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Misclassification and emotional logic – a case report and a narrative review of Capgras syndrome

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Abstract

Quick, standardised, and often fragmented and adapted medical histories, have become the norm in the health service, including neurology, with procedures, forms, scales, etc. - modern medicine lacks elucidation, thus depicting soullessness and a lack of identity. Psychological theories are difficult to apply universally, as they are often affected by culture and contemporariness; and the mechanisms of the brain are unknown. Although delusions are common, the discipline of psychiatry has perhaps not shown sufficient interest in psychopathology which affects the imagination system. Capgras syndrome, a delusional misclassification syndrome, in a patient with Parkinson's disease, illustrates the blurred boundaries between psychiatry and neurology.

Introduction

Neurology (from Greek *neuron, nerve and logos,* doctrine) is the medical discipline concerned with diseases of the nervous system, whilst psychology (from Greek *psyche,* soul and *logos*) and psychiatry (from Greek *psyche, iatreia,* medical science) concern the mind and related diseases. Neuropsychiatry is at the intersection of these specialities, but disciplinary divides do not separate the understanding of the brain and human beings in the same way that the terms 'psychogenic', 'functional' and 'organic' do not individually provide a satisfactory explanation of complex disorders. The well-known neurologist, Oliver Sacks (1933-2015) and author of 'The Man who Mistook his Wife for a Hat', called for a new professional discipline: *neurology of identity* [1]. Ultimately, what we imagine and our scientific explanations might just boil down to logic. The ability of humans to imagine makes sensory experiences meaningful, it houses feelings, earlier experiences and interpretations, and makes sensing more than just physical stimulation of the sense organs [2]. Images are formed in the same way in the sensory system, but through the perspective of the individual. The reality, that we perceive to be authentic and true, is the image that arises when our expectations correspond with what we see, hear, smell and sense in general. The truth probably also lies within the logic of emotions, which the following case will illustrate.

Case Description with Discussion and Review of the Literature

An ordinary patient

Mr I was 63-years-old when I diagnosed him with Parkinson's disease; a progressive condition caused by the deterioration of dopamine producing brain nerve cells (basal ganglia) with subsequent weakening of the affected cell networks. The reason for this is unknown. He showed typical symptoms of the disease with reduced fine motor skills and tremors in one hand while resting. The following year he had the classic somewhat expressionless face, low voice, slightly forward bent posture, and in addition to hand tremors, his movements were slower. The patient was given *Levodopa*, which is an amino acid that easily penetrates the brain, converts to dopamine and replaces some of that lost. The treatment, which has been standard since the 60s, had a good symptomatic effect. He was furthered monitored by colleagues and the treatment was uncomplicated. Eight years was to pass before I saw him again, at which time he was admitted to my department for acute deterioration. It is not uncommon for patients who have suffered from Parkinson's disease for many years to develop psychiatric symptoms, but this patient was so bizarrely delusional that it awoke my interest.

A strange symptom

He gave me a warm greeting when I met him again and he said my name before I introduced myself. Nonetheless, he added that he could not remember what I looked like. His wife said that her husband had encountered orientation problems for the last six months, but only in his own apartment. He had a tendency to place things where they were not normally placed and when she called him from another room he found it very difficult to ascertain where her voice was coming from. Despite this, he had no difficulty in going shopping, paying bills online, travelling to his country home for a week on his own and he was an active follower of current affairs. Regrettably, however, the patient had been admitted earlier that day, because he did not recognise his wife.

The married couple had talked and done normal everyday things as usual, but early that afternoon, whilst sitting drinking

coffee, Mr I asked if it was alright if he blew in a bottle (an exercise he normally did for his disease). His wife said he could, of course, but added that it was not necessary to do it in public or when guests were present. To this, Mr I responded, *"My wife usually says that too."* When she tried to convince him that she was his wife, he asked her to telephone from an adjacent room. He did not perceive her as a stranger during the conversation, but as soon as she returned he became wary again.

Mr I gave a detailed account of his day, and he seemed intellectually and mentally normal. He also confirmed his wife's story. During the consultation, he said that the person accompanying him was extremely like his wife and acted like her, but expressed uncertainty by saying, *"Even though she looks like her, it doesn't feel like it's her."*

It did not appear that the problem arose from a lack of recognition, but a lack of identification. Nonetheless, could it be the case that his vision was impaired? He said that he found it harder to read and his wife had noticed that he sometimes put his coffee cup at the side of the saucer. An eye doctor's examination revealed reduced visual acuity in the left eye (1/3) due to age-related macular degeneration. On the MRI of the brain, no specific damage was seen, but a radiologist noticed a slight cerebral atrophy in relation to his age. Electroencephalography (EEG) recordings did not show any specific pathology either, but in general the activity of the brain waves was slightly slower than during an entirely normal examination. The patient was treated for an uncomplicated urinary tract infection and given a secondary diagnosis of a Capgras-like syndrome caused by delirium, and he was sent home.

Capgras

Capgras syndrome, named after Jean Marie Joseph Capgras (1873-1950), who in 1923 described a patient with this type of monothematic delusion, is the illusion that one or more of the people in one's close environment (parents, spouse, children) have been replaced by an imposter. In the original description, Capgras suggested that the imposter is a creation of *logique des emotions,* the logic of emotions [3]. Nonetheless, explanations deeply rooted in psychoanalysis were characteristic in later explanatory models, including his own until well into the 70s [4,5]. Other forms of delusional misclassification syndrome, or *syndrome of subjective doubles* as it also has been called, also exist, but they are much rarer. These could include confusion about pets, personal positions and buildings, for example, one's home [6].

Phenomenon or syndrome?

A year later the patient came for a check-up. He could move reasonably well, but it was clearly seen that he had Parkinson's disease. He was in a good mood and aware of time and space, but more reticent than earlier. This time he confirmed that his vision was disturbed. He had visual hallucinations quite often, especially at his country home. He saw 'dogs running' or 'armies marching', but added, *"They disappear when I approach them."* He continued to perceive his wife as a stranger, but someone who looked very similar, *"A third person,"* he stressed. *"Especially when I raise my voice,"* interrupted his wife. She said that on one occasion her husband would not go to bed in the evening, because he thought someone else was in the bed. Other times, he would suddenly walk a few metres away from her when they were walking and then call for her. The problem with orientation around their home had escalated, especially in the evenings, and arrows and instructions had gradually been put up in the apartment. Despite these problems, he continued to perform everyday chores and did not appear to be suffering from dementia. Changing the dose of Levodopa did not help and his GP referred him for a psychiatric assessment, which he underwent six months later. The report was half-a-page long: *The patient speaks of troublesome visual hallucinations. He says he sleeps quite well with the occasional nightmare. The most distressing aspect for the patient and his wife is that he does not always recognise his wife. This phenomenon is only connected to his wife - not his children or grandchildren. Before initiating any other measures, I am referring the patient to the neurological outpatient clinic for an assessment on the patient's medication for his basic disorder.*

From psychiatry to neurology?

In all honesty, I must admit that I was disappointed with the psychiatrist's short and 'organic' assessment, but in general he was correct. Visual hallucinations are common with Parkinson's disease and a relatively frequent side effect of Levedopa. Nonetheless, another explanation could also explain the visual hallucinations. Some people with impaired vision due to an eye disease, especially the elderly with age-related retinal degenerative disease, can experience repeated lively visual hallucinations despite having normal cognition and insight. These tend to appear with poor lighting in a blind spot in the visual field. The hallucinations are usually complex and varied, and generally involve animals and people. It is assumed that the phenomenon is caused by compensatory activity in the brain (the anterior occipito-temporal cortex) as a result of lost information from the primary visual pathways known as a release phenomenon. The syndrome was first described in 1760 by Swiss philosopher, Charles Bonnete (1720-1793), and as such was named after him [7].

An MRI did not detect any damage to Mr I's brain, yet he had agnosia (from Greek *a,* without and *gnosis,* knowledge), which is the inability to interpret some sensory impressions despite having assumingly adequate primary sensory functioning. He struggled with recognising/identifying his wife (visual agnosia) and orientation in familial surroundings (topographical agnosia). Nonetheless, could the pathology be connected to a damaged visual system?

Seeing and feeling

It is not difficult to imagine that ambivalent (dualistic) feelings might form the basis for Capgras syndrome. An

earlier prevailing theory was that the patient was to create two split objects to allow the imposter to project unpleasant feelings without affecting the original. In Freudian theory, this is a defence mechanism to camouflage sexual orientation, incestuous feelings or an expression of regression to the infantile [4,5]. It can also be considered an unresolved conflict; a division of the conscious and unconscious part of the superego (the self). The imposter in this medical case did not perceive that she was the object of aggression, but she noticed that her husband's delusion tended to appear after she raised her voice. In literature, patients with Capgras syndrome react in different ways to their imposters, i.e. from hate to friendship.

In the middle of the 80s after being inspired by studies on so-called split-brain patients, an American neurologist launched an organic explanatory model. By then, it had also become known through a number of published case reports, that Capgras syndrome did not only appear in connection with traditional psychiatric diseases, such as paranoid schizophrenia, but also with many somatic brain diseases and conditions. Today, the list is long, and ranges from migraines and medicines to structural brain damage. It seems that delusional misidentification syndromes appear relatively often in connection with neurogenerative diseases, especially Alzheimer's disease and Lewy body dementia [8]. A few cases in Parkinson's disease have also been described [9]. In other words, Capgras syndrome may appear as a result of non-interaction between the two hemispheres of the brain (*cerebral hemisphere disconnection hypotheses*).[10] Two images of a face are created in the brain, one in each brain hemisphere. The face that is normally seen, interpreted, and potentially recognised and identified, is a fusion of these two images. Any damage prevents patients with Capgras syndrome from integrating successive memories with real time sensing. The face is recognised, but an unconscious inner conflict results in the conclusion that it must belong to someone else – an imposter. In 1990, Ellis and Young initially used knowledge about the brain, and how it conveys and processes visual information about objects, including faces, to postulate a model based on *logique des emotions* – a theory which to some extent could also be tested [10,11].

Prosopagnosia

Raw data from the retina is conveyed via the optic nerve to the brain's primary visual cortex in the occipital lobe. From here, the signals are distributed to a number of other areas, the extrastriate visual areas, where they are processed and integrated before obtaining a conscious visual experience. The signals follow two main pathways, of which, one leads to the parietal lobe where information about the whereabouts of an object is stored, i.e. an area for spatial analysis. In simple terms, this pathway provides us with unconscious knowledge about the object and the possibility to register it. The other pathway leads to an area in the occipitotemporal gyrus (gyrus fusiformis) for recognition and classification of the object. It is then analysed more thoroughly and integrated with previously acquired information (memories, knowledge, semantics) in the two other temporal lobe areas. This pathway enables us to recognise what we see. It has been found that gyrus fusiformis is especially important to the recognition of facial features. Any damage to this area (mostly when in both brain hemispheres) could result in an isolated inability to consciously recognise a face and to whom it belongs. This is known as prosopagnosia (from Greek *prosopon,* face and *a gnosis,* no knowledge). Hence, prosopagnosia is a specific form of visual agnosia, i.e. facial agnosia. In its most pronounced form, the patient must use non-visual information (for example, voice) to identify a person. In milder cases, normally after one-side damage to the right brain hemisphere, the patient is able to determine age, gender and facial expressions.

A third pathway

People are not robots. Visual impressions awaken emotions and bodily reactions. A central pathway probably exists from the gyrus fusiformis (found on the inside of the temporal lobe), which runs via the upper part of the temporal lobe (superior temporal sulcus) to the almond-shaped nucleus (amygdala). This part of the brain is most strongly connected to emotions and emotionally charged behaviour. Knowledge and previous experiences, both affective and rational, are integrated in the amygdala and give emotional stimulation to both images and sounds. In other words, such a pathway could charge and convey emotional responses to objects.

A number of studies on people with prosopagnosia in the 80s proved that they would excrete micro-perspiration (measured with galvanic skin response (GSR)) when they looked at images of familial faces even though they did not recognise them. There were also reports of brain-damaged patients recognising faces, but they did not have this autonomous response to familial visual stimuli. Later studies on patients with congenital prosopagnosia indicate that they also have covert facial recognition and that it does not differ qualitatively from the norm [12]. So, could damage to a neuro-anatomic pathway that conveys affective information, with insufficient emotional stimulation of facial data as a result, lie behind the Capgras delusion?

A mirror image of prosopagnosia?

If people with prosopagnosia could get increased autonomous response (increased GSR) by looking at the faces of relatives they cannot visually identify, this could be deemed inconsistent with Capgras syndrome, since patients with Capgras syndrome will recognise the faces of their relatives, but their GSR would only slightly increase or not at all. This was also the case in one study of five patients.[11] The ten controls, five without a disease and five with a different psychiatric disease, had the expected autonomic response. How is this relevant? The important paradox of the Capgras syndrome delusion is that the face is indeed recognised, but the authenticity, or identity if preferred, is denied. The idea is that a lack of connection between the amygdala and visual areas results in an 'error' and that the delusion is allowed by another

dysfunction. Reduced metabolism (activity) in the right-brain hemisphere (lateral prefrontal cortex) has been associated with delusions, for example, with Lewy body dementia, and is also suspected with Capgras syndrome [13,14]. Thus, the presence of damage, other psychiatric disease, delirium or dementia also seems to be necessary. However, is this true?

Impaired ability to identify rather than impaired facial recognition?

Let it be clear, a lack of autonomic response to familial faces has been detected in psychiatric patients who do not suffer from Capgras syndrome and during acute psychological crises.[7] In addition, several case reports have been published in which the delusion of an imposter is not based on visual stimuli, for example, in blind patients [15]. As mentioned, delusional misclassification syndrome can also be connected to objects, animals, places and one's self. The common denominator is a monothematic illusion created by the sensory system about something familial that results in a bizarre conclusion. Patients with prosopagnosia can identify people by using other information than faces, such as hairstyle, voice, walk and apparel. Neural networks that allow the perception of something 'familial' are probably influenced by a number of sensory modalities that are possibly integrated into the perception of one's own identity (the self). Objects that we are particularly attached to might be specially processed in the brain to give an automatic individual type of 'glow'. An important cognitive process such as losing this glow and discovering that one's expectations have been let down (in the prefrontal cortex) might be disturbed in those who develop Capgras delusions. New data supports this type of model [16].

Through so-called lesion network mapping in studies on familial perceptions, it was possible to prove in 17 patients with damage-associated syndrome of subjective doubles that all damage (lesions) was localised in networks functionally linked to the left retrosplenial cortex; the region with greater activity during functional magnetic response imaging in studying what is perceived familiar. All lesions, except one, were also functionally linked to the right prefrontal cortex. This is the most active region during functional magnetic response imaging of the brain with the presence of deviations related to expectations based on earlier experiences and known patterns (*belief evaluation and prediction error*) [17]. There was also evidence of impairment in the right prefrontal cortex in fifteen of those with lesions associated with delusions not connected to Capgras syndrome, however none of the lesions were connected to areas associated with the perception of 'familial'. In terms of neuroanatomics, these findings do not fully correspond with Ellis and Young's model, but the hypothesis holds true [10]. Could the findings indicate that one lesion could explain it all?

Complex neuropsychiatric syndrome – one lesion?

The retrosplenial cortex is a small area along the centre line of the brain just behind the splenium of corpus callosum, as indicated by its name, i.e. in the posterior section of the corpus callosum. It also includes some of the posterior cingulate gyrus, an area associated with focused attention when activated. Retrosplenial cortex is involved in a myriad of functions involving integration of information from the cortices (prefrontal cortex, and parietal and occipital cortices) and sub-cortical areas for memory (hippocampus, thalamus). Reduced activity in the region (proven with a PET scan) has been observed with mild cognitive failure and is one of the first signs of Alzheimer's disease. Pathological changes in the area have also been associated with diseases such as schizophrenia and bipolar disorder. Even more interesting is that isolated damage to the retrosplenial cortex could cause a selective defect in spatial orientation. In most of the reported cases, the patient had problems with orientation in previously known surroundings, but can adequately navigate around non-familial areas following landmarks [18]. That is, they have topographic agnosia similar to Mr I.

Conclusion and Reflection

Twelve years have passed since I first met Mr I. There are definitely no signs of an isolated brain damage. He still has the same neuropsychiatric problems, but his cognitive ability has deteriorated and the traits of Parkinson's disease are more pronounced. His brain is deficient in dopamine and he needs supplements. He has now developed dementia, a condition associated with lack of acetylcholine in various networks. Therefore, he started treatment with a cholinesterase inhibitor (rivastigmine), which probably had minor temporary positive effect on his cognition, and the Capgras phenomenon disappeared for a few months, but returned, and still seems to be triggered most often by his wife's tonal change in voice. Good effects on Capgras-like syndrome in patients with similar form of dementia and parkinsonism, Lewy body dementia, have been reported previously [19]. Non-surprisingly, it has been reported that drugs affecting other signal substances, such as dopamine receptor antagonists and selective serotonin reuptake inhibitors, have good effects on some patients as well.

I was a student in the early 90s when I came across the term prosopagnosia, but it did not register until a few years later after reading "The Man who Mistook his Wife' [1]. Since then, I have devoted a considerable amount of time studying and pondering over the human brain. The reason for who we are and the illusions we have is undoubtedly coded in the network of the brain. Important *locus in quo* (place where things happen) can be identified, but our knowledge of the human ability to imagine can still be compared to glimpses of far distant planets in a telescope.

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