Case Report

Visual Hallucinations (Charles Bonnet Syndrome) Associated with Neurosarcoidosis

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ABSTRACT

The Charles Bonnet syndrome (CBS) refers to lucid and complex visual hallucinations in cognitively normal patients with acquired vision loss. It can be associated with any type of vision loss including that related to macular degeneration, corneal disease, diabetic retinopathy, and occipital infarct. Neurosarcoidosis, a multi-systemic inflammatory granulomatous disease affecting both the central and peripheral nervous systems, is rarely associated with CBS. We report a patient with biopsy-confirmed neurosarcoidosis who experienced visual hallucinations following the development of a right seventh-nerve palsy, right facial paresthesia, and bilateral progressive visual loss. This case highlights the importance of recognizing that the CBS can occur in visual loss of any etiology.

Key words: Charles Bonnet Syndrome, Neurosarcoid, Sarcoid, Visual Hallucinations

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INTRODUCTION

The Charles Bonnet syndrome (CBS) refers to lucid and complex visual hallucinations in cognitively normal patients with acquired vision loss. It can be associated with any type of vision loss including macular degeneration, glaucoma, corneal disease, diabetic retinopathy, and occipital infact.[1] CBS is also associated with older age, social isolation, decreased quality of life, and a history of stroke.[1] Neurosarcoidosis is a multi-systemic inflammatory granulomatous disease affecting both the central and peripheral nervous systems.[2] We report a patient with biopsy-confirmed neurosarcoidosis who experienced CBS. To our knowledge, this is the first such case reported in the English language ophthalmic literature.

CASE REPORT

A 46-year-old African American female presented in March 2010 with a 1-year history of bilateral progressive vision loss accompanied by right facial paresthesia in the V2 distribution, a right seventh-nerve palsy, eye pain, and photophobia. Over the next several weeks, the patient developed lucid visual hallucinations consisting mostly of an unfamiliar little girl. The images did not speak to the patient nor did she attempt to interact with the patient. The patient recognized that the hallucinations were not real and she was not disturbed or frightened by them. The hallucinations were not accompanied by any delusions, paranoid ideation, mental status changes, seizure activity or change in the level of consciousness. Past medical history was significant for a colonoscopy in 1990. The patient was not on any medications and denied the use of hallucinogens (i.e., anti-psychotics, barbiturates, sedatives, illicit drugs). The patient had been treated with topical antibiotic drops for her eye pain without relief. She had no prior psychiatric history.

On neuro-ophthalmic examination in March 2010, the patient was awake, oriented, and alert, but continued to complain of visual hallucinations. Best corrected visual acuity was 20/200 in both eyes. Pupils were 4.0 mm in the dark and 2.0 mm in the light with a left relative afferent pupillary defect and light-near dissociation in both eyes External examination revealed right

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partial peripheral facial nerve palsy. Motility and intraocular pressure exams were normal. Slit lamp bio-microscopy was normal without uveitis or granuloma formation. Dilated funduscopic examination showed bilateral optic nerve atrophy with temporal pallor in both eyes. Automated (Humphrey 24-2) visual fields revealed a cecocentral defect in the right eye and a central defect with superior arcuate nerve fiber layer defects in the left eye. Optical coherence tomography of the optic discs showed diffuse nerve fiber layer loss measuring 57 microns in the right eye and 70 microns in the left eye on the global indices.

Angiotensin-converting enzyme levels were markedly elevated (>200 ug/L). A cranial magnetic resonance imaging study showed enhancement of both optic nerves [Figure 1], both oculomotor nerves, the left V2 branch of the trigeminal nerve, and the right facial nerve as well as right cavernous sinus meningeal enhancement extending anteriorly and inferiorly into the pterygopalatine fossa, the orbital apex and the infratemporal fossa with minimal involvement of the occipital cortex. A full-body positron emission tomography scan and chest computed tomography scan revealed mediastinal lymphadenopathy as well as multifocal areas of hypermetabolic activity involving the head, neck, chest, abdomen, and pelvis with no evidence of occipital cortex involvement. Cerebrospinal fluid was significant for inflammatory pleocytosis (19 WBC's mm^3, elevated protein, normal glucose). Mediastinal lymph node biopsy showed non-caseasting granulomas consistent with the diagnosis of sarcoidosis. The patient was started on 50 mg of prednisone daily for 3 months that was subsequently tapered to a maintenance dose of 5 mg.

The patient's visual hallucinations had completely resolved by September 2010. At last follow-up in April 2011, best visual acuity had improved to 20/25 bilaterally and the central visual field defects improved bilaterally with some residual mild nerve fiber layer defects and optic atrophy in both eyes. There was complete resolution of the patient's peripheral nerve palsy.

DISCUSSION

Sarcoidosis is a multisystem inflammatory disease of unclear etiology characterized by the hallmark histologic finding of non-caseating epithelioid granulomas.[2] Neurosarcoidosis occurs in up to 2-5% of all sarcoidosis patients and can cause cranial neuropathy (particularly peripheral facial nerve palsy), neuroendocrine dysfunction, seizures, encephalopathy, and hydrocephalus.[2] Anterior uveitis and optic neuropathy are the most common ophthalmic manifestations of sarcoidosis, though the disease can affect any component of the eye. Visual hallucinations are rarely associated with neurosarcoidosis and a literature search indicated one report of a patient who developed hallucinations secondary to neurosarcoidoisis-induced meningoencephalitis with cognitive impairment, but had no intraocular complications except for mild papilledema.[3] Our patient exhibited no evidence of encephalitis, seizure activity or psychological manifestations of neurosarcoidosis and had complete resolution of her hallucinations following visual recovery.

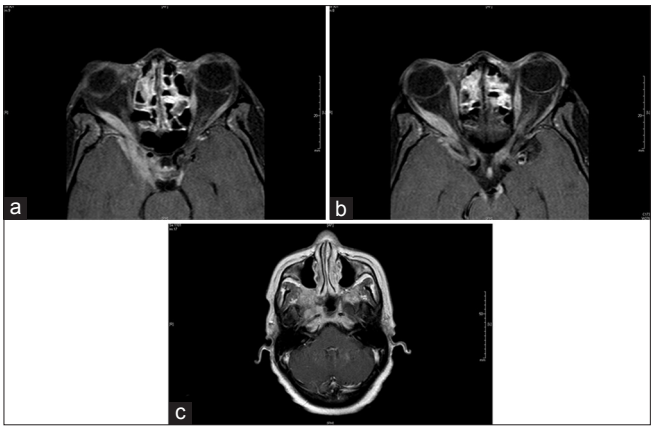

Figure 1: T-1 axial magnetic resonance imaging post gadolinium contrast at the level of the cavernous sinus (a) mid-midbrain (b) and foramen magnum (c) showed enhancement of the bilateral proximal optic nerves and the bilateral third nerves. There is also enlargement of the proximal optic nerves and the right lateral rectus muscle. Enhancement of the right cavernous sinus is seen extending anteriorly and inferiorly into the pterygopalatine fossa, the orbital apex and the infratemporal fossa

CBS is described as lucid visual hallucinations in patients with acquired vision loss and lack of cognitive impairment.[1] The visual loss is generally bilateral and patients with CBS often retain insight into the unreal nature of their hallucinations. These hallucinations often vary in type and normally do not involve other sensory modalities. The hallucinations often resolve with recovery of visual acuity or visual fields, such as after pituitary adenomectomy.[4] However, resolution of the hallucinations can occur spontaneously, even in the absence of visual improvement.

The pathogenesis of CBS remains unclear. One theory suggests that lesions of visual pathway result in the transmission of abnormal signals to the visual cortex.[5] These signals, when added to normal visual cortex activity are thought to account for the complex visual hallucinations experienced by these patients. Another theory suggests that visual deprivation leads to the production of spontaneous images from the visual cortex, creating visual hallucinations in a manner similar to the phantom limb pain phenomenon (release hallucinations).[5] Age related macular degeneration remains the most common etiology for CBS while other neuro-ophthalmic causes of visual loss have not been reported to cause CBS at the same frequency.[1] Whether or not this represents ascertainment/selection bias, failure of recognition or a true difference in incidence of CBS is unknown, but raises further questions about the pathogenesis of this disorder with regards to the importance of genetic, immunological, and environmental factors.

Treatment of CBS mainly involves management of the underlying ophthalmic pathologies. Patients should be reassured that the hallucinations are inherently benign and not the harbinger of psychiatric disease. Several case reports have suggested that treatment with various pharmacologic agents, including olanzapine, donepezil, and pegaptanib, can be beneficial in resolving hallucinatory symptoms.[1] Clinicians should be aware that CBS can be a symptom of visual loss of any etiology, including sarcoidosis.

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