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Charles Bonnet Syndrome: Rapid Resolution of Visual Hallucinations with Low Dose Risperidone

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Abstract

Charles Bonnet syndrome (CBS) comprises of complex visual hallucinations secondary to visual impairment in the presence of preserved cognition. Age related macular degeneration and other factors causing visual deterioration lead to visual hallucinations in 10-15% of patients. Sensory deprivation and social isolation are risk factors for development of CBS. The distressing hallucinations are underreported by patients and to date there are no robust evidence based guidelines to manage these hallucinations. Since visual hallucinations are associated with multiple psychiatric disorders as well, these patients are often referred to mental health providers. Here, we present the case of CBS development after macular degeneration. Rapid and sustained resolution of visual hallucinations and associated distress was observed with low dose Risperidone.

Keywords: CBS; Visual hallucinations; Pharmacology

Introduction

CBS is an underdiagnosed syndrome with prevalence rates as high as 15% [1]. The syndrome is defined by the presence of complex visual hallucinations in visually impaired individuals in the absence of cognitive deficits or other major psychiatric issues. The patients are insightful regarding the artificial nature of the visual hallucinations. The cluster of symptoms, seen more frequently in elderly individuals are often underreported. According to Lange *et al.* (2007) [2] up to 63% of patients with CBS worry about being called crazy. CBS management lacks firm evidence base regarding effective treatment and requires interdisciplinary collaboration.

Presence of visual hallucinations and distress related to their complexity prompts referral to psychiatrists in addition to ophthalmology follow up. This manuscript will highlight an interesting case of CBS which responded to low dose Risperidone resulting in rapid and sustained resolution of visual hallucinations. Furthermore, the discussion will provide concise guidelines for psychosocial and pharmacologic management of patients with CBS.

Case Report

Ms. A is an 88-year old Caucasian female with a six year history of age related macular degeneration(ARMD) who had been living alone since the death of her husband four years ago. She presented to inpatient psychiatric facility after transfer from a medical floor where she was admitted secondary to visual hallucinations, mild paranoia related to the visual hallucinations and severe distress secondary to the hallucinations. The patient and family reported that visual hallucination experiences were on and off through the day and lasted for hours at a time. While the exact visual acuity could not be determined during her psychiatric hospitalization, the patient and her family confirmed that she had presbyopia and myopia for which she used corrective glasses with good response. She had experienced similar symptoms a year ago and achieved complete remission of symptoms with 1 mg of Risperidone at that time. She had stopped taking the Risperidone a few months prior to this admission and soon afterwards re-experienced symptoms but did not share them with anyone secondary to embarrassment. A comprehensive psychiatric evaluation ruled out other psychiatric causes leading to paranoia. The classic presentation of complex visual hallucinations with age related macular degeneration in the absence of other medical or psychopathology were deemed diagnostic of CBS. The patient was initiated on a trial of 0.25 mg of Risperidone at night while on the medical floor which resulted in minimal relief of symptoms and patient was eventually transferred to the psychiatric facility for further management.

Prior to transfer to the psychiatric facility multiple neuroimaging tests were done to rule out other organic causes for her symptoms. Her neurology workup including CT head & face, EEG and MRI head were all within normal limits. Other workup including Ammonia, urine drug screen (UDS), C-reactive protein, Vitamin B12, Rapid plasma reagin, Hemoglobin A1C and Comprehensive metabolic profile were also within normal limits. Her EKG showed mild QTc prolongation at 439 but she was otherwise asymptomatic. Her thyroid stimulating hormone (TSH) was mildly elevated secondary to poor compliance with levothyroxine; it was rechecked multiple times over the course of her admission to rule out psychotic symptoms secondary to hypothyroidism and was found noncontributory.

On presentation, the patient stated that she has been very distressed secondary to visual hallucinations which recurred after discontinuation of Risperidone. Pt did have clear insight that the hallucinations were unreal and maintained good insight throughout her hospital stay. Her mental status exam including memory, focus and concentration was normal. Patient stated that she was seeing cartoons and pastel colors on the walls; she stated that the vivid pastel colors had rock like patterns and she could see them on multiple surfaces. She expressed severe distress related to these symptoms and stated that at home she hid herself in the closet for safety due to fear and paranoia related to the hallucinations.

During the hospital course, she received supportive, individual & group therapy with psychopharmacologic management which comprised of Risperidone titration to achieve maximum therapeutic benefit. The therapies focused on engaging the patient in activities of daily living, strategies to engage in community and decrease sensory deprivation, coping skills to improve frustration tolerance and cognitive strategies to reframe negative thoughts. The comprehensive treatment plan resulted in rapid and sustained relief of symptoms. Treatment team also contact family members who verified the patient's report of symptoms. They were engaged in the family therapy sessions and received psychoeducation regarding the possible effects of social isolation, grief and lack of social support in the development of CBS symptoms. They remained engaged as the Risperidone dose was slowly titrated up and discharge planning ensued. Patient was initially given twice daily dose which was later changed to 1 mg every night secondary to complaints of sedation. The once daily dosing was well tolerated by the patient. She was discharged home after complete remission of symptoms within a week. The patient continued outpatient follow up for several months with sustained resolution of symptoms.

Discussion

Complex visual hallucinations are seen in about 10-15% of individuals following reduced visual acuity [1]. Age related macular degeneration predisposes elderly individuals to develop CBS secondary to vision loss.

ARMD is the leading cause of blindness in people older than 50 years. It is a degenerative disease of the macula leading to painless loss of central vision, metamorphopsia and scotomas. The pathogenesis is poorly understood but the risk factors like increasing age, Caucasian race, family history, smoking, hypertension, cardiovascular risk factors and obesity seem to play an important role.

Other risk factors for CBS include social isolation, loneliness, decreased extroversion and shyness [3]. Experience of loss has also been implicated in the development of CBS. Poor quality of social interactions is also considered a contributory factor. Sensory deprivation and changes in the visual cortex functioning underlie development of visual hallucinations [4].

Spontaneous activation of the occipital cortex after vision loss and increased activity in fusiform gyrus has been postulated as the core neurobiological mechanisms underlying CBS [4]. Another theory postulates that loss of inhibitory mechanism in occipital cortex to eliminate undesired images from conscious perception might contribute to development of visual hallucinations [4]. Asymmetric blood flow in the temporal lobes has also been implicated in development of visual hallucinations of CBS. CBS affects both genders equally and mean age of onset is over 75 years. Only 15% of patients reports symptoms to medical professionals while 21% do not disclose them to anyone due to embarrasement [4].

Comorbid issues in advanced age pose another challenge for timely and accurate diagnosis. Interdisciplinary approach can reduce likelihood of fragmented care and avoid unnecessary tests. Ophthalmological, neurological and psychiatric evaluations are critical to rule out other organic causes. Management strategies include psychosocial and pharmacological modalities with management of comorbidities. Psychosocial interventions must encompass supportive therapy, social support to decrease isolation, grief or interpersonal therapy to alleviate the untoward effects of loss and reality testing exercises. Additionally psychoeducation about the organic nature of CBS hallucinations can likely reduce subjective distress. Family support can be garnered by educating family members about the nature of the disorder. They can help patient by improving lighting in the living area, reminding them to blink eyes during distress, reassurance, asking questions to improve reality testing and appropriate social interactions [5,6].

Pharmacological interventions target the hallucinations and comorbid psychiatric issues and mainly comprise of antipsychotic medication use for hallucinations and serotonergic agents to alleviate depressed mood or anxiety.

To date there are no randomized placebo controlled clinical trials for CBS management. Pharmacotherapy options include atypical antipsychotics, antiseizure medication, SSRIs, ondansetron and Diazepam to manage the symptoms [7-10]. Atypical neuroleptics have shown promise in a few case reports as their D2 receptor blockage eliminates hallucinations. Risperidone blocks 5-hydroxytrytamine-2 ($5\text{-}HT_2$) receptors in addition to D2 receptors affecting dopaminergic as well serotonergic neurotransmitter systems resulting in rapid relief of hallucinations as well as decrease in subjective distress and anxiety. Risperidone has been used for management of CBS symptoms with favorable outcomes for some time [7-10]. and the literature is suggestive of timely alleviation of CBS symptoms with Risperidone.

Conclusion

In our case, the rapid and sustained relief of distressing visual hallucinations with administration of Risperidone, substantiates the growing body of successful clinical cases following use of atypical neuroleptics in CBS along with the above mentioned psychosocial supports.

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