

Charles Bonnet Syndrome: A Precursor To Alzheimer Disease

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ABSTRAK

Laporan kes ini mengetengahkan Syndrom 'Charles Bonnet' sebagai faktor dorongan pembentukkan penyakit neurokognitif utama disebabkan oleh penyakit Alzheimers di kalangan warga tua yang juga mempunyai masalah penglihatan dan juga diagnosis penyakit yang lain. Kami melaporkan kes tentang seorang wanita tua yang tiada masalah kesihatan dan perubatan buat pertama kalinya hadir ke klinik psikiatri dengan halusinasi visual kompleks di mana pesakit melihat imej manusia yang jelas dan objek selama 18 bulan. Selepas lebih kurang setahun, terdapat kemerosotan kognitif secara beransur-ansur dengan gejala psikotik seperti halusinasi suara dan delusi persekutori. Tiada gejala afektif atau obsessi dilaporkan. Beliau mempunyai kurang kesedaran terhadap penyakit beliau. Pemeriksaan mata menunjukkan kemerosotan akuiti visual di kedua-dua belah mata. Selain daripada itu, pemeriksaan fizikal adalah didapati normal. Beliau menerima rawatan ubat-ubatan Rivastigmine patch 4.6 mg/24 jam and Zydis 10 mg waktu malam. Kemerosotan kognitif dan gejala psikotik secara beransur-ansur pulih dalam tempoh masa 2 minggu sejak menerima rawatan dan juga rawatan susulan di klinik pesakit luar. Beliau juga semakin menyedari akan penyakit beliau. Syndrom 'Charles Bonnet' boleh menjadi faktor pencetus dan pendorong masalah penyakit neurokognitif disebabkan oleh penyakit 'Alzheimer's' di kalangan warga tua yang mempunyai masalah penglihatan yang sering terlepas pandang, atau tersilap diagnosis dan justeru itu kurang dilaporkan.

Kata kunci: penyakit 'Alzheimer's', Syndrom 'Charles Bonnet', halusinasi visual

ABSTRACT

This case report highlights Charles Bonnet Syndrome as a precursor to the development of major neurocognitive disorder due to Alzheimer's disease in the elderly with visual impairment and the possible differential diagnoses that could be considered. We report a case of an elderly lady with no known previous medical

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illness, who presented for the first time to the psychiatric clinic with complex visual hallucinations consisting of well-formed images of people and inanimate objects of long standing duration of 18 months. About a year later, there was emergence of cognitive impairment which progressed gradually accompanied by other psychotic symptoms such as auditory hallucinations and persecutory delusions. There were no affective or obsessional symptoms. She had poor insight towards her illness. Ophthalmological examination revealed significant reduction in bilateral visual acuity. Otherwise, physical examination was unremarkable. She received inpatient treatment with Rivastigmine patch 4.6 mg/24 hours and Zydis 10 mg noctre. Her cognitive impairment and psychotic symptoms gradually improved over a period of 2 weeks upon commencing treatment and on subsequent follow-ups at outpatient clinic. She had also gained some insight into her illness. Charles Bonnet syndrome could be a possible precipitating factor and precursor to the development of major neurocognitive disorder due to Alzheimer's disease in the elderly with visual impairment which is often overlooked or misdiagnosed and hence under reported.

Keywords: Alzheimer's disease, Charles Bonnet Syndrome, visual hallucinations

INTRODUCTION

Charles Bonnet Syndrome (CBS) is a condition characterised by the occurrence of complex visual hallucinations which consists of formed images of objects or persons, predominantly observed in visually handicapped individuals and those with normal psychology especially in elderly. Its aetiology and pathogenesis are not well understood. CBS increases with age of the individual and ophthalmological diseases (Jacob et al. 2004). The prevalence also increases and cerebral diseases which includes age-related macular degeneration, cataract, glaucoma, cerebrovascular disorders and Alzheimer's disease (Rovner 2006). Estimates of prevalence among older adults range from 0.4% to 27% (Shiraishi et al. 2004). This case illustrates the presence of complex visual hallucinations of long standing

duration in an elderly lady with visual impairment and later onset of cognitive impairment accompanied by other psychotic symptoms as well as the possible differential diagnoses that could be considered.

CASE REPORT

An 81-year-old lady was brought by her daughter-in-law to Psychiatric Clinic, Universiti Kebangsaan Malaysia Medical Centre (UKMMC) with complaints of visual hallucinations for the past 18 months. Her visual hallucinations consisted of images of men, women, children, babies and inanimate objects. The images were smaller in size than normal (liliputian hallucinations). She was also noted to be talking to herself since past 6 months whereby patient claim she was communicating with the images that she visualised. She would go to the

extent of preparing food and serving it to them and hosting the images that she visualised.

Her family members initially tolerated her behaviour but few months prior to clinic visit, patient started to visualise dead bodies in front of house and she insisted that her family members must perform prayers for the cremation ceremony. She would also wake up in the middle of the night, open the front door of her house and wait for the 'people' she visualised in order to serve them food. She also had persecutory delusion towards her maid whereby she accused her maid of trying to seduce one of the images of men that she visualised. There was history of forgetfulness since past six months whereby patient frequently misplaced her belongings and sometimes had difficulty recognizing her own family members but it was not of utmost concern to the family members.

Patient denied having any depressive symptoms. There were no symptoms suggestive of mania. No history of alcohol consumption. There was no family history of psychiatric illness or dementia. She had no known medical illness. There was history of bilateral cataract surgery done 10 years ago. At that time, she had presented with blurring of vision of both eyes which was worsening over a period of 3 years. Premorbidly, patient was an introvert type of person, had few friends, hardworking and preferred to stay indoors spending time with her family.

Mental state examination revealed an elderly lady, moderately built and neatly dressed. She had poor eye contact, was not forthcoming and

there was only superficial rapport established. She spoke in local Malay language. Her speech was of normal tone, rate and volume. Her speech was mostly coherent and relevant but it was irrelevant at times. Mood was euthymic and affect was congruent to thought. She had visual hallucination whereby she visualised images of people and inanimate objects. There was persecutory delusion towards her maid. She denied having any suicidal ideation. Cognitive assessment revealed that she was orientated to person but not to time, date and place. Her attention and concentration was poor. Her immediate, recent and remote memory was impaired. A Mini Mental State Examination score of 18 indicated moderate cognitive impairment. Her judgement was impaired and she had poor insight towards her illness.

Physical examinations were unremarkable. Ophthalmological examinations revealed reduction in bilateral visual acuity with measurement of 6/30 on right eye and 6/60 on left eye using Snellen chart. Neurological examinations were grossly normal. Her blood investigations were within normal range. Computerised Tomography (CT) scan of the brain was normal.

Patient was admitted for assessment and treatment. She was prescribed rivastigmine transdermal patch 5 mg once daily with olanzapine zydis 5 mg at night, which was then optimised to 10 mg at night a week later. Her behavioural and psychological symptoms showed improvement after two weeks of treatment. She also showed improvement in insight and activities of daily living on her subsequent follow-

ups at the Psychogeriatric Outpatient Clinic.

DISCUSSION

The diagnosis made clinically based on DSM V was major neurocognitive disorder due to Alzheimer's disease with behavioural disturbance. It is evidenced by the presence of significant cognitive impairment which progressed gradually accompanied with behavioural and psychological symptoms of dementia (visual and auditory hallucination with persecutory delusion). Delirium was unlikely in this patient as she did not have fluctuation in her consciousness level. Another possible differential diagnosis worth considering could be very late onset schizophrenia.

Eventhough CBS is a less frequently diagnosed condition; it is a rather common cause of complex visual hallucination. CBS has even been reported in elderly patients without visual impairment (Arya 1995). Its prevalence in individuals with visual impairment varies from 10% to 15% (Menon et al. 2003). CBS occurs in 1.85-3.5% of psychogeriatric patients referred to psychiatrists by general physicians, family physicians, and Ophthalmologists for visual hallucinations. CBS frequently remains undetected because of lack of awareness among doctors and patients' reluctance to admit to hallucinatory episodes, with the fear of being labelled as mentally unstable (Menon et al. 2003).

In this patient, the visual hallucinations were well formed, vivid and elaborate but pleasant to her, as described by de Morsier who stated that most of the

patients found the experience to be pleasant (Finucane 2006). This patient had experienced CBS for duration of a year prior to the onset of neurocognitive disorder and emergence of other psychotic symptoms such as second person auditory hallucinations and persecutory delusions. These psychotic symptoms and cognitive impairment gradually improved with antipsychotics and anti-dementia drugs which made the diagnosis of major neurocognitive disorder with behavioural disturbances a likely provisional diagnosis. In Alzheimer's disease, the visual hallucinations are more frequent than auditory while delusions are mainly paranoid, non-bizarre and simple type (Bassiony & Lyketsos 2003). A study by Pliskin et al indicated that patients diagnosed with CBS often manifest evidence of neuropsychological changes frequently associated with the early stages of dementia. Thus, CBS might be a predisposing factor to cognitive impairment, Alzheimer's disease or neurological deficit, or it may be an early marker of these conditions (Pliskin et al. 1996).

A differential diagnosis worth considering in this patient would be Schizophrenia (Very late Onset). The estimated one year prevalence rate of schizophrenia in individuals over 65 years of age is much lower than adults, between 0.1 to 0.5% (Copeland et al. 1998). Studies have shown that later onset is associated with a low prevalence of thought disorder and blunting of affect and a higher prevalence of visual hallucinations (Hafner 1998). This patient had predominantly positive symptoms of schizophrenia such as

visual and auditory hallucinations with persecutory delusions as well as cognitive impairment. However, in this patient, major neurocognitive disorder is a more likely diagnosis than very late onset schizophrenia as auditory hallucinations in the latter are more common than visual, delusions are more bizarre and complex, cognitive deficits are less severe, past history of psychosis is more common, symptoms are more persistent and the patient needs a long-term higher dose of antipsychotic drugs (Wetherell & Jeste 2004) as compared to dementia with behavioural disturbances. This patient showed favourable response to low dose antipsychotic and anti-dementia drugs.

Even though several cases of CBS were reported in the literature, CBS is still often overlooked or misdiagnosed and hence under reported. Therefore, this case report highlighted the importance of CBS as a possible precursor to the development of major neurocognitive disorder, especially in the elderly population with impaired vision.

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