

SLEEP MEDICINE PEARLS

## A Girl Who Sees Dead People

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### CASE REPORT

A 10-year-old African American girl with no significant past medical or surgical history and taking no regular medications presented with complaints of seeing her recently deceased uncle while resting on her living room sofa in the afternoon or in her bedroom at night. In these visual experiences, she saw her uncle walk through the house. They were very upsetting to the patient, causing her to cry and seek comfort from her mother. The mother was concerned about voodoo or satanic influences on her child. The patient denied hearing her uncle speak to her during these events or any history of auditory hallucinations. She denied illicit drug/alcohol or tobacco use; there was no suggestion of physical, emotional, or sexual abuse. The patient also had excessive daytime sleepiness (EDS), with sudden episodes of rapid sleep onset during the day while in school. These episodes were brought to the mother's attention by the patient's teachers. The patient denied episodes of loss of muscle strength in her legs, arms or face associated with either laughing or cry-

ing. Her physical examination, including neurological examination, was unremarkable. Laboratory tests including a basic metabolic panel, complete blood count, liver function tests, and thyroid function tests, were all unremarkable.

The patient underwent an extensive neuropsychological evaluation for cognitive functioning (Wechsler intelligence scale for children), academic achievement (Wechsler individual achievement test), attention/executive functioning (D-KEFS verbal fluency test), visual spatial functioning/visual monitoring (developmental test of visual-motor integration), memory functioning (NEPSY-II developmental and neuropsychological assessment), and behavioral/social emotional functioning (behavioral assessment for children, childhood depression inventory, house-tree-person projective drawing). Beyond a few episodes of sleepiness during the neuropsychological evaluation, the patient's results were all within normal limits. No evidence of thought disorder or disturbed mood was seen.

**Question: What is the diagnosis?**

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**Table 1**—Differential Diagnosis of Complex Visual Hallucinations in a Young Patient

Disease	Key diagnostic features
Insufficient sleep with hypnagogic/hypnopompic hallucinations	Sleep disordered breathing and/or nocturnal leg movements on PSG, or Narcolepsy on MSLT, or Insufficient sleep on sleep log Hallucinations upon falling asleep or awakening
Substance abuse	LSD: visual features only Amphetamines/cocaine: visual and auditory features
Schizophrenia/schizoaffective disorder/mood disorder with psychotic features, PTSD with “re-experiencing”	Visual and auditory hallucinations Stimulants may exacerbate psychosis
Hysteric conversion, malingering for stimulants	Diagnosis of exclusion
Complex partial seizures	Temporal lobe epilepsy can produce visual hallucinations, auditory hallucinations, persecutory delusions and ideas of reference—all features of schizophrenia. Temporal lobe epilepsy can be differentiated from schizophrenia by a lack of family history and preserved affect between ictal events. Complex visual hallucinations with previously experienced complex color images are usually produced from the occipitoparietal junction of the non-dominant parietal lobe. These hallucinations can also include micropsia (images appear smaller) and morphopsia (objects appear distorted).

CPAP, continuous positive airway pressure; PSG, polysomnogram; MSLT, multiple sleep latency test; SSRI, selective serotonin reuptake inhibitor; PTSD, posttraumatic stress disorder; LSD, lysergic acid diethylamide. (Modified from Manford, et al.<sup>1</sup>)

**Answer: Narcolepsy without cataplexy.**

## DISCUSSION

Complex visual hallucinations have a broad differential diagnosis spanning multiple neurologic and psychiatric conditions (Table 1).<sup>1</sup> The differential diagnosis of complex visual hallucinations in our patient includes insufficient sleep with hypnagogic/hypnopompic hallucinations, substance abuse, schizophrenia/schizoaffective disorder/mood disorder with psychotic features, hysteric conversion, posttraumatic stress disorder (PTSD) “re-experiencing,” malingering for stimulants, alcohol/barbiturates withdrawal, and complex partial seizures. The differential diagnosis of complex visual hallucinations in older adult patients may include peduncular hallucinosis associated with lesions in the midbrain, pons or thalamus; Parkinson disease<sup>2</sup>; dementia with Lewy bodies; and Charles Bonnet Syndrome. Charles Bonnet syndrome is characterized by complex visual hallucinations in the presence of normal cognition in elderly individuals. It almost always occurs with profound loss of vision or interruption of visual input into the occipital cortex anywhere along the visual pathway.<sup>3</sup> Affect and executive function are usually preserved in Charles Bonnet Syndrome. Awareness of the broadness of the differential allows for a more complete assessment. Each disease has a distinct therapeutic approach. For example, the hypnagogic/hypnopompic hallucinations of narcolepsy do not respond to treatment with antipsychotic medications. Treatment for one disorder may result in increased sedation and worsen others.

A particularly difficult diagnosis to make is simultaneous narcolepsy and schizophrenia. A clue to the presence of both

of these diseases in a single patient would be the coexistence of hypnagogic/hypnopompic complex visual hallucinations with auditory hallucinations. However, auditory hallucinations and out-of-body experiences/autoscopy can occur in narcolepsy. Treatment is also problematic, since the drug therapy for each disease worsens the other. Stimulants such as amphetamine or modafinil may exacerbate psychotic symptoms in patients with schizophrenia.<sup>4</sup> In addition, while treatment of narcolepsy with stimulants may reduce hypnagogic/hypnopompic hallucinations by reducing EDS, schizophrenic symptoms, such as auditory hallucinations, would persist.<sup>5</sup> Typical antipsychotics, such as chlorpromazine and haloperidol are sedating and promote weight gain, which can, in turn, lead to increased EDS and sleep disordered breathing. Of the atypical antipsychotics, apiprazole and ziprasidone are the least sedating; quetiapine, olanzapine, risperidone and clozapine are the most sedating—particularly quetiapine. Apiprazole and ziprasidone do not promote weight gain, while quetiapine, olanzapine, risperidone, and clozapine do.

Negative symptoms of schizophrenia are characterized by the absence or elimination of certain behaviors; and include *avolition* (loss of motivation, diminished drive, and apathy), *alogia* (talks very little and uses few words), *anhedonia* (reduced ability to experience pleasure in previously pleasurable hobbies or interests), and affective flattening (resulting in a loss of facial expression, eye contact, and expressive gestures or vocal inflections). The differentiation between schizophrenia and narcolepsy in a patient with prominent negative schizophrenic symptoms is challenging. Negative symptoms may easily be confused with the EDS and fatigue of narcolepsy. While EDS associated with narcolepsy responds well to amphetamines and modafinil, the negative symptoms of schizophrenia do not. Modafinil, at 200 mg/day has been shown to be ineffective in

treating the negative symptoms of schizophrenia.<sup>6</sup> Another tool for differentiation is that patients with schizophrenia tend to have cognition disorders. As a result, neuropsychological assessment by neuropsychologists, who are aware of the effect of EDS on the tests used, may be of particular importance in differentiation.

Adult norms for pathological levels of sleepiness are generally applied to adolescents, but no widely accepted norms for children currently exist. In adults, mean sleep latency on the multiple sleep latency test (MSLT)  $\leq 8$  minutes, with at least 2 sleep onset REM periods (SOREMPs), helps confirm a diagnosis of narcolepsy. However, in children from 6 to 12 years of age, mean sleep onset latency of  $<16$  minutes is considered “at risk” for having significant daytime sleepiness. Given the limited validation of the MSLT in 8-year-old patients, neuropsychological testing was performed to help substantiate the diagnosis. A diagnostic polysomnogram (PSG) was performed with a subsequent MSLT (4-nap protocol), with normal sleep before her PSG and MSLT. PSG total sleep time (TST) was 410 minutes with a TST apnea-hypopnea index of zero. Periodic limb movements of sleep were not present. MSLT revealed an average sleep latency of one minute and four SOREMPs. Normal neuropsychological testing results combined with these MSLT results helped us make the diagnosis of narcolepsy in this patient. Scheduled naps helped relieve the patient’s EDS and visual hallucinations.

#### CLINICAL PEARLS

1. Complex visual hallucinations have a broad differential diagnosis; each disease has a distinct therapeutic approach (see Table 1).
2. Hypnagogic/hypnopompic hallucinations can be associated with insufficient sleep from a variety of causes, including sleep disordered breathing, narcolepsy, sleep related movement disorders, and behaviorally induced insufficient sleep syndrome.
3. Narcolepsy and schizophrenia may coexist in the same patient, making treatment difficult. Amphetamines and modafinil may induce psychosis, and many typical and atypical antipsychotics lead to sedation/weight gain.

#### DISCLOSURE STATEMENT

This was not an industry supported study. The authors have indicated no financial conflicts of interest.

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