%), hallucinations (15% to 49%) and misidentifications (20%-50%). The affective component includes depression (up to 80%), mania (3-15%) and apathy. The personality category comprises of personality change (up to 90%), behavioural symptom (up to 50%), aggression/hostility (up to 20%) (Lawlor, 2002). Various BPSD occur at different phases of illness. Research has indicated that these symptoms either: appear to occur increasingly as the dementing disorder progresses or, may occur more commonly during specific periods in the dementing disorder. Wandering and agitation have been shown to be the most enduring BPSD over a 2-year period. Untreated BPSD contribute to: premature institutionalization (Colyer & George, 1986; Sellers et al., 1998; Steele, Rovner, Chase, & Folstein, 1990), increased financial cost, decreased quality of life for both the caregiver and the patient (King, Collins, Given, & Vredevoogd, 1991), significant caregiver stress, stress to nursing staff in residential facilities (Rodney, 2000) and excess disability (Brody, 1982) i.e. people with BPSD function at a lower level than those without. Once symptoms are ameliorated or removed, functional level improves (reducing patient and caregiver distress and improving quality of life).

The BPSD in dementias varies according to the subtypes, for example agitation, apathy and depression are more common in Alzheimer’s dementia and vascular dementia whereas in DLB (Dementia with Lewy bodies), psychotic symptoms are the hallmark of presentation. In DLB, some patients initially present with parkinsonism alone, the frequent falls, orthostatic hypotension, transient disturbance of consciousness occurs. Episodes of confusion, progressive cognitive decline and it finally a full blown dementia occurs. The fluctuation in cognitive performances and functional abilities may be confused with transient ischemic attack. Visual hallucinations occur in about 2/3 of the patients with DLB. The hallucinations as described by the patient may be vivid, colourful fragmented figures of people and animal. The emotional response in return to these hallucinations may range from intense fear to indifference (may be due to apathy or parkinsonism) or even amusement.

The patient’s response to visual hallucinations may seem very realistic, for example they try to feed an imaginary dog, but insight is preserved in these individuals. Delusions may also occur and it may be secondary to the amnestic symptoms. Auditory hallucinations are least common, and secondary to BPSD or the dementia per se, depression can occur in about 40% of DLB patients.

CASE VIGNETTE

In this article we present a case of Dementia of Lewy Bodies. A 65/F, a housewife with no formal education living with spouse and children, belonging to middle socio economic status and a rural background was brought by her husband to psychiatry OPD with complaints of talking to self, irritability and aggressiveness for past 3 months. History revealed that patient had forgetfulness for the past one year. She initially had difficulty in remembering things and also often misplaced things. She used to forget to turn off the gas stove and switch off the lights. She used to misplaced things and blamed family members for her actions. She gradually started having difficulty in dressing and also remembering names. The
severity of her symptoms increased to an extent that she required assistance in day to day activities like bathing, eating and finding the restroom. The patient also had sleep disturbances with more of delayed onset and early morning awakening. She kept wandering at night and needed constant monitoring.

Gradually she used to pass urine in her clothes. She was also found talking to herself. Many times, she would be found making gestures and values enquired by family members, she used to say that somebody was sitting in front of her though others could not visualise them. Patient also had frequent unprovoked anger and was irritable towards family members. Symptoms were more profound during evening.

There was no history of head injury, seizures or substance use. Patient was a known hypertensive for the past 5 years and on regular treatment with Tab Amloidpine 5mg OD and the blood pressure was under control. There was no history of any other physical morbidity.

There was no past history of mental illness. There was no significant family history. Premorbidly, she was a well-adjusted, responsible, cheerful person and there was no history suggestive of any deviant personality traits.

General physical examination revealed no significant abnormalities. Pulse rate was 70 bpm and Blood pressure was 120/80 mm Hg. Fundus examination was normal. Systemic examination was also normal with regards to respiratory, cardiovascular and gastrointestinal systems. CNS examination revealed impaired higher mental functions. Mini Mental Status Examination score was 17 out of 25. Patient had hard of hearing but there was no focal neurological deficit.

On Mental Status examination, patient was alert, ambulant and cooperative. Rapport could be established with initially difficulty. Eye contact was adequate. Psychomotor activity was normal. During interview, patient was observed to scan the environment and make gestures. Hallucinatory behaviour was present. Talk was relevant and coherent. Quantum, tone and rate was decreased. Reaction time was prolonged. Affect was irritable especially towards family members. Patient had ideas of persecution. Visual hallucinations were present. Insight was absent.

With the above history and mental status findings, a provisional diagnosis of Dementia with psychotic features was made. According to ICD-10 F03.x2 Unspecified dementia, Other symptoms- predominately hallucinatory. This corresponds to DSM 5 diagnosis of Major Neurocognitive disorder with behavioural disturbance.

All investigations were done to rule out the causes and differential diagnosis of dementia such as delirium, endocrine causes, metabolic disorders, nutritional deficiencies, trauma, toxicity, HIV and infections.

**DIAGNOSIS AND DISCUSSION**

A final diagnosis of Dementia with lewy bodies was arrived based on the "Revised criteria for the clinical diagnosis of probable and possible dementia with Lewy bodies (DBL)" formulated in 2017 by the Dementia with Lewy Bodies (DLB) Consortium (McKeith et al., 2017).

According to the above cited criteria, the core features for diagnosing dementia with lewy bodies are

1. Fluctuating cognition with pronounced variations in attention and alertness.
2. Recurrent visual hallucinations that are typically well formed and detailed.
3. REM sleep behaviour disorder, which may precede cognitive decline.
4. One or more spontaneous cardinal features of parkinsonism: these are Bradykinesia (defined as slowness of movement and decrement in amplitude or speed), rest tremor, or rigidity.

**Indicative biomarkers are:**
1. Reduced dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET.
2. Abnormal (low uptake) 123iodine-MIBG myocardial scintigraphy.
3. Polysomnographic confirmation of REM sleep without atonia.

**By the criteria, Probable DBL can be diagnosed if:**

a. Two or more core clinical features of DBL are present, with or without the presence of indicative biomarkers, or
b. Only one core clinical feature is present, but with one or more indicative biomarkers.

In this case our patient satisfied two core clinical features, and hence a diagnosis of DBL was made. Indicative biomarkers could not be assessed because of the technical feasibility. The neurophysician was consulted regarding the DBL management and the BPDS was managed with tab.quetiapine 12.5mg at night initially. The patient is being followed up regularly.

This patient is being emphasized to emphasize the significance of the behavioural and psychological symptoms in dementia and to highlight the need for appropriate diagnosis in dementia. Since patients with DBL usually present with gross psychotic features rather than typical dementia features like amnesia or aphasia, there is a possibility of misdiagnosing and treating as pure psychosis. This can play a role in the progression and further management of the patient. In our case, the patient presented with primarily visual hallucinations but did not have aphasia or amnesia. Only on detailed evaluation, amnestic features could be elicited. MMSE aided in confirming a clinical diagnosis. This stresses the importance of a detailed history from patient and caregiver and mental status examination in elderly patients presenting with psychotic symptoms.

Although DBL ranks as second most common dementia in world next only to Alzheimer’s disease, the prevalence of DBL in India is less and it is only the 3rd most common dementia. A possible explanation could be a dearth of clinical criteria for diagnosis of DBL which may lead to classification of DBL as Alzheimer’s dementia. The recently released “Revised criteria for the clinical diagnosis of probable and possible dementia with Lewy bodies (DBL)” formulated by the Dementia with Lewy Bodies (DLB) Consortium offers a simple and effective means of diagnosing DBL.

Behavioural and Psychological symptoms in DBL not only causes a lot of morbidity the patient but also cause significant caregiver burden. Symptoms like visual hallucinations and episodes of inattention can be difficult to interpret. Some symptoms may require constant monitoring of patients. REM sleep behaviour disorders in patient can also affect caregiver’s sleep. Coping with behavioural and psychological symptoms can cause significant distress in patient’s caregiver (King et al., 1991). Psychoeducation of caregivers and early management of behavioural and psychological symptoms can significantly reduce caregiver burden.

Hence, this case report emphasizes the importance of using the latest “Revised criteria for the clinical diagnosis of probable and possible dementia with Lewy bodies (DBL)”. It also signifies the importance of management of behavioural and psychological symptoms in DBL.

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