Persistent Cotard’s Syndrome following Surgery for Right Temporoparietal Meningioma

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ABSTRACT

Patients with Cotard’s syndrome (CS) deny their own existence or the existence of their internal organs. The condition is controversial in terms of etiology and pathophysiology and is mainly described in psychiatry. We present the case of a patient with no previous history of psychiatric disorder, manifesting chronic CS secondary to neurological disease. We analyze the neurobiological basis of the syndrome as a complex perception disorder.

Key words: Cotard’s syndrome, right hemisphere, pathogenesis

INTRODUCTION

Cotard’s syndrome (CS) is a rare condition whose main feature is a nihilistic delusion in which patients believe that they lack some parts of the body or that their organs are malfunctioning. Patients with more severe forms can even deny their own existence.[1,2] Classically described in patients with psychiatric disorders, the condition is usually transient and responds to treatments targeting the underlying disorder.[1,3] Its character as a nosological entity has been questioned. Whether CS should be considered as a syndrome in the context of psychiatric disorders is also debated; in fact, it is not listed as a specific disorder in the fifth revision of the diagnostic and statistical manual of mental disorders (DSM-5).[4] Previous studies have described isolated cases of CS in the context of systemic disease (typhoid fever and adverse drug reactions) and mainly of neurological diseases of varying etiology (dementia, cerebral infarction, venous thrombosis, tumor, epilepsy, multiple sclerosis, migraine, vascular malfunction, focal brain atrophy, encephalitis, and trauma).[5-6] In these cases, researchers have tried to associate CS with lesions to or hypofunction of specific cortical areas. For some authors, the lesion would facilitate delusion in some patients with a certain previous personality, which could also be a determining factor in the appearance of delusion in the context of a psychiatric disorder.[7] We report a case of CS (manifesting persistently in a patient with no history of psychiatric disorder) linked to a post-operative lesion to the right temporoparietal association cortex. Considering this case and the available literature, we analyze the pathophysiology of CS as a complex perception disorder.

CASE REPORT

Our patient is a 54-year-old woman, who has no relevant psychopathological history and is well-adapted socially and professionally.

She was assessed due to decreased level of consciousness and left hemiparesis, with ipsilateral supranuclear facial paresis preceded by intense headache and difficulty walking, which had persisted for several months. Brain computed tomography (CT) and magnetic resonance imaging (MRI) scans revealed a large expansile process in the right temporoparietal region with subfalcine herniation and homogeneous contrast uptake, radiologically compatible with meningioma. She underwent

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total exeresis of a tumor measuring 8 cm in diameter and attached to the dura mater of the convexity. At a histological level, the tumor was formed by spindle cells with no atypical features or mitosis and corresponded to Grade I meningioma (WHO). After surgery, the patient showed a positive neurological outcome with no sequelae. 2 months after the intervention, she began to experience anxiety and depressive symptoms, together with cenesthopathic hallucinations which she described as “a tingling sensation under the skin” and “itching” on the left side of the body and the head. Symptoms progressed to the point that she experienced nihilistic delusion of the absence of organs, which she expressed as: “I have no brain, esophagus, or bones,” “I cannot eat because I have no stomach,” “I cannot feel the air on my skin or when I breathe,” “I cannot see any colors,” “how could I walk if I don’t have a brain?,” and “My body is melting.” A complete blood test (including thyroid hormones, vitamins, and serology) and an electroencephalography yielded normal results. Follow-up CT and MRI showed a post-operative malacic area in the right temporoparietal region, with no findings suggestive of tumor recurrence [Figure 1]. A single-photon emission CT scan (single-photon emission CT with Tc-99m) showed hypoperfusion in the same region [Figure 1]. We started treatment with intramuscular haloperidol and fluoxetine, which improved depression symptoms but did not resolve the delusional ideation. 3 years after the first hospitalization, the patient experienced a severe exacerbation due to treatment discontinuation. The patient still presented Cotard-like delusional ideation: “My body is burnt on the inside,” “I cannot walk because I have no legs,” “I do not have feelings,” and “I have the brain of a sick person.” She also manifested behavioral disorganization and refused to eat; she was hospitalized once more for these symptoms. Treatment was changed to olanzapine and sertraline, which improved her condition; she began eating and caring for herself once more and was discharged. However, delusional ideation persisted for the following 2 years, becoming chronic. After that time, the patient was lost to follow-up at outpatient consultations and her outcome is unknown.

DISCUSSION

CS is an infrequent delusion, which has awakened great interest since its description in 1880; however, published studies are limited to very short series or isolated cases. A 1995 review found 100 cases in the literature, although another review of cases published before 2007 includes only 35 cases of CS with neurological correlate.[1,5] A recent publication identified only 12 CS cases in the Mayo Clinic database over the past 20 years; however, it is possible that the entity was underdiagnosed.[4]

Since its initial description, CS has been conceptualized in two fundamental forms: As a subtype of psychotic depression...
and as a non-specific syndrome unrelated to melancholia. Most cases correspond to patients with another psychiatric disorder. Three main psychiatric conditions have basically been associated to CS: Depression, schizophrenia, and psychiatric disorders with an organic brain lesion.\[8\] Despite having specific features, CS currently lacks nosological specificity.\[4\]

Numerous hypotheses have been discussed concerning its etiopathogenesis, reflecting the underlying complexity of the condition.

Among neurobiological theories, some authors have linked CS with specific brain lesions, especially at the temporoparietal junction, an important multimodal area which incorporates inputs from the tactile, proprioceptive, visual, and vestibular systems. This area is also associated with bodily self-consciousness.\[8-13\] Thus, most articles on CS which include neuroimaging findings report bilateral lesions affecting the prefrontal and temporoparietal cortices, with the most common being the right parietal lesions.\[10\] In fact, there have been published cases of CS with lesions affecting the right parietal cortex exclusively.\[14,15\] Our case would support the hypothesis that the delusion derives from a disruption of the circuit contained in that region.

Nihilistic delusion and delusional misidentification syndromes, particularly Capgras delusion, have frequently been associated with the right hemisphere lesions.\[3,5,9\] Young et al. studied the relationship between Capgras delusion and CS, concluding that both delusions may originate from a delusional interpretation of an altered perception; the key element would be a loss of the emotional feeling of familiarity.\[16\] The fact that some patients with neurological lesions to the right hemisphere present delusions of death together with misidentification delusions suggests that abnormal feelings of familiarity may be the common basis for the co-occurrence of both delusions.\[14\] Alexander et al. argued that the pathophysiological explanation may be a disconnection of the right temporolimbic region from both frontotemporal lobes, which would distort the integration of feelings and emotional memory.\[17\] Another hypothesis, proposed by Hirstein et al., is that CS may involve a dysfunctional connection between the limbic system, especially the amygdala, and the parietal sensory cortex.\[18\] Therefore, there would be no access to emotional memory and the delusion would manifest as a response to the pathological situation of not experiencing feelings of familiarity, giving rise to an erroneous interpretation of perception experiences.\[3,10,11,18\] Several authors conclude that the right temporoparietal junction is a key to connect the flow of information from sensory areas to the limbic system, facilitating affective processing of perception experiences and originating the integration of a structural model of the body.\[11,13]\n
Other neurological conditions which should be differentiated from CS but which have some similar elements are asomatognosia (loss of recognition of one’s own limbs) and anosognosia (inability to recognize a disability or deficit or integrate it into bodily experience), manifesting mainly after lesions limited to the right hemisphere, especially to the right temporoparietal and posterior parietal cortices.\[11,14,19\] Regarding the relationship between asomatognosia and CS, some studies insist on the differential characteristics between them, although case reports of patients with both CS and asomatognosia are also available on the literature.\[9,11,20\] However, similarities between asomatognosia and depersonalization have been known for years: The term “total asomatognosia” to refer to depersonalization was coined by L’Hermitte. Likewise, the significance of depersonalization in the origin of delusions of negation had already been mentioned by Ségas (1889), who thought that an alteration in the mental synthesis process would prevent the patient from relating images with other feelings and memories, leading to a modification of personality, and finally to the appearance of nihilistic ideas.\[21\] The most recent neurobiological model of depersonalization points to the existence of a corticolimbic dysfunction.\[22\] In this sense, depersonalization and nihilistic delusion may be interpreted as a “visceral anesthesia” or “the extreme end of agnosia,” leading to an inability to recognize oneself and create a correct image of one’s own body schema (self-consciousness).\[21\] A recent functional neuroimaging study positron emission tomography (PET) scans revealed significant hypermetabolism in the parietal, temporal, and occipital sensory cortices of patients with depersonalization compared to healthy controls.\[22\] Other studies have reported the right fronto and temporoparietal hypometabolism in patients with CS.\[23,24\] These findings support the idea that the posterior parietal cortex and the temporoparietal junction are the areas where the body schema is integrated (multisensory areas where visual, auditory, and somatosensory information converge).\[13\] Other studies highlight the importance of the insula as the central processor of interoception and structure responsible for the integration and conscious awareness of internal sensations. The connection of the insula with frontal, parietal, and limbic areas (especially the anterior cingulate cortex) would be the circuit which constitutes the underlying basis of conscious experience of selfhood and one’s own body schema.\[10,12\]

We report a case of CS with an organic cause, secondary to a right temporoparietal glial tumor. The manifestation of delusion may require a latency period from the appearance of the noxa, as described in other case reports.\[8,14\] Furthermore, given the absence of previous depressive symptoms or psychodynamic factors, the patient developed pure CS (Cotard Type 1, according to Berrios).\[1\] In comparison with delusions of negation of existence, the predominant clinical symptoms were nihilistic ideas concerning parts of her body and body functions. She also manifested
Cenesthesiopathic hallucinations. Refusal to eat is also in line with delusions. We should highlight that our patient did not manifest asomatognosia or visual involvement at any time. We found few cases in the literature of CS related to conditions exclusively affecting the right hemisphere.\textsuperscript{[14,20]} A very interesting report described a case of sudden-onset CS associated with the right temporoparietal glioblastoma.\textsuperscript{[15]} However, we should mention that we have found no other case in the literature with such a prolonged progression of delusion as in the case described (minimum 5 years of follow-up).

CONCLUSIONS

CS is an infrequent and possibly underdiagnosed condition, which is of significant clinical relevance due to its high vital risk (self-injury, refusal to eat, and suicide). The pathophysiology of the syndrome remains uncertain, although several interesting neurobiological mechanisms have been proposed and merit further study. In light of this case, it is important to highlight the need for a thorough etiological study including screening for organic brain lesions. Furthermore, researchers have already stressed the importance of publishing CS cases including neurological findings to better understand the underlying correlate.\textsuperscript{[19]} As suggested by other authors, CS should be reconsidered as an entity itself, especially in its pure forms, as it is a very characteristic delusion which can predominate among clinical symptoms.

REFERENCES
