

CASO CLÍNICO/CASE REPORT

Cotard's Syndrome in Adolescence: A Case Report Síndrome de Cotard na Adolescência: Um Relato de Caso

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Abstract

Cotard's syndrome is a rare neuropsychiatric condition characterized by nihilistic and hypochondriacal delusions that generally occur in individuals with some predisposition or in association with a psychiatric or organic disease. Its development is even more unusual in pediatric population.

The purpose of this paper is to briefly review the available scientific evidence, based on the description of a clinical case. Data were obtained from clinical records and a careful search and analysis on the electronic databases PubMed and Embase.

We illustrate the case of a 15-year-old female adolescent with Cotard's syndrome in the context of an adjustment disorder. Cotard's syndrome can be associated with chronicity, self-neglect and suicide, but a prompt diagnosis and treatment can promote a good outcome. This case shows how the use of antipsychotics complemented by psychotherapeutic and psychoeducational interventions can promote a complete remission of the condition.

Resumo

A síndrome de Cotard é uma doença neuropsiquiátrica rara caracterizada por delírios niilistas e hipocondríacos que ocorre geralmente em indivíduos com alguma predisposição ou outra doença psiquiátrica ou orgânica. O seu desenvolvimento é ainda mais raro na população pediátrica.

Este artigo tem como objetivo rever brevemente a evidência científica disponível, a partir da descrição de um caso clínico. Os dados foram obtidos a partir de registos clínicos e da pesquisa criteriosa nas bases de dados electrónicas PubMed e Embase.

Ilustramos o caso de uma adolescente de 15 anos com síndrome de Cotard no contexto de uma perturbação de ajustamento. A síndrome de Cotard pode associar-se a cronicidade, auto-negligência e suicídio, mas o diagnóstico e tratamento precoces podem promover um bom prognóstico. Este caso mostra como o uso de antipsicóticos complementado por intervenções psicoterapêuticas e psicoeducativas pode promover a remissão completa do quadro clínico.

Keywords: Adolescent; Delusions

Palavras-Chave: Adolescente; Delírios

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INTRODUCTION

Cotard's syndrome (CS) is a rare and severe neuropsychiatric condition in which the central feature is the existence of nihilistic and hypochondriacal delusions.¹⁻³ CS is more common in middle-aged or older females,^{1,4} being extremely rare in adolescents.⁵⁻⁷ Studies in adult population suggest a prevalence of 0.57% - 3.2%.^{7,8}

Given its rarity, undefined neurobiology and frequent association with other conditions, CS is frequently described as part of an underlying disorder³ and it does not appear as a specific nosological classification in Diagnostic and Statistical Manual of Mental disorders Fifth Edition (DSM-5) or International Classification of Diseases 11th Revision (ICD-11). In DSM-5 there is only a reference to nihilist delusions in the description of depressive episodes with psychotic symptoms congruent with the humor.⁴

CS was firstly described in 1880 when Jules Cotard proposed that depressive and anxiety symptoms would serve as a ground for the development of a systematized negative delusion, with hypochondriacal and nihilistic ideas of denial, malfunction or non-existence of the body, body parts or organs, including the belief of being dead.^{1,2,5,6} These symptoms may be associated with self-starvation, dehydration and disinvestment in self-care.^{1,5} Severe forms can present with behaviors of self-injury, suicide ideation or suicide attempts.^{9,10} Some patients can experience hallucinations, including olfactory, frequently describing a bad smell caused by body decomposition, but also kinaesthetic, auditory, visual, or tactile hallucinations. Frequently patients demonstrate feelings of hopelessness, worthlessness and uneasiness.⁹ Symptoms can either be sudden or emerge progressively.^{2,6} Currently, CS is mostly described as a symptom cluster that occur in individuals with some predisposition or in association with a psychiatric or organic disease.² Recent epidemiological studies show that CS is frequently associated with depressive, bipolar and anxiety disorders or schizophrenia, but has also been reported in patients with multiple organic conditions, such as neurosyphilis, typhoid fever, epilepsy, cerebral infarction, subdural hemorrhage, cerebral arterio-venous malformation, parietal lobe tumors, Parkinson's disease, dementia and multiple sclerosis.^{1,2,4,6,9} A systematic review found a relationship between the age >25 years and the diagnosis of major depression or an organic illness.¹⁰ There is evidence that CS may emerge from an hypochondriac setting, particularly in the context of a newly identified diagnosis, a recent surgical intervention or when a new symptom or complaint appears.¹¹ An association with stress events, such as the death of a family member or economic problems was also described.⁹

In literature, some authors identify three distinct groups of patients: (1) patients with psychotic depression, with symptoms of melancholic depression and rare nihilistic delusions, (2) patients without depression (CS type I), whose nosology is closer to a psychotic disorder (functional or organic), and (3) patients who show anxiety and/or depressive symptoms associated with hallucinations (CS type II).^{2,5,7} Nevertheless, this distinction is not consensual in the scientific community.⁹

Despite the fact that some authors suggest the implication of fronto-temporo-parietal cortical circuits in the pathophysiology of CS, most studies describe absence of abnormalities in the neuroimaging exams.^{8,12} Even in the most serious cases, CS can resolve spontaneously and suddenly. Therefore, it is recommended to initiate treatment as soon as possible, including specific treatment towards the underlying condition if that's the case.⁷ First line treatment is poorly established, but antipsychotics appear to be effective and can be combined with antidepressants and anxiolytics.⁶ Electroconvulsive therapy is described as an alternative in severe cases.^{7,8} Some authors report potential benefit of combining psychotherapy and pharmacological treatment.⁴ The prognosis seems to be determined mostly by the establishment of an early medical intervention and the evolution of the underlying disorder.^{1,6}

CASE REPORT

The case is about a migrant 15-year-old female adolescent living with her parents and brother in Portugal for two months.

She was brought to the emergency room (ER) by her mother with complaints of tingling on the left side of her body with one month of evolution, initially on her face with descending progression through her neck, torso, abdomen and pelvis during the last week, accompanied by reduction of food intake and initial insomnia. The patient was in the 9th grade, but was not yet attending school in Portugal, which made her apprehensive about her future academic performance. Her former karate instructor had recently died of suicide, fact which she learned a month previously to the observation. Pregnancy, childbirth and psychomotor development occurred without complications. She was not taking any medication. There was no personal history of medical or surgical conditions, nor substance use. There was no family history of medical or psychiatric conditions.

In clinical observation the patient was awake, oriented in person, time and space, complaining of discomfort. She had psychomotor retardation and an induced speech that was limited to the description of hypochondriacal concerns. The patient described ruminative thoughts and nihilistic delusions related to herself, stating that her body was "inflamed" and "decomposing" and that she was dying. There were also present kinesthetic, somatic and olfactory hallucinations including the sensation of "bugs walking" inside her body, the feeling of fetid cold air coming out of her mouth, nose, ears and vagina, and the sensation of having the left arm detached from the rest of her torso. She had little insight for her condition.

Physical and neurological examinations at the ER were normal. An analytical routine screening was carried out (blood count, renal function, ionogram, liver profile and drug testing), as well as a chest x-ray, an electrocardiogram and a brain computerized tomography (CT) scan, all of them without any alterations.

The patient and her family accepted the treatment plan. Antipsychotic medication was initiated and the patient was

admitted to the inpatient child and adolescent psychiatry unit for clinical stabilization and extended etiological study. During hospitalization, an initial comprehensive complementary assessment was carried out. The patient was again evaluated by the pediatrics and neurology teams, with normal physical and neurological complete examinations. It was also performed an electroencephalogram (EEG), a brain magnetic resonance imaging (MRI), an abdominal ultrasound and multiple analytical evaluations, including hemogram, renal and thyroid function, ionogram, liver profile, viral serologies, VDRL test, folate and vitamin B12 dosages, all without alterations. A psychological assessment was also carried out, which suggested a borderline intellectual functioning. This evaluation had some limitations due to different cultural background since the patient was from a developing country.

Initial pharmacological treatment consisted of olanzapine 5 mg/day and diazepam 10 mg/day. Treatment plan also included daily therapeutic group activities and 3 to 5 therapeutic consultations per week with the attending child psychiatrist. Four days following the admission the patient showed significant symptomatic improvement, maintaining only cervical tingling and sensation of fetid cold air coming out of her nose, with insomnia resolution. On the 7th day after admission complete remission was observed and diazepam was changed to a SOS regimen.

On the 8th day of hospitalization, the adolescent described a toothache on the left inferior dental arch, revealing concerns about going to the dentist that improved with a psychoeducational intervention. She was observed by stomatology team and underwent a careless tooth extraction, with total symptomatic remission.

On the 18th day of hospitalization a switch to aripiprazole 10 mg/day was made following a reduction of thyroxine (TH) levels. A thyroid ultrasound was performed which was considered normal. After 6 days TH and TSH levels were in the normal range.

The child and adolescent psychiatry team concluded that the patient presented with a set of symptoms compatible with CS including nihilistic delusions and congruent hallucinations, in the context of (1) a possible adjustment disorder, (2) a recent medical symptom (toothache) associated with hypochondriac concerns, and (3) a mild intellectual development disorder.

On the 25th after the admission the patient was discharged, oriented in all references, with euthymic mood and referring no concerns. The delusional ideas of body decomposition and death, as well as the hallucinations, were absent. She recovered from sleep difficulties and developed partial insight regarding the symptoms. She was referred to a psychiatric outpatient clinic in her residence area. Before the first appointment, she had 2 post discharge consultations in the 6 weeks period after hospitalization, where it was possible to confirm maintenance of symptomatic remission with pharmacological treatment with aripiprazole 10 mg/day. After 6 months, the patient showed complete symptomatic remission after tapering off the medication.

DISCUSSION

CS is a rare disease without a specific nosological classification.² Nevertheless, psychiatrists but also other doctors, should be aware of CS in order to be able to diagnose and initiate the appropriate treatment as soon as possible.⁹ Since CS is frequently associated with another psychiatric and organic disorders, a comprehensive initial approach of the patient is essential.^{1,2} It should include a physical and neurological examination, and a psychiatric assessment, including previous history. It also includes imaging exams and a complete laboratory analysis to exclude the presence of an organic condition.⁹ When an underlying disorder is identified, attention should be primarily directed to its treatment. If there's a self-injury or suicidal risk, appropriate security measures must be ensured.⁴ The use of antipsychotics in monotherapy has proved to be effective in the management of CS.⁹ The combination with antidepressants and/or anxiolytics has shown to be more effective than antipsychotics alone in the presence of depressive and anxiety symptoms.⁹ ECT is reserved for severe or refractory cases, although its use is described in the literature as a first-line treatment in a variety of patients with an underlying mood disorder.^{1,9} Although studies describe cases refractory to treatment with chronic evolution particularly in patients with depression and schizophrenia, spontaneous recovery is also reported in CS associated with organic conditions.¹³ Evidence shows that an early medical intervention is associated with better outcomes and it can lead to a complete recovery.^{1,7,9} Given the high risk of young patients with CS to exhibit a bipolar outcome, they should be closely monitored for the onset of symptoms at follow-up consultations.¹⁴

We illustrate a rare case of CS in an adolescent patient that included the typical self-related hypochondriacal and nihilistic delusions associated with multimodal hallucinations. The clinical picture seems to correspond to a CS type II given the presence of anxiety symptoms and hallucinations, in association with an adjustment disorder (moving to Portugal, not attending school and having received the news about the death of her former karate professor) and the recent emergence of a toothache causing hypochondriacal concerns. This case illustrates that pharmacological treatment with antipsychotics, accompanied by psychotherapeutic and psychoeducational interventions, can be an adequate treatment of CS without other specified medical condition. The expected prognosis was favorable since treatment was started early, with a good and fast response, and CS was not associated with a psychiatric disorder like schizophrenia or depression.¹³ Limitations of this case report include the lack of previous research studies on this topic, the different culture of the patient and her family and the short follow-up duration.

Since CS is rare and its underlying pathophysiology is still largely unknown, its description in case reports has special relevance in helping to answer many questions about epidemiology, associated diseases and prognosis.

Declaração de Contribuição

NAB: Desenho do trabalho, interpretação de dados, redação, revisão e aprovação final.

MMN e MM: Desenho do trabalho, interpretação de dados, revisão e aprovação final.

Contributorship Statement

NAB: Manuscript conceptualization, data interpretation, writing, review and final approval.

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