A case of obsessive symptoms in Huntington's disease

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Un caso de síntomas obsesivos en la enfermedad de Huntington

Summary

Background. *Recently, some authors have postulated the existence of the so-called «obsessive spectrum». This would include disorders that have the common point of the presence of repetitive thoughts and actions, which may either have a «compulsive» or «impulsive» nature. Certain neurological disorders with repetitive movements have also been grouped within this spectrum, including some forms of epilepsy, Parkinson's disease, Sydenham's Chorea or Huntington's disease. There is evidence that physio-pathological connections may exist between some of these conditions.*

Methods. *A case of a patient with Huntington's Chorea with obsessive signs and symptoms is presented. The patient is a 50 year old male, diagnosed of Huntington's Chorea who is being treated with 25 mg/day of clozapine and 10 mg/day of clorazepate. The patient refused to eat certain foods for «fear of choking». He began to display other obsessive behaviors such as refusing to touch objects that might have been touched by others, and developed ritual checking methods. Treatment based on the administration of 40 mg/day of paroxetine was begun. With this treatment the compulsive symptoms have disappeared, as has the fear of choking. However, the choreic movements typical of his condition persist and the signs and symptoms associated with a process of dementia are becoming increasingly evident.*

Conclusions. *It is concluded that the case described may well be a good example of obsessive spectrum, and that the underlying physio-pathology should be studied.*

Key words: *Obssesive spectrum. Huntington's disease. Obsessive-compulsive disorder.*

Resumen

Fundamento. *Recientemente, algunos autores han postulado la existencia del llamado «espectro obsesivo». En él se encontrarían trastornos que tendrían como punto en común la presencia de pensamientos o actos repetitivos, ya sean estos de tipo «compulsivo» o «impulsivo». Ciertos trastornos neurológicos con movimientos repetitivos se han incluído también dentro del espectro, como son algunas formas de epilepsia, la enfermedad de Parkinson, la corea de Sydenham o la enfermedad de Huntington. Existen indicios de que pudieran existir vínculos fisiopatológicos entre algunas de ellas.*

Métodos. *Se presenta el caso de un paciente de corea de Huntington con sintomatología obsesiva. Se trata de un varón de 50 años, diagnosticado de corea de Huntington, y en tratamiento con clozapina 25 mg/día y clorazepato 10 mg/día. El paciente se negaba a tomar ciertos alimentos por «miedo a atragantarse». Comenzó a presentar comportamientos tales como evitar tocar objetos que pudieran haber tocado otras personas, además de rituales de comprobación. Se instauró un tratamiento de 40 mg/día de paroxetina. Con ello, la sintomatología compulsiva ha desaparecido, como también el miedo a atragantarse. Persisten, no obstante, los movimientos coreicos propios de su enfermedad y cada vez son más evidentes los signos y síntomas propios de un proceso demencial.*

Conclusiones. *Se concluye que el caso reportado puede ser un buen ejemplo del espectro obsesivo, y se estudia la fisiopatología subyacente.*

Palabras clave: *Espectro obsesivo. Enfermedad de Huntington. Trastorno obsesivo compulsivo.*

INTRODUCTION

Huntington's disease (HD) is a picture of autosomal dominant genetic transmission characterized by choreic movements and progressive dementia[1]. Other frequent psychiatric manifestations are depression, psychosis, and personality disorders[2]. This disease was recognized as a nosological entity after its description by George Huntington in 1872, although there are previous descriptions of hereditary choreas such as those of Amstrong (1783), Bernt (1810), Mongeot in 1815, Coste in 1827, Elliotson in 1832, Waters in 1841 or Duglison in1842[3].

The obsessive-compulsive disorder (OCD) is characterized by the presence of obsessions (persistent ideas, thoughts, impulses or images that the individual considers to be intrusive or inappropriate) or compulsions

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(recurrent behaviors or mental acts, whose purpose is to prevent or alleviate anxiety or malaise), that are serious enough to cause significant loss of time, marked deterioration of the general activity or clinically significant malaise. The first descriptions of that known as OCD at present can be found in the *Traité des scrupules* of Jacques-Joseph du Guet (1717), that speaks about the «scruples disease». The term obsession appeared in the psychiatric language in the middle of the XIX century, the obsessive phenomena being described under multiple denominations: reasoning monomanias (Esquirol, 1838), fixed ideas (Parchappe, 1851), lucid insanity (Falret, 1864), disease of doubt with delusion of touching (Legrand du Sulle, 1875), or onotomamia (Charcot and Magnan, 1885). In addition, the term compulsion comes from the German term *Zwangvorstellung,* proposed by Krafft-Ebing in 1867 to name the «compulsive representations»[4].

The existence of the so-called «obsessive spectrum»[5] has been hypothesized in recent years. In the «obsessive» end of the spectrum, clinically characterized by the presence of repeated ideas, we find OCD, body dysmorphic disorder or hypochondria, while in the compulsive end, we find ludopathy, kleptomania or trichotillomania. Certain neurological disorders with repeated movements have also been included within the spectrum, such as the HD itself, some forms of epilepsy, Parkinson's disease, or Sydenham's chorea. There are signs that these disorders share a common neurobiological substrate, involving the brain circuits in which orbitofrontal connections, basal and limbic ganglia participate.

CLINICAL CASE

A 57 year old male from rural setting. He belongs to a family with abundant background of HD (fig. 1). He was referred to the Mental Health Care Team (MHCT) by his family doctor in February 2001 because he was having problems in obtaining authorization from drug inspection for treatment with clozapine, a drug that had been prescribed by a specialist in Madrid. At the time of referral, he was receiving treatment with: clozapine 25 mg/day; tetrabenazine 120 mg/day and nicardipine 60 mg/day.

Since his youth, his behavior was characterized by impulsivity, some sexual disinhibition and sporadic alcohol abuses. At 40 years of age, he began to suffer motor problems (choreic movements of hands and legs), and was studied and diagnosed of HD.

In the examination of the first interviews in the MHCT, the patient was conscious and oriented both allo- and autopsychically. He showed a mildly subdepressive mood. He reported anxiety and anguish. There were no disorders of the course, thought content or sensoperception. There was mild deterioration in immediate memory and fixation. Choreic movements of the lower limbs and phonatory difficulties were observed. In the first visit in the MHCT, anxiety was already detected, so that 15 mg/day of cloracepate were added.

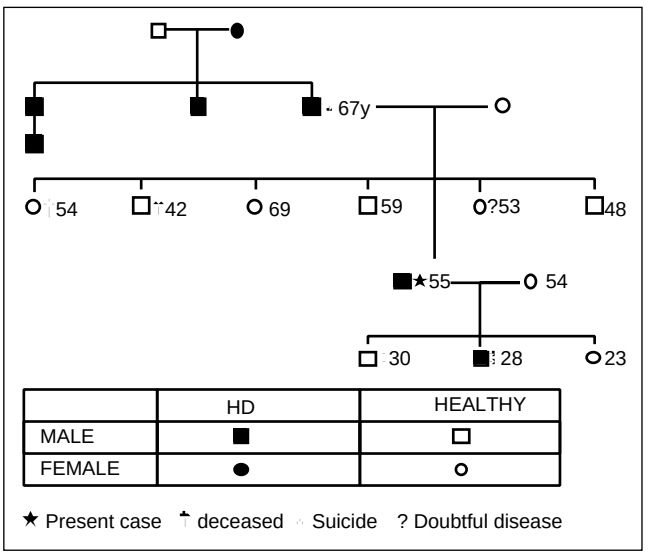

Figure 1. Genealogical tree of the patient.

In January 2002, his depressive symptoms worsened after the suicide of a child who also suffered HD. He suffered was significant social withdrawal, so that it was decided to add paraxetine (20 mg/day) and reduce the cloracepate, since he complained of excessive sedation.

During the summer of 2002, he began to refuse to eat certain foods due to fear of choking, even though the neurologist had discarded the existence of pharyngeal or esophageal neuromuscular problems. He also avoided touching certain objects, such as the toilet seat cover, the stair railing, or the doorknob, and refused to have contact with his dog «to not get stained». He was very observant of the times that he had to take his medication, while he had completely ignored this subject before (until them, the entire responsibility of the treatment was held by his wife). The paroxetin dose was increased (40 mg/day). With this treatment, the avoidance behaviors disappeared completely and he no longer was concerned about the time schedules. His mood tone has improved, although social withdrawal persists. In regards to cognitive deterioration, this has worsened, episodes of disorientation, even within the home itself, appearing.

DISCUSSION

Phobic symptoms, with avoidance behavior, and fears of infection that could lead to the diagnosis of a phobia appear in this patient. However, it is frequent that obsessive patients with contamination ideas (which are present in the case described, since he fears «becoming stained») develop avoidance behaviors that can lead them to try to not enter into contact with contaminated objects or those that can transmit contamination[6,7]. On the other hand, Marks[8] has shown the phenomenological proximity between obsessive disorders and pho-

bias, establishing a classification in which the following are distinguished: a) fear of external stimuli (equivalent to phobic neurosis): phobias of animals, agoraphobia, social phobias and other specific phobias, and b) fear of internal stimuli: nosophobias (halfway between hypochondria and other specific phobias) and obsessive phobias (in relationship with the obsessive-compulsive disorder)[9]. Thus, the patient could be diagnosed according to the ICD-10 as: F42.1 Obsessive-compulsive disorder, with predominance of compulsive acts.

There seems to be some common points in the physiopathology of the HD and the OCD. On the one hand, from the neuroanatomical point of view, the pathogenesis of the HD includes the loss of striate neurons, and their molecular pathology is focused on the selective vulnerability of the neurons of this brain region[11]. From the study of the obsessive symptoms associated to neurological diseases (Gilles de la Tourette, epidemic encephalitis of Von Economo, Sydenham's chorea and even the same HD syndromes) and the data from the neuroimaging studies, a neuroanatomical model of OCD has been proposed in which the frontal cortex, striate cortex, complex formed by the internal nucleus pallidus and substantia nigra, basal ganglia and thalamus in the OCD disease (fig. 2)[12,13]. Following this anatomical model, the obsessive signs and symptoms would appear due to abnormalities of these circuits, either due to a primary hyperactivity of the frontal-strio-pallido-thalamic loop, that would cause excessive thalamic disinhibition, or due to the increase of the primary activity of the orbitofrontal-thalamic interconnection, or perhaps due to a relative increase of the direct pathway activity, in comparison with the indirect pathway. In the case in question, as the striate is injured, there could be hyperfunction of the indirect circuit, which would give rise to obsessive thoughts.

Cummings[14] published the case of two patients suffering from comorbid HD with OCD. The latter was manifested by repeated, stereotypal and complex behaviors that were egodystonic and incapacitating for the patients. The explanation of this author was coherent with the model described, since it stated that HD and other comorbid neurological syndromes with OCD (Gilles de la Tourette syndrome, neuroacanthocytosis, post-encephalic parkinsonism, caudate nucleus infarctions, carbon monoxide poisoning, manganese poisoning, anoxia, progressive supranuclear paralysis, Sydenham's chorea, and other frontal lobe lesions) would indicate that the frontal lobe, the caudate nucleus, and the globus pallidus form a part of a complex circuit that has an important role in the OCD.

Patzold and Brüne[15] have recently described a case of OCD in HD, that was treated satisfactorily with sertraline. They concluded that the degeneration of the caudate in HD may lead to the appearance of the obsessive-compulsive symptoms.

However, there is another explanation for the appearance of obsessive symptoms in this patient. It has been described that certain atypical neuroleptics, among which clozapine is found, may lead to the appearance of obsessive symptoms[16], which would be explained by the 5-HT_2 serotoninergic antagonism, above all when these drugs are taken at low doses[17], as is the case of this patient.

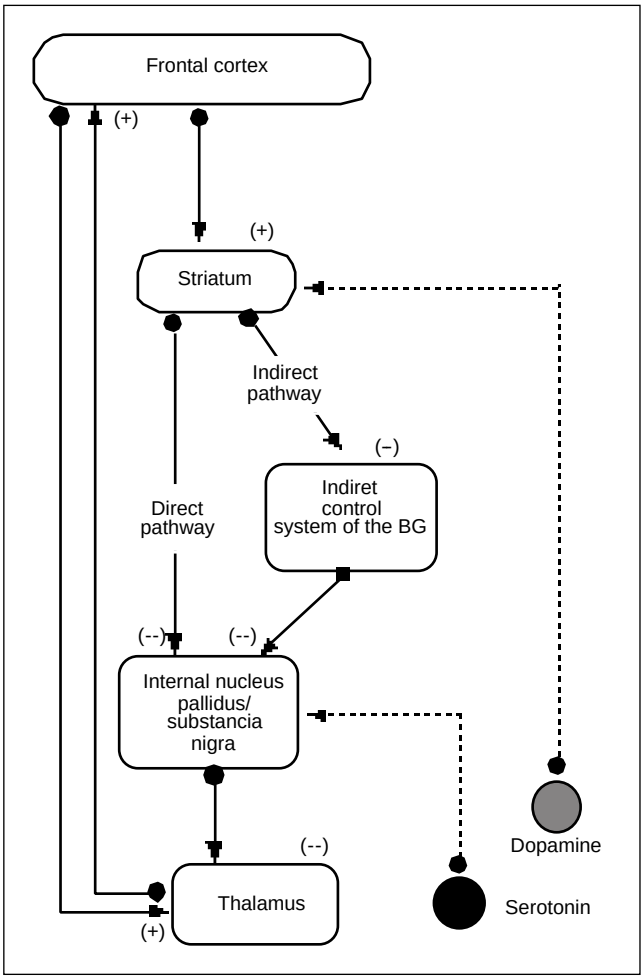

Figure 2. Frontal-subcortical circuits supposedly involved in the pathophysiology of the obssesive-compulsive disorder[13].

CONCLUSIONS

The case presented is an example of how the different disorders and diseases that are included within the obsessive spectrum may present the obsessive-compulsive symptoms, as a reflection of a common physiopathology.

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