The Charles Bonnet syndrome: a case report and a brief review

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Síndrome de Charles Bonnet: exposición de un caso y breve revisión

Summary

The Charles Bonnet syndrome (CBS) is a rare disease *that also seems to be generally misdiagnosed. Initially described in the XVIII century by the philosopher with the same name, it consist in complex visual hallucinations in elderly people* who suffer *sensory deprivation with no other psychopathology. The hypothesis on the neurophysiology of hallucinations suggests, as in other* diseases *that present hallucinations, some implication of thalamus-cortex pathway release. Some authors have proposed CBS as an early marker of dementia and Parkinson's disease. Nevertheless, the results in functional neuroimaging are not conclusive. Regarding treatment, typical and atypical neuroleptics do not seem to be useful and recent studies suggest that the new anticonvulsants could be effcient. The patient should be informed about CBS as a «non-psychiatric disease».*

In the case reported, the patient presented complex hallucinations with normal SPECT and neuropsychological examinations. *The patient did not respond to treatment with risperidone, presenting a favorable evolucion with valpromide. Although further research is needed, this case report supports the efficacy of valpromide in CBS.*

Key words: *Charles Bonnet syndrome. Visual hallucinations. Anticonvulsants. Valpromide.*

Resumen

El síndrome de Charles Bonnet (SCB) es una patología poco frecuente y que además suele ser infradiagnosticada. Descrita inicialmente en el siglo XVIII por el filósofo del mismo nombre, consiste en la aparición de alucinaciones visuales complejas en personas de edad avanzada y con deprivación sensorial sin otra psicopatología concomitante. Las hipótesis acerca de la neurofisiología de las alucinaciones responde, al igual que en otras patologías que presentan alucinaciones visuales, a una liberación de la vía tálamo-cortical. El SCB también se ha propuesto como marcador precoz de demencia y enfermedad de Parkinson. Sin embargo, los resultados en pruebas de neuroimagen funcional no son concluyentes. En cuanto al tratamiento, los neurolépticos, tanto típicos como atípicos, no parecen útiles, y últimamente se ha señalado la eficacia de los nuevos anticomiciales. Por otra parte, hay que señalar la importancia de informar al paciente del carácter «no psiquiátrico» de su trastorno.

En el caso que se presenta, el paciente refiere alucinaciones visuales complejas tras la pérdida completa de la visión, siendo las pruebas neuropsicológicas y el SPECT realizados normales. El paciente no respondió al tratamiento con risperidona, presentando una evolución favorable con valpromida. Aunque con reservas por tratarse de un único caso, esta exposición sugiere la eficacia del tratamiento anticomicial en el SCB.

Palabras clave: *Síndrome de Charles Bonnet. Alucinaciones visuales. Anticomiciales. Valpromida.*

INTRODUCTION

The Charles Bonnet syndrome (CBS) consists in an organic hallucinosis that appears in elderly persons who present sensorial deprivation. It is characterized by the appearance of complex visual hallucinations in patients no other psychopathology[1,2]. Classically considered a rare disease, some recent studies seem to contradict this idea, even though there are no solid data in this regards[2]. The treatment is controversial, generally because of the failure of neuroleptics and its evolution is variable[3]. In the following, based on one case, the data available for the diagnosis, therapeutic management and prognosis of this disorder are reviewed.

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CLINICAL CASE

This is an 81 year old woman, widow since four years ago, who has two children and who lives with a caretaker in the same building as her children. The patient is referred to the Psychiatry Office from Ophthalmo-

logy due to visual hallucinations after complete loss of sight.

It seems that after the complete loss of sight due to suffering endolphthalmitis, the patient began to «see rare things»: «people I do not know, and they greet me; children who sit at the table while I eat and they take my food; the physicians and nurses of the hospital, etc.». Even though the images are perceived as real images, there is a certain criticism on their existence: «although I know that they are not there, I see them exactly as if they were there, etc.». The image that is repeated the most is «the green corridor» (popular name of a typical walking area in the center of Madrid, an old train track that led to the Delicias station and that was reconverted into a park. The place is very near to the patient's home and she walked their daily with her husband for many years). These images intensified when the patient is alone and mostly at night, which makes it difficult for her to fall asleep and to stay asleep: «it does not let me sleep, all day with the green corridor». In addition, she experiences the hallucinations with intense anguish and presents behavior adequate to them (she fights with the «children» so that they do not take her food).

Since the surgery, she is more discouraged, she does not feel like doing anything and finds it difficult to manage to perform daily life tasks. She has many somatic complaints and the incapacity caused by her blindness is increased by the presence of significant arthrosis. She has no appetite alterations and does not report failures in the memory functions. Recently, she has begun rehabilitation treatment in a geriatrics center that she goes to daily.

In spite of the anxiety caused by the hallucinations, the patient is reluctant to receive psychiatric treatment. After a few minutes of interview, when she was questioned about the onset of the symptoms, the patient answered in the following way «I am not crazy or depressed, any normal person would be the same as I am in my situation». When asked about specific subjects, the patient focuses on her complaint, and when an attempt is made to lead to an answer, she tires «so many questions confuse me!»

In the psychopathological examination, the patient appears conscious and auto as well as allopsychically oriented. She is approachable and collaborates acceptably. Her mood is dysphoric, with tendency to irritability in the interview due to her lack of trust in the psychiatric treatment. Floating anxiety is observed and it intensifies with the hallucinations. There is some speech burden, with circumlocutions and answers not related with the questions. Her discourse is focused on the hallucinations, on the past and on her loss of sight. She presents vivid and complex polychromatic visual hallucinations which the patient criticizes but which create great anguish due to their realism, intensity and frequency. The picture becomes worse at night. There are no other sensoperceptive disorders, or delusional ideation or phenomena of theft/diffusion of thought. Occasional death ideas without ideation or suicide planning are identified. Judgement and abstraction ability and intelligence are apparently conserved. Persistent conciliation Insomnia and early waking type. Conserved appetite. No alterations are observed in memory function in the gross examination. The patient is aware of her somatic disease but not of mental one.

Regarding her background, no personal or family psychiatric background is identified. Regarding medical background, the existence of high blood pressure under treatment and significant arthrosis stands out. She has had right ophthalmopathy since childhood. She was operated on one month ago for cataracts in her left eye. The surgery was complicated by an endophthalmitis that required evisceration of the eye with complete loss of vision. Some days after the surgery, she presented an episode compatible with delirium that was resolved during her hospital stay with neuroleptic treatment with 1 mg of risperidone (treatment that was maintained on discharge).

The treatment she had when she came to the first visit included anti-hypertensive drugs, analgesics, risperidone 1 mg at night and lormetazepan 2 mg at night. Risperidone was prescribed on discharge after the acute confusional episode.

Regarding biographical background, the patient has a 76 year old healthy living sister. Her two other brothers died during the war when she was an adolescent. She married at 35 years of age and has two children (a 44 year old single son and a 39 year old married daughter) and two grandchildren. She has been widowed since four years ago. She worked as a dressmaker. She has good family support and her degree of functional activity is limited by the blindness and arthrosis.

No significant findings were found in the physical examination and, except for hypercholesterolemia, the laboratory analysis data were normal. Neuroimaging test, specifically a brain SPECT (fig. 1), were performed. The SPECT was performed after the intravenous administration of a dose of a bicasate dichlorhydrate compound, labeled with ^{99m}Tc. The examination was performed under conditions of sensorial rest, assessing the regional brain flow and the neuronal metabolic activity and transversal, coronal and sagittal sections were studied. The analysis showed a pattern of regular and symmetrical cortical perfusion, the cortical neuronal activity being considered within normality.

Finally, although no neuropsychological tests were performed due to lack of collaboration by the patient, these tests were described as normal in the geriatrics report.

With the clinical history and the data obtained in the complementary examinations, the initial diagnosis of organ hallucinosis, Charles Bonnet Syndrome, was made.

THERAPEUTIC PROCEDURE AND CLINICAL COURSE

In the first visit, the patient was informed about the organic character of her disorder and the neuroleptic treatment (risperidone 3 mg per day) was intensified, discon-

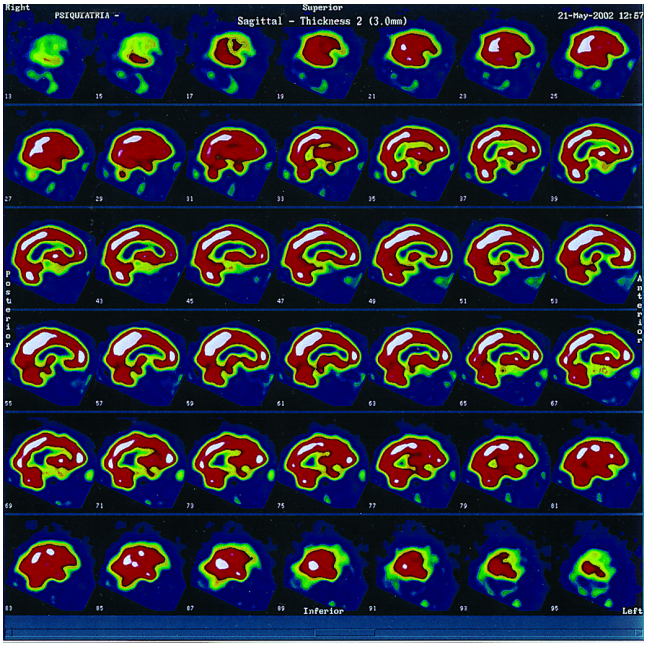

Fig. 1. Baseline brain SPECT (with ^{99}Tc). Saggital cuts. Regional brain flow pattern and neuronal metabolic activity are normal.

tinuing benzodiazepines. Furthermore, treatment was established with sertraline 50 mg in order to improve initiative and motivation for rehabilitation in the first moments. However, the patient did not come to the successive visits and returned the following month by referral from her geriatrist without having followed the pharmacological regimes. It seems that sertraline worsened her hallucinatory symptoms after three days of treatment, so that it was discontinued. In regards to the neuroleptics, 3 mg of risperidone during one month improved the quality of the hallucination (they are only one type now «the green corridor»), which was perceived by the patient as improvement. However, the hallucinations continued to be as frequent, intense, invalidating and egodistonic. Due to the appearance of extrapyramidal symptoms (tremor and rigidity) the neuroleptic treatment was slowly withdrawn and treatment was established with valpromide 600 mg/day. After discontinuing the antipsychotics, the extrapyramidal symptoms disappeared, a possible diagnosis of Parkinson's disease being discarded.

At one month of treatment with valpromide, the clinical picture improved significantly. The hallucinations disappeared in frequency and intensity («the hallucinations occur more slowly»), there being asymptomatic periods in which the patient reports that «she sees black». The patient is less anguished, her quality of sleep has also improved and her awareness of disease and trust in the treatment has increased progressively. No side effects have appeared.

Finally, given the course and therapeutic response to epileptic treatment, the diagnosis of Charles Bonnet Syndrome was verified.

DISCUSSION

Charles Bonnet, Swiss philosophist from the XVIII century (1720-1793) belonged to the naturistic school. He discovered the syndrome that carried the name of his elderly father, after a cataract operation. Charles himself was blind and deaf since his youth.

The definition of CBS coincides with the case presented: the appearance of complex visual hallucinations (polychromated, in movement) in a recently blind elderly person, without other psychopathology[1,2]. Classically, it has been considered a rare disorder, but in the light of recent studies (although not conclusive), it seems to be an underdiagnosed disease[2]. Only five cases have been published in our country[3-5]. Although few data are available, certain risk factors are pointed out (table 1), some of which are fulfilled by the patient presented: advanced age, blindness, recent operation, widowhood, with a previous episode of delirium.

In regards to the clinical symptoms, the picture presents hallucinations with the characteristics described in the CBS[6]. In general, complex hallucinations have been described that represent symmetric, geometric, distorted drawings, etc. (table 2), whose nature could correspond with the hallucination production mechanism. At present, there are different hypotheses to explain the neurobiology of complex hallucinations. One of them proposes that the different diseases that they give rise to (epilepsy, schizophrenia, Parkinson, etc.) could have a phenomenon of release or deafferentiation of the thalamocortical pathways in common (irritative phenomena such as epileptic focus, release or inhibition of the optic pathway by structural involvement, serotonin-dopamine involvement in Parkinson) [7,8].

According to the organic nature of the disorder, isolated cases have been described in relationship with the different medical pathologies such as temporal arteritis[9], HIV without findings of brain involvement in the neuroimaging tests[10], hormone replacement treatment with estrogens[11], treatment with erythropoietin in patients

TABLE 1. Risk factors in the Charles Bonnet syndrome

Physiological	*Neurological*	*Psychiatric*	*Iatrogenic*
Advanced age	*Optic pathway lesions*	*Delirium associated to surgery*	Anticholinergic
Altered sleep cycles	Epilepsy	Neurodegenerative diseases (i.e. Alzheimer, Lewy body dementia...)	L-Dopa
Sensorial deprivation	Migrane	Psychosis	
Photostimulation	Parkinson's disease	*Bereavement*	

In table 1, the risk factors proposed for the Charles Bonnet syndrome are shown. The characteristics which the patient have in the case presented are shown in italics. Source: Thomas E. Seminar on macular disease. University of Nottingham. The Royal College of Ophthalmologist Congress; 2001.

TABLE 2. Nature of the visual hallucinations in the Charles Bonnet syndrome

Category	*Description*	*%*
Simple, unformed	Flashes, lines, dots	82
Geometrical (tessellopsia: see mosaics)	Grids, lattice, fence, brickwork	74
Disembodied faces (prosopometamorphosia)	Grotesque, prominent, distorted eyes	56
Figures	Costumes, uniforms, hats	41
Branching forms (dendropsia)	Trees, branches, shrubs	41 27
Poliopia	Multiple copies of the same object in rows, columns	27
Perserveration	Real objects continue when looking away	27
Texts	Letters, notes	27
Particulate	Visual field covered with particles, dots	23
Vehicles	Cars, trucks, buses	18

Source: Thomas, E. Seminar on Macular disease. University of Nottingham. The Royal College of Ophthamologists Congress; 2001.

with renal dialysis[12] or leprosy[13]. The appearance of hallucinatory syndrome is especially frequent and is clinically indistinguible from CBS in early phases of Parkinson's disease[14] (in addition to the tendency of L-Dopa, fundamental treatment of Parkinson, to produce visual hallucinations). In the case presented, it was verified that the extrapyramidal symptoms that appeared in the patient were iatrogenic, secondary to the neuroleptic treatment. Finally, some authors propose CBS as a marker of early dementia[15,16]. However, there is great controversy regarding this proposal[15,16]. In the cases presented, the neuropsychological evaluation was normal, discarding, in the beginning, the existence of cognitive deterioration.

The diagnosis of the syndrome is basically clinical, there not being any complementary diagnostic test up to the present. The results found with structural neuroimaging are normal. Specifically, the SPECT technique does not present conclusive results: increase of flow in the lateral temporal cortex, striatum and thalamus[17]; decrease of the cerebral blood flow in the left occipital cortex[18]. Finally, the functional magnetic resonance techniques have demonstrated hyperactivation in the extrastriated ventral visual cortex in some cases that persisted when the hallucinatory activity stopped[19]. In the case described, the SPECT revealed that the cortical neuronal activity was within normality.

Regarding treatment, the use of typical and atypical neuroleptics does not seem to be effective[20]. In some isolated cases, it has been reported that treatment with cisapride[21] and melperone has been useful[20] and finally some studies point out the efficacy of the anti-epileptic agents carbamazepine[22] and valproic acid[23] at low doses. In our case, risperidone had to be discontinued due to lack of efficacy and, as pointed out in the literature, treatment with valproic acid was beneficial. In any case, the prognosis varies greatly. Cases have been described in which the hallucinations have subsided spontaneously and others in which they never do so, not even with pharmacotherapy. It is essential, in any case, to inform the patient of the «non-psychiatric» nature of their disorder as well as to transmit to them the need for pharmacological treatment due to possible therapeutic benefit.

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