

# Psychosis and Capgras delusions in a patient diagnosed with Cogan's syndrome

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## Clinical presentation

Vincent is a 39-year-old Hispanic American gentleman who was admitted to our acute inpatient service on a legal hold as a danger to self, danger to others as well as being gravely disabled. He presented with a chief complaint: "I need to sleep." A 5150-legal hold was initiated by the Police Department. The family had called law enforcement because the patient was trying to attack family members with a kitchen knife. When the police arrived, he was sitting on a couch with a large kitchen knife behind his back. The patient struggled with the officers when they attempted to retrieve the weapon. Remarkably, Vincent has total visual loss and near total hearing loss. The patient was unable to provide much of the history, which was obtained mostly from collateral sources, his family and previous medical records. The patient's brother reports that Vincent had become convinced that family members had been substituted by "intruders." Vincent was not convinced that neither his mother, nor his brother were "real" and had been substituted by imposters. We observed this quite dramatically during a family meeting. His brother leaned over to Vincent and spoke into his right ear (in which Vincent has some residual hearing), announcing his name to get Vincent's attention. Vincent began violently waving his arms protesting "no, no," indicating that he did not accept his brother's identification. Vincent also believed that his brother's girlfriend was in fact his own wife and was frequently angry because she did not join him in his bed at night. In addition, he believed that his nephew was in fact his own child.

Vincent had one previous admission to our facility approximately 10 months prior to this admission. He was admitted on a legal hold as being a danger to others. He had become aggressive with family members, stating that he worked for the CIA and that he was a friend of the governor's. Vincent stated during that admission, that his mother, who he lives with, had been replaced by an imposter. He was quite agitated during the admission procedure and briefly required to be placed in restraints. Characteristic of Capgras delusions, Vincent did not appear to be concerned about the safety and whereabouts of his loved ones who had been replaced by imposters and described his mood as "calm".

Vincent did not have a prior history of mental health problems. His difficulties began in August of 2010 when he experienced pain in his eyes, blurred vision and photophobia. He was initially diagnosed with conjunctivitis. He suffered from watery and painful red eyes for about three weeks. He was prescribed gentamicin sulfate ophthalmic drops (0.3%). He denied any history of eye trauma and his physical examination was unremarkable. His body mass index, however, was increased at 33.6. He was observed to be alert and oriented to time, place and person. It was noted that the onset of his illness was acute

early in August of 2010. He experienced a headache and complained of loss of vision. He was seen in the emergency department where he was found that he could differentiate light and dark, but just minimally. During the emergency room visit, it was noted that his hearing was "okay." Sudden hearing loss ensued, and by September 2010 he had marked auditory deficits. His physical examination revealed very small pupils, markedly injected sclera and normal intraocular pressure. A CT scan of the head without contrast was unremarkable. It was not until September 2010, when a rheumatologist diagnosed him with Cogan's syndrome and initiated steroid therapy with prednisone 60mg orally daily. The rheumatologist comments in his report that "Cogan's syndrome is an inflammatory disorder affecting young adults, characterized by ocular and vestibular auditory dysfunction. Ocular disease includes red and painful eyes, photophobia, blurred vision retinal vasculitis and posterior scleritis." The patient had dizziness and nausea, suggesting vestibular manifestations, which are common in Cogan's syndrome. There was no evidence for generalized systemic vasculitis in this patient. Vasculitis in Cogan's syndrome usually affects the large vessels, for example aorta. The patient had good pulses in his arms and had no leg complaints. His cardiac exam was unremarkable. For insurance reasons, no extensive workup including echocardiogram and other tests was feasible. In January of 2011, a specialty referral to ophthalmology was made which further confirmed the diagnosis of Cogan's syndrome based on the typical findings of keratitis and posterior synechiae.

## MRI and CT scan of the brain

There was a vague increased T2 signal within the medial aspect of the right temporal lobe, suspect more artifactual than actual. Nasal septal deviation to the left appreciated. Subtle scalp soft tissue contour abnormality over the left frontal convexity noted, which could reflect the sequela of prior trauma. Left V4 segment of the left vertebral artery is dominant over the right side, felt congenital.

**Impression:** No convincing definite acute focal intracranial pathology.

CT scan was also unremarkable.

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## Diagnostic considerations for our patient Vincent

### Cogan's syndrome

The patient's family reports to us that Vincent did not have any psychiatric symptoms prior to the onset of his visual and auditory deficits. Vincent suffered a very rapid loss of vision, followed soon by deficits in auditory and vestibular function. The association of interstitial keratitis and audio- vestibular auditory symptoms has been described originally by David Cogan in 1945 [1]. In the subsequent 68 years many more cases have been described and a variety of symptoms have been detailed. A recent review from the Mayo Clinic describes experiences of 60 patients collected over the last 50 years [2]. The average age of onset of disease was thirty eight. Imaging studies of the brain by either CT scan or MRI were uniformly negative. Corticosteroid therapy, although moderately effective in some patients if treated early in the disease, was rarely effective when instituted later than four weeks after the day of onset. The Mayo clinic cohort had a variety of vestibular complications such as ataxia and oscillopsia. There are no reports in this cohort of any psychiatric symptoms or diagnoses.

Cogan's syndrome can be associated with vasculitis and lacunar brain infarcts [3]. Both CT scan and MRI, however, were read as being within normal limits in our patient.

Vincent showed the hallmarks of the "typical" Cogan's Syndrome [4], *i.e.* ocular involvement with interstitial keratitis, and audio-vestibular involvement similar to that of Meniere's accompanied by rapidly progressive and profound hearing loss. He had ocular pain, redness, photophobia and loss of visual acuity. He experienced dizziness, nausea and rapid hearing loss.

### Capgras delusion

Vincent's psychiatric symptoms included the rapid onset of grandiose and persecutory delusions as well as a "delusional misidentification" syndrome (DMS) originally described by Capgras and Reboul-Lachaux [5]. Patients suffering from DMS consistently misidentify persons, places, objects, or events [6]. Capgras-type delusions are not uncommon in a variety of psychiatric [6-8] and neurological disorders [9,10]. There is a significant risk of violence in patients with Capgras delusions [10-14]. Our patient Vincent had become threatening to family members whom he misidentified as imposters and had armed himself with a kitchen knife, which he held behind his back. His blindness may have mitigated the risk of harming his mother and brother.

A number of neurological correlates have been associated with the development of misidentification delusions [6] such as direct damage to right temporal lobe regions involved in the visual processing of faces by ischemic stroke [15] or damage to connections of this area to the amygdala, which is important in the emotional processing of facial images [16]. A number of disconnection syndromes [17] have been described resulting in impairment of facial recognition and the neuronal pathways involved have been reviewed [18]. Anatomical disconnection [16] may cause an inability to match current experience with autobiographical memories resulting in the under-identification of people and places [6].

There are few reports describing Capgras syndrome developing after sensory loss.

A 75 year old woman rapidly developed complete blindness as a result of giant cell arteriitis and treatment complication. She believed

that her spouse of 25 years had been replaced by another man. No evidence of focal brain lesions was found by CT scan [19].

A 66 year old woman had lost her hearing after ear infections and surgical treatment of her ears and started to believe that her daughter had been replaced and was being held captive in a secret chamber. She intermittently heard the voice of her daughter on her right side only [19]. A 73 year old man with childhood onset retinitis pigmentosa was totally blind by age 65. He developed Parkinson's disease at age 67 which improved with levodopa therapy. He developed visual hallucinations at age seventy-one. He believed that a number of imposters, including men replaced his wife [20]. A 26-year-old Afro-Brazilian married housewife became blind at age 16 years, after a bilateral Tuberculous uveitis. She was admitted to hospital in a severe depressive episode, with marked apathy, weight loss, sadness, and suspiciousness, especially of her husband. She believed that her spouse had been replaced by an impostor, an almost-identical person who was trying to take his place [21]. A 32 year old male suffering from total blindness as a result of diabetic retinopathy and recovering from hypoglycemic coma, metabolic encephalopathy and haptic and visual hallucinations, believed that his mother was a double because the skin of her hand felt softer [22].

### Discussion

The syndrome specific and neurological manifestations of Cogan's syndrome have recently been reviewed [2,23,24]. Cogan's syndrome is rare. By 2004, fewer than 250 cases have been reported in the literature [24]. Surprisingly, these reviews and a search of the psychiatric literature showed little evidence for psychiatric complications of this devastating disorder that leads to sudden severe visual impairment and profound deafness within a few months. To the best of our best knowledge, this is the first case of Capgras misidentification delusions in a patient with Cogan's syndrome.

Capgras syndrome is one of the fascinating types of a family of delusional misidentification syndromes (DMS). When Capgras and his colleague Reboul-Lachaux, first reported it in 1923, they suggested the term "illusion des sosies," which was essentially an identification agnosia [5]. The patient described in their report had no trouble recognizing faces, but failed to identify these faces as the faces of her dear ones.

Capgras syndrome is associated with both psychiatric disorders and various neurological conditions. Psychiatric conditions such as paranoia, derealization, and depersonalization and neurological illnesses such as seizures, cerebral infarction, traumatic brain injury, Parkinsonism and dementia have been associated with Capgras delusion [6]. A retrospective study comprising 47 subjects with Capgras syndrome, examined possible relationship of Capgras syndrome with neurodegenerative diseases [9]. About 80% of patients with a Capgras syndrome had a neurodegenerative disease, most commonly Lewy Body disease.

Capgras delusions are often linked to lesions in the parieto-temporal regions of the right hemisphere [25]. Feinberg and colleagues conducted a case series analysis of previously reported DMS cases and included a total of 29 applicable reports of DMS involving 27 different individuals [6]. Their analysis suggested that DMS is strongly associated with right hemisphere damage. Roughly 35% of cases with DMS had injuries limited to frontal lobe and this association was shown to be statistically significant. Interestingly, no single case with parietal or temporal damage alone produced DMS. Furthermore,

almost all patients who had *neuropsychological* testing evidenced a pattern of memory (primarily nonverbal), perceptual, and executive impairments, in lieu of what is to be expected from predominantly right hemisphere pathology [6]. However, it is to be noted that not all patients with right hemisphere pathology develop Capgras syndrome.

Various hypotheses have been suggested to explain the phenomenology of Capgras syndrome since its first report nine decades ago. Draaisma, a Dutch psychologist in his paper suggests that soon after the initial publication, Capgras published a series of case reports championing psychoanalytical explanations [26]. In essence, the Freudian psychodynamic model proposes that Capgras delusion evolves as a means of conflict resolution. The conflict itself is due to the unsettling sexual attraction towards the mother and the consequent jealousy and envy towards the father for sexual attention. Although plausible, this theory does not explain all cases of Capgras syndrome. Moreover, reports of Capgras delusion towards a pet dog seriously undermine the credibility of a Freudian explanation [27].

As Draaisma points out, the explanatory theories of Capgras syndrome underwent a “gestalt switch” in 1980s (26). Clinicians and researchers alike increasingly came to an understanding that an organic lesion in specific areas of the brain is the primary cause for the delusions [28]. The initial attempts at biological explanations came from studying another fascinating and perhaps related clinical phenomenon. Individuals with prosopagnosia are unable to recognize familiar faces, but have no difficulty in recognizing other visual objects. Bauer [29,30] initially suggested the existence of two pathways for face recognition; overt face recognition and a covert emotional recognition. In this study, he demonstrated that although conscious face recognition was impaired in patients with prosopagnosia, they were able to show autonomic arousal to familiar faces. Ellis and Young [31] proposed the idea that patients with Capgras delusion may have a “mirror image” of prosopagnosia. They hypothesized that individuals with Capgras delusion are consciously (overtly) able to recognize faces, but lack automatic (covert) emotional arousal to familiar faces. They suggested a visual-anatomic disconnection, which could account for cases with prosopagnosia and Capgras delusion. The dorsal route runs between the visual cortex and the limbic system through the inferior parietal lobe, carries the affect-laden signals and if damaged, could result in intact face recognition without feeling of familiarity, thus resulting in Capgras delusion. A few years later, they published their study on five patients with Capgras delusion, which supported the above hypothesis [32]. Using skin conductance responses they showed the lack of autonomic emotional arousal to familiar faces in their subjects. A similar theory proposed by Hirstein and Ramachandran [16,27] suggested that disconnection between the inferotemporal cortex and the amygdala could result in intact face recognition without evocation of appropriate emotion attached to the face, thus resulting in delusional misidentification. Furthermore, Ramachandran suggests a relationship between the Capgras syndrome and a more general difficulty in linking successive episodic memories because of the crucial role emotion plays in creating memories.

However, the pattern of decreased automatic emotional arousal responses described by Ramachandran has been reported in patients with no evidence of Capgras delusion [33]. Therefore, a lack of autonomic responses is in itself insufficient to produce the delusion [19]. Moreover, various case reports describing DMS as a result of sensory loss published since the 1990s appear to challenge this theory [19-22].

There are reports in the literature of Capgras syndrome as a result of drug abuse. A retrospective study identified two subjects who developed Capgras syndrome immediately after methamphetamine abuse [9]. Since it is well established that methamphetamine causes the loss of dopamine transporters [34,35], this was a novel finding suggesting a possible dopamine dysregulation in the pathogenesis. Others have also independently suggested dopamine dysfunction in Capgras syndrome [36]. There is also a published report of a case of transient Capgras delusion induced by ketamine in a healthy subject [37].

Collectively, these case reports challenge the visual- anatomic disconnection hypothesis. Our own case report is another example of Capgras syndrome occurring in the absence of demonstrable anatomical disconnection of visual pathway and limbic structures mediating affective responses to visual representations.

It is well known that individuals experiencing sensory deprivation often report perceptual disturbances such as hallucinations. The earliest work on this was done by Ziskind who explored the possibility of extreme prolonged sensory deprivation leading to psychotic experiences [38]. There is some debate whether personal predispositions play a role in evolution of frank psychotic states in response to sensory deprivation. McCreery and Claridge demonstrated that highly schizotypal individuals reported anomalous perceptual experience during a period of sensory deprivation [39]. A recent study (2009) explored the psychotomimetic effects of short-term sensory deprivation using a specially constructed anechoic chamber, which produced near total auditory and visual sensory deprivation [40].

The case reported here presents the impact of progressive sensory deprivation occurring over a time period of a few months leading to increasing distortions and culminating in an almost complete cessation of sensory (auditory and visual) input. This scenario may be understood in the emerging Bayesian model of psychosis [41], which postulates that a mismatch between “prior expectations” encoded in brain circuits and noisy or meaningless sensory input may lead to a failure of the brain to minimize prediction-errors and draw correct inferences from environmental stimuli.

Sensory deprivation [19-21], neuronal disconnection syndromes [16,27] and neuronal changes as a result of ketamine [37] and amphetamines [9] are seen as antecedents to the emergence of Capgras delusions by mechanism with converge on the disruption on the Bayesian mechanism invoked for brain/environment interaction [41].

Although we have been able to advance our understanding of Capgras syndrome substantially since the publication of the seminal article almost nine decades ago, we are yet to fully comprehend the phenomenology involved. Recognizing this very complexity, Stone and Young [25] proposed that patients with Capgras delusions suffer an impairment that leads to faces being perceived as drained of their normal affective significance, and an additional reasoning bias that leads them to put greater weight on forming beliefs that are observationally adequate rather than beliefs that are a conservative extension of their existing stock.

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