Phenomenological and Diagnostic Implications of Paraschemazia: A Case Report

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SUMMARY
Paraschemazia is characterized by distortions of body image, and has been observed in neurological disorders (epilepsies, migraine, and non-dominant cortical lesions), retinal damage, and hallucinogenic drug use. This complex symptom cluster of subjective and objective perceptual abnormalities forms an essential feature of Todd’s Alice in Wonderland syndrome. This neuropsychiatric manifestation must be phenomenologically differentiated from hallucinations, illusions, and agnosias. Herein we report a patient with paraschemazia that occurred during the ictal phase of temporal lobe epilepsy; the patient was referred to the department of psychiatry due to symptoms of transient intense fear and abnormal perceptions. We also discuss the diagnostic difficulty and phenomenological implications associated with this rare phenomenon.

A 45-year-old female presented with brief episodes (30-45 s) of sensory distortion, during which the faces and limbs of other people and her own appeared distorted (e.g. protruding eyes, elongated nose, and oval or square head). The episodes of sensory distortion were associated with intense fear and were followed by loss of consciousness for 5 min. She had been experiencing these episodes since the age of 12 years and responded poorly to several antiepileptic medications. 3-Tesla MRI performed using a complex partial seizure protocol showed altered signal intensity and volume loss in the right hippocampus, suggestive of right mesial temporal sclerosis. Neuropsychological assessment showed impaired sustained attention, working memory, and verbal and visual learning and memory, suggestive of bilateral dorsolateral prefrontal and temporal lobe deficits. She improved in response to a combination of levetiracetam and clobazam, and was seizure free at the 2-year follow-up.

Key words: Visual perception; psychopathology; epilepsy, temporal lobe; neuropsychiatry; Alice in Wonderland syndrome

INTRODUCTION
Sensory distortions (Sims 2002) are perceptual abnormalities of intensity, quality, spatial form, and affect. Paraschemazia is characterized by rare sensory distortions, described as fluctuations and distortions in the size and shape of body parts. Paraschemazia occurs in association with the use of hallucinogenic drugs, epilepsy, migraines, and non-dominant cortical infarcts (Casey and Kelly 2007), and are also an essential feature of Todd’s Alice in Wonderland syndrome (Podoll et al. 2002; Todd 1955). Clinically, the distortions must be phenomenologically differentiated from illusions, hallucinations, and visual agnosia for accurate diagnosis. Herein we report a patient with paraschemazia associated with temporal lobe epilepsy that was mistakenly referred to the department of psychiatry for visual hallucinations, as well as a discussion of the diagnostic and phenomenological implications of this rare presentation.

CASE REPORT
Mrs. X, a 45-year-old, right-handed female from northern India, was referred to the department of psychiatry from the neurology department to be evaluated for possible panic attacks and visual hallucinations; she had symptoms of brief intense fear and anxiety, with autonomic arousal and perceptual abnormalities. Upon evaluation she recounted a history
of episodes of abnormal behavior (described below) followed by loss of consciousness that began at age 12 years, and poor treatment response to antipsychotics, anxiolytics, and multiple anti-epileptic drugs. Each episode had a 2-d prodrome characterized by heightened threat perception, referential ideas and elementary auditory hallucinations, i.e. door knocking, footsteps approaching, and clock ticking, followed by sudden onset of abdominal discomfort with sensations of gas rising through her thorax, dry mouth, thirst, palpitations, tremors, sweating, and smelling incense and roses (in instances in which nobody else was able to smell them) (olfactory hallucinations). These symptoms usually lasted about 2 min and were followed by a period of 30-45 s during which any person near her appeared to have distorted faces (protruding eyes, elongated and deformed nose, and oval or square head) and distorted limbs (irregular shapes and varying sizes). She also perceived her own limbs as distorted and deformed. Together, these phenomena were suggestive of paraschemazia. She reported being extremely frightened by and trying to get away from anyone that approached her, screaming and crying in fear, and pleading with family members to go away, whereas inanimate objects appeared to her normal in size and shape. This was followed by loss of consciousness for about 5 min, headache for about 15-30 min, and then complete recovery. She was able to recall events until the paraschemazia episode, but had amnesia of all subsequent events. She had a family history of epilepsy (2 of her siblings) and she did not abuse any medications or recreational drugs.

The duration of each episode (pre-ictal, ictal, and post-ictal phases together) was approximately 3 d. She remained asymptomatic the remaining days, except for apprehension about the recurrence of episodes. A review of her treatment history showed that she had always exhibited an initial response to anti-epileptic drugs, only to relapse while continuing treatment. Adequate trials of carbamazepine, sodium valproate, and phenobarbitone were unsuccessful, and she was taking levetiracetam 2 g d⁻¹ when she presented to the referring neurologist. She was evaluated via cranial CT and MRI (1.5 Tesla), and measurement of metabolic, endocrinological and hematological parameters; all findings were normal. We repeated cranial MRI using a 3-Tesla machine and a complex partial seizure protocol used at our institution), which showed altered signal intensity in the right hippocampus with volume loss, altered internal architecture, and loss of pes cavus appearance of the head. T2 relaxometry values of the head of the hippocampus were 90 ms on the right and 70-80 ms on the left. These features were suggestive of right mesial temporal sclerosis (Figure a and b). Electroencephalography performed during an inter-ictal period, however, showed no epileptiform discharges.

Neuropsychological assessments were conducted using the National Institute of Mental Health and Neurosciences Neuropsychological Battery (Rao et al. 2004). Motor speed (finger tapping test), verbal comprehension (token test), category fluency (animal naming test), and visuospatial construction (copying scores from the complex figure test) performance were adequate; however, she exhibited impaired (score 2 standard deviations below her age, gender, and level of education standardized norms) sustained attention (digit vigilance test), working memory (verbal 2-back), verbal (Rey’s Auditory Verbal Learning Test) and visual (recall scores from the complex figure test) learning and memory, and paired association (word association test). This profile suggested bilateral dorsolateral prefrontal involvement and temporal lobe deficits. A diagnosis of complex partial seizures was considered, and clobazam 20 mg d⁻¹ was added as an adjuvant to levetiracetam 2 g d⁻¹, and subsequently the frequency of seizures was reduced. At the 2-year follow-up for treatment with the anti-epileptic combination of clobazam and levetiracetam she was seizure free.
DISCUSSION

The presented patient experienced sensory distortions of body image, illustrating the phenomenological dilemma that led to her referral to the psychiatry department with visual hallucinations. The patient’s ictal experiences with body images were sensory distortions, as there were visual stimuli (presence of people) that were subjectively perceived in a distorted manner. As such, these experiences were not hallucinations because sensory stimuli were always present. Illusions were also unlikely, as the objects of perception (faces and body parts) retained their identity and were not misconstrued for other objects. The patient was able to recognize the objects of perception correctly, thus ruling out the possibility of visual agnosia. The Table illustrates the conceptualization of various visual perceptual abnormalities that were considered as differential phenomenological entities. Paraschemazia is one such example of sensory distortion in which body parts appear distorted; however, patients are able to recognize them as body parts and are able to appreciate the distorted quality of their perception.

The presented patient had a negative history of hallucinogen/other substance abuse that could explain her symptoms. Retinal damage was ruled out based on normal ophthalmological examination (including fundoscopy) findings. Sensory distortions of size (e.g. micropsia and macropsia) have been described in patients with depersonalization syndrome. Brief intense fear associated with symptoms or autonomic arousal (e.g. sweating, palpitations, and tremors), as noted in the presented patient, can be mistaken for panic disorder; however, these diagnostic possibilities were unlikely in the presented patient because her symptoms were followed by brief periods of unconsciousness, suggesting a neurological etiology. Moreover, the periodic, consistent, and stereotyped nature of her episodes, typical semiology, MRI findings of mesial temporal sclerosis, and good response to anti-epileptic drugs together suggested a possible clinical diagnosis of complex partial seizure.

Paraschemazia in the presented patient typically involved the face and extremities. Functional MRI studies indicate that regions in the right temporal cortex respond preferentially to faces and extremities (Pinsk et al. 2005; Kanwisher et al. 1997). The extrastriate body area (EBA)—a part of the lateral occipitotemporal cortex, predominantly in the right hemisphere—is specifically involved in processing bodies (Downing et al. 2001). On the other hand, the fusiform face area (FFA)—again, a part of the occipitotemporal cortex—is specifically responsive to facial stimuli and insensitive to non-facial body parts (Grill-Spector et al. 2004). Based on neuro-imaging and neuro-psychological findings in the presented patient, we surmised that the patient’s episodes of paraschemazia were due to epileptiform discharges from the right hippocampus, which is closely connected to the FFA and the EBA via the hippocampo-fusiform pathway (Smith et al. 2009).

In conclusion, the presence of paraschemazia should warrant a thorough neurological work up because of its association with complex partial seizures and migraine. Attention toward subtle phenomenological differences may potentially help delineate paraschemazia from other more common presenting visual hallucinations, illusions, and agnosia, thus preventing misdiagnosis.

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REFERENCES


