Abstract A 61-year-old patient suffered from Charles Bonnet syndrome (CBS) while his visual acuity declined, whereas CBS subsided after he became blind. These findings suggest that reduction of visual acuity (dynamic or acute impairment) has a greater impact on the onset of CBS than low visual acuity (static or chronic impairment) per se in some patients. They may also explain why patients with low visual acuity do not always suffer from CBS. Although further studies are required, the present case highlights the importance of the differentiation between lowering and low visual acuity in the etiology of CBS.

Key words Charles Bonnet syndrome · visual hallucinations · visual acuity · sensory deprivation

Introduction

Charles Bonnet syndrome (CBS) was named after the Swiss philosopher who first described the occurrence of colored, dynamic and highly organized visual hallucinations in his visually impaired, otherwise psychologically normal, grandfather Charles Lullin (Damas-Mora et al. 1982). Since then, the syndrome has been widely reported but estimates of prevalence vary considerably from around 1 % (Adachi 1996; Berrios and Brook 1984; Norton-Wilson and Munir 1987; Shiraishi et al. 2004) to about 10 % (Brown and Murphy 1992; Nesher et al. 2001; Olbrich et al. 1987; Teunisse et al. 1996). To date, all of the studies that have shown high prevalence of CBS (Brown and Murphy 1992; Nesher et al. 2001; Olbrich et al. 1987; Teunisse et al. 1996) involved ophthalmologic patients, whereas those studies that have found low prevalence (Adachi 1996; Berrios and Brook 1984; Norton-Wilson and Munir 1987) did not. Consequently, the presence of visual impairment may be at least partially associated with CBS although CBS may have heterogeneous causes (Terao 2002).

Reduced or absent visual-system stimulation leading to sensory deprivation and release phenomenon is the most widely accepted explanation for CBS (Fernandez et al. 1997). However, patients with visual impairment do not always suffer from CBS. Taking the close chronological relationship between sudden decrease in visual acuity and onset of visions, and the rarity of visual hallucinations in the totally blind (Damas-Mora et al. 1982) into consideration, there may be a possibility that lowering visual acuity (dynamic or acute impairment) has a greater impact on the onset of CBS than low visual acuity (static or chronic impairment) per se in some patients. In this report, a case of CBS is presented in support of the above possibility.

Case report

The patient is a 61-year-old male with severe visual impairment in his left eye (right visual acuity = 1.0, left visual acuity = 0.04) following an accidental eye injury at the age of 19. Although he was diagnosed with diabetes mellitus at the age of 40, he was not compliant with treatment. He had a wife and three children and lived with his wife and a younger son. From January 2001, the patient noticed that his right visual acuity had clearly decreased and reported repeated visual hallucinations involving rabbits being attacked by a tiger and the transformation...
of the attacked rabbit into a tiger. Subsequently, the patient's right visual acuity further decreased (50 cm hand moving) and the frequency of visual hallucinations increased. In August 2001, he was seen by an ophthalmologist and diagnosed with diabetic retinopathy. After this time, the patient reported seeing visions such as a daimyo gyoretsu (a feudal lord's procession in the Japanese Edo era). Following retinal photocoagulation therapy that did not affect the visual acuity, he was admitted to our university hospital in September 2001. At the interview by a psychiatrist (Y.S.), the patient was fully oriented without memory disturbance. He had no other psychiatric symptoms or a past history of mental illness. He retained full insight to his visual hallucinations and confessed that he had concealed his experience of visual hallucinations because he did not want to be regarded as a mad person. He was neurologically normal and the findings of electroencephalography showed normal background activities without any paroxysmal patterns, and magnetic resonance imaging of the brain was also normal.

On admission to our ophthalmological ward, visual acuity was 0.02 (corrected, 0.4) in the left eye and 50 cm hand moving in the right eye. In October 2001, the patient underwent a vitrectomy and endophotocoagulation in the right eye, which improved his right visual acuity, and he was discharged. Following this, his visual hallucinations vanished with the exception of one vision, consisting of a landscape, which occurred just after the operation. Subsequently, right visual acuity improved to 0.6 and thereafter he experienced no further visual hallucinations. However, two months later, the patient suffered from right retinal detachment, and visions of a landscape and those of small rabbits and a tiger recurred. In December 2001, he underwent two operations to correct the retinal detachment, but unfortunately he became blind in his right eye. Thereafter, the patient reported no visions.

Discussion

There have been several diagnostic criteria for CBS (Damas-Mora et al. 1982; Gold and Rabins 1989; Podoll et al. 1989, 1990; Ball 1991; Teunisse et al. 1995, 1996). Most of these require the presence of complex visual hallucinations with insight and the present case meets these criteria. However, none of these criteria necessitates the presence of optic pathology, and visual impairment is not necessary for diagnosing CBS. Nonetheless, previous reports (Teunisse et al., 1995, 1996; Podoll et al., 1989) and the present case suggest the association between visual impairment and CBS in some patients.

In this case, the patient's left visual acuity was continuously low for 42 years whereas right visual acuity decreased from 1.0 to 50 cm hand moving in the space of approximately 8 months. In parallel with the decline in right visual acuity, the patient began to experience visual hallucinations. Following the operation in October 2001, right visual acuity improved to 0.6 and the visual hallucinations subsided for 2 months. However, in parallel with the drop of right visual acuity caused by retinal detachment, the visual hallucinations returned once again. Thereafter, the patient's right eye became blind and he did not experience further visual hallucinations.

It is significant that this patient suffered from complex visual hallucinations while his right visual acuity was in decline. Conversely, he did not experience visual hallucinations while right visual acuity was static. In particular, it is noteworthy that no visual hallucinations were reported after losing sight in his right eye. These findings suggest that lowering but not low visual acuity brought about CBS in this patient. This supports the above-mentioned possibility that reduction in visual acuity (dynamic or acute impairment) has a greater impact on the onset of CBS than low visual acuity (static or chronic impairment) in some patients. Also, this possibility is supported by the fact that CBS more often occurs in the evening or in situations with inadequate lighting conditions (Teunisse et al. 1996). Interestingly, this possibility may explain why patients with low visual acuity do not always suffer from CBS. Probably, a process of adaptation may gradually compensate for sensory deprivation in static or chronic visual impairment, and thereby visual hallucinations are eliminated. This is also supported by the fact that the onset of the hallucinations was abrupt but thereafter they became less prominent as the patient's sight further deteriorated (White 1980).

With regard to other risk factors of CBS, Podoll et al. (1989) and Teunisse et al. (1999) pointed out the importance of loneliness as a sociodemographic risk factor for CBS. In the present patient, however, it seems unlikely because he lived with his wife and a younger son.

In conclusion, the present case suggests that lowering but not low visual acuity may induce visual hallucinations in some patients, and that the differentiation between lowering and low visual acuity is important in the etiology of CBS.

References