

Charles Bonnet syndrome in hemianopia, following antero-mesial temporal lobectomy for drug-resistant epilepsy

Sara Contardi¹, Guido Rubboli¹, Marco Giulioni², Roberto Michelucci¹, Fabio Pizza¹, Elena Gardella¹, Federica Pinardi¹, Ilaria Bartolomei¹, Carlo Alberto Tassinari¹

¹ Neurology Unit, Department of Neurosciences, Bellaria Hospital, Bologna

² Neurosurgery Unit, Department of Neurosciences, Bellaria Hospital, Bologna, Italy

Received January 16, 2007; Accepted May 2, 2007

ABSTRACT – Charles Bonnet syndrome (CBS) is a disorder characterized by the occurrence of complex visual hallucinations in patients with acquired impairment of vision and without psychiatric disorders. In spite of the high incidence of visual field defects following antero-mesial temporal lobectomy for refractory temporal lobe epilepsy, reports of CBS in patients who underwent this surgical procedure are surprisingly rare. We describe a patient operated on for drug-resistant epilepsy. As a result of left antero-mesial temporal resection, she presented right homonymous hemianopia. A few days after surgery, she started complaining of visual hallucinations, such as static or moving “Lilliputian” human figures, or countryside scenes, restricted to the hemianopic field. The patient was fully aware of their fictitious nature. These disturbances disappeared progressively over a few weeks. The incidence of CBS associated with visual field defects following epilepsy surgery might be underestimated. Patients with post-surgical CBS should be reassured that it is not an epileptic phenomenon, and that it has a benign, self-limiting, course which does not usually require treatment.

Key words: Charles Bonnet syndrome, mesial temporal lobe epilepsy, hemianopsia, visual hallucination, epilepsy surgery, temporal lobectomy

The Charles Bonnet syndrome (CBS) is a disorder characterized by stereotyped, persistent or repetitive, complex visual hallucinations (such as moving objects or human beings, animals, vivid landscapes), not accompanied by psychotic symptoms, in patients with acquired impairment of vision due to ocular diseases or visual field abnormalities (Bonnet 1769, Schultz and

Melzack 1991, Ffytche 2005). Additional diagnostic criteria include full or partial retention of insight into the unreal nature of these phenomena, absence of hallucinations in other sensory modalities, exclusion of substance abuse or sleep disorders (Ffytche 2005). The visual disturbances characterizing CBS can occur for variable periods of time (from days to years),

Correspondence:

G. Rubboli
Neurology Unit,
Bellaria Hospital,
Via Altura, 3
40139 Bologna
Italy
Tel.:(+00 39) 051 6225738
Fax: (+00 39) 051 6225369
<guido.rubboli@ausl.bo.it>

Clinical observation presented at the 28th National Congress of the Italian League Against Epilepsy held in Bari on May, 18-21, 2005. This study has been partially supported by a grant from the Italian Ministry of University and Research (years 2004-2006) (Prof. C.A. Tassinari).

with the hallucinations changing both in frequency and complexity during this time (Schultz and Melzack 1991). Several factors may trigger or stop the hallucinations, presumably through a general arousal mechanism (Kolmel 1985, Schultz and Melzack 1991).

Visual field defects, mainly represented by upper quadrantanopias, are a relatively common consequence of anterior temporal lobe resection for refractory temporal epilepsy, with an incidence that can vary from more than 50% (Tecoma *et al.* 1993) to about 100% (Hughes *et al.* 1999). In spite of this high incidence of visual field defects, descriptions of CBS in patients who have undergone epilepsy surgery for drug-resistant temporal lobe epilepsy have been reported only rarely. To our knowledge, CBS has been reported in only four patients with upper quadrantanopia following amygdalohippocampectomy (Freiman *et al.* 2004, Tan and Sabel 2005). In this paper, we describe CBS in an additional patient presenting with right homonymous hemianopia resulting from left antero-mesial temporal lobectomy performed for the treatment of drug-resistant left temporal lobe epilepsy associated with a left temporo-mesial cavernous angioma.

Case report

A 49-year-old, right-handed female reported that, at the age of 35, she had started to experience brief, daily episodes of “*déjà-vu*” and “*jamais-vu*”, followed, after few weeks, by two generalized tonic-clonic seizures. Following this, she began to suffer from seizures characterized by an initial epigastric sensation, loss of contact, oralimentary automatisms, and repetitive upper limb movements; recovery of consciousness was accompanied by transitory aphasia lasting few minutes. Seizure frequency was two-three seizures per week. Interictal EEG displayed left temporal spikes. Brain MRI showed a cavernous angioma in the mesial portion of the left temporal lobe (*figure 1*).

She was admitted to the Department of Neurosciences at Bellaria Hospital in Bologna to undergo a presurgical evaluation that included prolonged video-EEG monitoring for seizure recording. She had never suffered from psychiatric disturbances or from disorders of vision; her visual field at admission was normal. She was on phenytoin, 400 mg/day; all previous medications had failed to satisfactorily control her seizures. The results of video-EEG monitoring indicated an epileptogenic area that extended from the left anterior lateral neocortex and temporal pole to the left mesio-temporal region; consequently, a left antero-mesial temporal lobectomy was performed (*figure 1*).

Immediately after surgery, she presented a transitory aphasic disorder lasting a few days; in addition, she started to complain of blurred vision in her right hemifield. A visual field examination demonstrated right hemianopia (*fig-*

ure 2). On the third day after surgery, she started to experience complex visual hallucinations in her right hemifield, with no impairment of consciousness: she usually referred to seeing coloured “Lilliputian” figures of women and children, either static or moving, but usually running in meadows or even lying in bed with her, or, occasionally, brightly coloured countryside scenes. She was concerned about the possible epileptic nature of these phenomena, although she acknowledged that they were completely different from the “*déjà vu*” episodes she had previously suffered from. These visual imageries were not accompanied by hallucinations involving other sensory modalities; in particular, no auditory sensations were referred to. She was aware of their fictitious nature and she could describe precisely what she was experiencing, without being frightened; in fact, she was sometimes amused by them. After a while, she noticed that she could make them disappear by closing her eyes. During these hallucinatory episodes, several EEG recordings failed to show any epileptiform abnormalities, but sporadic bursts of theta activity in the left anterior temporal leads, unrelated to the visual imageries, were observed. These phenomena were almost continuous in the first days, but then started to occur intermittently, more often in the evening, lasting from a few seconds to a few minutes. They became progressively less frequent; after two months, they disappeared spontaneously.

Discussion

Although anterior temporal lobectomy for drug-resistant temporal epilepsy is complicated, in a high percentage of cases (from more than 50% to nearly 100%) (Tecoma *et al.* 1993, Hughes *et al.* 1999), by post-surgical visual field impairment, reports of CBS in epileptic patients with visual field defects following this surgical procedure are actually rather rare. In the literature, only four patients have been described, by Freiman *et al.* (2004) and Tan and Sabel (2005): in all of them CBS was associated with upper quadrantanopia following amygdalohippocampectomy for the treatment of mesial temporal lobe epilepsy. In the two patients reported by Freiman *et al.* the visual hallucinations consisted of, in one case, moving geometrical objects, and of unfamiliar, moving human faces restricted to the specific region of the visual field defect, in the other; these phenomena appeared the day after surgery, vanishing after a few days. In our patient, the surgical procedure to remove the cavernoma and the epileptogenic area, caused a complete homonymous hemianopia, presumably due to more extensive damage of the optic radiations beyond Meyer's loop, the most anterior portion of the optic radiation, and possibly of the optic tract. Complex visual hallucinations in the defective hemifield appeared few days after surgery and persisted, although with decreasing frequency, for about two months. The character-

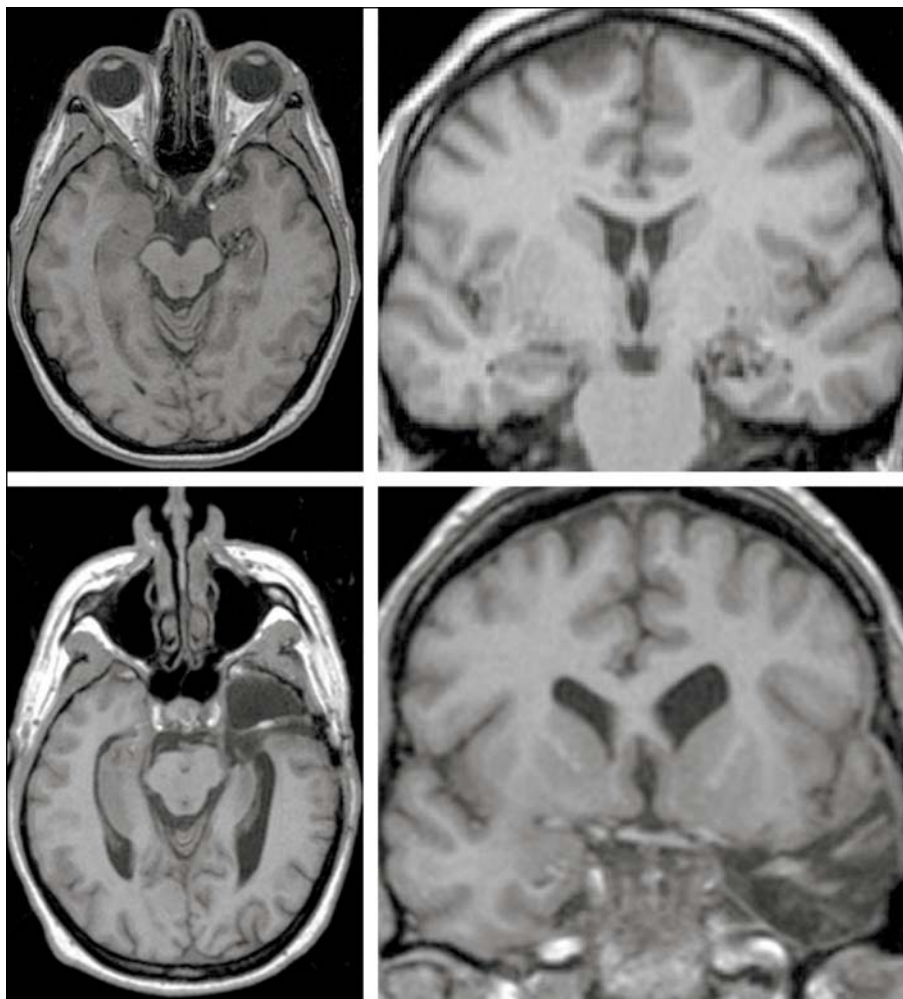


Figure 1. Upper panel: axial and coronal MRI images showing a cavernous angioma in the antero-mesial portion of the left temporal lobe. Lower panel: axial and coronal images showing the extent of the left antero-mesial temporal resection.

istics of the hallucinatory manifestations fulfilled the criteria for CBS (Schultz and Melzack 1991, Fytche 2005): in fact, they consisted of complex images, generally women and children, reduced in size (referred to as “Lilliputian” by the patient), brightly coloured and either static or moving; more rarely, panoramic views were perceived. The patient recognized their unreal nature, as was not frightened by them; non-visual hallucinations were never reported.

As reported elsewhere (Kolmel 1985), she could make them disappear, almost at will, by closing her eyes; the effectiveness of eye closing at stopping this kind of visual phenomenon has been suggested to depend upon an arousal mechanism, modulated by eye movements (Kolmel, 1985; Schultz and Melzack, 1991); an alternative explanation suggests that inhibitory impulses originating from the proprioceptors of the extraocular muscles might reach the occipital cortex via superior colliculi, interrupt-

ing visual imageries (Kolmel 1985). Of course, a possible epileptic nature was considered; however, epileptic visual hallucinations are usually brief and remarkably stereotypical, whereas in our patient, they could last several minutes and could differ in content from episode to episode. Moreover, visual phenomena in occipital epilepsy exhibit different characteristics, whereas when occurring in temporal lobe seizures, they are most often accompanied by other symptoms or signs (such as, for instance, confusion, mental slowing, clouding of consciousness) (Bien *et al.* 2000). Finally, in our patient, we never observed epileptiform abnormalities in any of the several EEGs performed during the hallucinatory episodes. Therefore, we concluded that an epileptic origin was extremely unlikely. In table 1, we summarize the main features of the visual hallucinations in epileptic seizures and in CBS.

Considering the high incidence of visual field deficits after antero-mesial temporal resection, it is somewhat

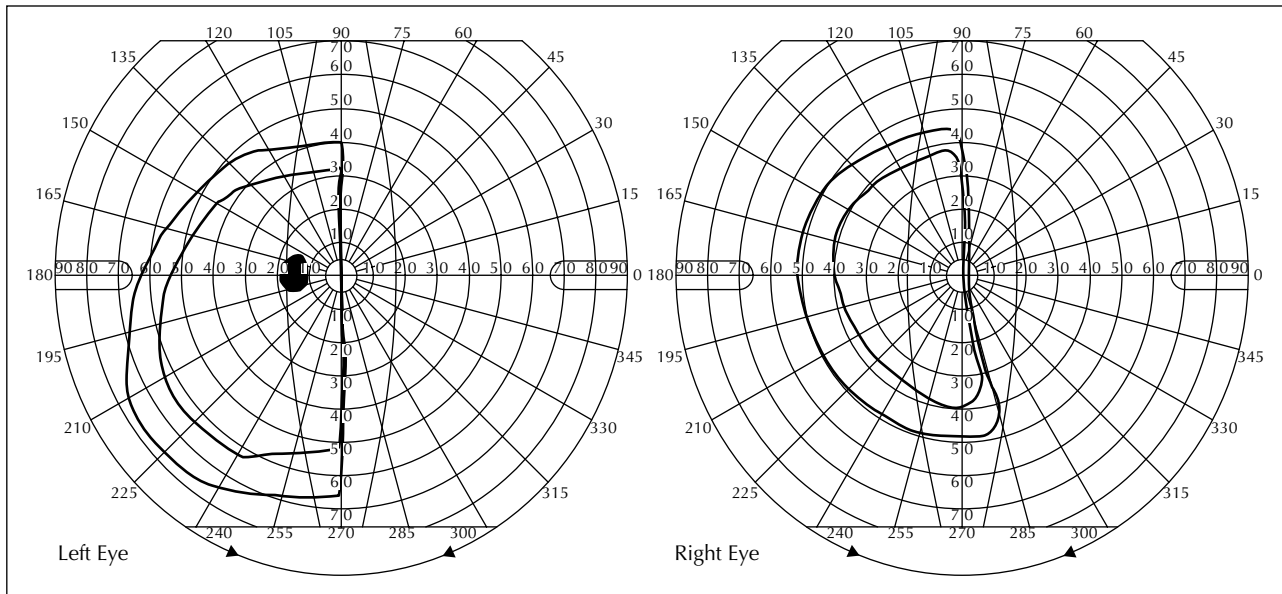


Figure 2. Post-operative visual field showing a right homonymous hemianopia.

surprising that CBS is reported so rarely in epileptic patients who underwent this surgical treatment. Indeed, it is possible that the incidence of CBS is underestimated because most patients suffering from this disturbance are reluctant to complain of it because of

the fear of being considered “mentally” disturbed (Freiman *et al.* 2004). In fact, in some instances it has been reported that the patients admitted that they were experiencing visual hallucinations only when specifically asked (Freiman *et al.* 2004).

Table 1. Main features of the visual hallucinations in epileptic seizures and in CBS.

	Visual hallucinations in epilepsy	Visual hallucinations in Charles Bonnet syndrome
<i>Duration</i>	seconds; rarely minutes	from seconds to several minutes
<i>Facilitating or triggering factors</i>	triggered by visual stimuli in occipital/occipito-temporal seizures	facilitated by darkness, or dim light; rarely by light.
<i>Voluntary modulation</i>	no	possible (by eye closure, or saccadic eye-movements)
<i>Visual field defects</i>	no	yes
<i>Contents</i>	- elementary hallucinations, illusions and visual loss in occipital/occipito-temporal seizures - elementary and complex hallucinations in temporal seizures	coloured complex hallucinations (moving objects, human beings, animals, vivid landscapes)
<i>Stereotyped</i>	yes	no
<i>Emotional valence</i>	unpleasant, pleasant, neutral	neutral, rarely pleasant
<i>Awareness of unreal nature</i>	sometimes	always
<i>Other symptoms or signs</i>	frequent (consistent with the spread of the epileptic discharge as in occipital, occipito-temporal, temporal seizures)	no
<i>Other sensory hallucinations</i>	possible	no
<i>Ictal EEG</i>	epileptiform abnormalities	normal

The pathophysiological mechanisms underlying CBS remain incompletely understood. Functional imaging studies in CBS patients have shown that hallucinations of colour, faces, textures and objects correlated with the time course of the fMRI signal in the ventral occipital lobe, with a correspondence between the nature of the hallucinatory phenomena and the specialization of the different visual areas (Ffytche *et al.* 1998). Regarding the processes causing the appearance of CBS in subjects with acquired abnormalities of visual function, Cogan (1973) postulated that missing inputs to the visual cortices (*i.e.*, a “deafferentation state”) might trigger the appearance of visual hallucinations by disinhibiting visual association areas through a “release” mechanism. Recently, Ffytche (2005) proposed the existence of two phenomenologic syndromes, one depending directly on visual system pathologies, the other on damage in the brainstem or in ascending neurotransmitter (particularly cholinergic) pathways. The characteristics of CBS in our patient, *i.e.* self-limiting hallucinations consisting of faces or unfamiliar figures or extended landscapes, lasting seconds or minutes, confined to a particular area of the visual field and progressively diminishing with time, fit with the features of the first syndrome proposed by Ffytche (2005), in which they result from excitation of visual cortical areas through local cortical pathology or secondary to functional/lesional visual deafferentation.

In conclusion, our report adds further evidence of the occurrence of CBS in visual field deficits following anterior temporal lobectomy for the treatment of drug-resistant temporal epilepsy. A proper diagnosis is necessary to reassure patients with post-surgical CBS that the manifestation is not an epileptic disorder and that it usually presents a benign, self-limiting course, not requiring treatment. □

Acknowledgements. We thank Mrs Clementina Giardini for her assistance in preparing the manuscript. This study has been partially supported by a grant from the Italian Ministry of University and Research (years 2004-2006).

References

- Bien CG, Benninger FO, Urbach H, *et al.* Localizing value of epileptic visual areas. *Brain* 2000; 123: 244-53.
- Bonnet C. *Essai Analytique sur les facultes de l’Ame* 2nd Ed, vol.11. Copenhagen and Geneva: Cl. Philibert, 1769; 176-178.
- Cogan DG. Visual hallucinations as release phenomena. *Albrecht Von Graefes Arch Klin Exp Ophthalmol* 1973; 188: 139-50.
- Ffytche DH. Visual hallucinations and the Charles Bonnet syndrome. *Curr Psych Rep* 2005; 7: 168-79.
- Ffytche DH, Howard RJ, Brammer MJ, Woodruff DP, Williams S. The anatomy of conscious vision: an fMRI study of visual hallucinations. *Nat Neurosci* 1998; 1: 738-42.
- Freiman TM, Surges R, Vougioukas VI, *et al.* Complex visual hallucinations (Charles Bonnet syndrome) in visual field defects following cerebral surgery. Report of four cases. *J Neurosurg* 2004; 101: 846-53.
- Hughes TS, Abou-Khalil B, Lavin PJM, *et al.* Visual field defects after temporal lobe resection. A prospective quantitative analysis. *Neurology* 1999; 53: 167-72.
- Kolmel H. Complex visual hallucinations in the hemianopic field. *J Neurol Neurosurg Psychiatry* 1985; 48: 29-38.
- Schultz G, Melzack R. The Charles Bonnet syndrome: “phantom visual images”. *Perception* 1991; 20: 809-25.
- Tan CSH, Sabel BA. Charles Bonnet syndrome in patients with visual field defects – are hallucinations good or bad? *J Neurosurg* 2005; 103: 193-5.
- Tecoma ES, Laxer KD, Barbaro NM, *et al.* Frequency and characteristics of visual field deficits after surgery for mesial temporal sclerosis. *Neurology* 1993; 43: 1235-8.