Visual Hallucinations in Giant Cell Arteritis: Association with Visual Loss

GIDEON NESHER, RONIT NESHER, YAACOV ROZENMAN, and MOSHE SONNENBLICK

ABSTRACT. ***Objective.*** To evaluate the frequency and characteristics of visual hallucinations (VH) in patients with giant cell arteritis (GCA) and to determine their relationship to other visual phenomena.

Methods. This prospective study included 31 consecutive patients with GCA. All were asked whether they had experienced recent visual phenomena. Patients with visual symptoms underwent a comprehensive ophthalmologic examination. When unusual visual phenomena were reported, patients were asked to describe their nature, duration, and frequency of occurrence.

Results. Visual symptoms occurred in 6 patients: permanent visual loss in 5 and amaurosis fugax in one. In 4 of the 5 patients with permanent visual loss, it was preceded by intermittent VH over a period of 1-10 days. Patients were aware of the unreal nature of the visions. Hallucinations disappeared within 2 weeks, but in one patient, recurred 6 months later in association with further visual deterioration.

Conclusion. The occurrence of visual hallucinations in patients with GCA-associated visual loss is more common than previously appreciated. As hallucinations preceded permanent loss of vision, this phenomenon may serve as a harbinger of imminent visual loss. (J Rheumatol 2001;28:2046–48)

Key Indexing Terms:
HALLUCINATIONS ISCHEMIC OPTIC NEUROPATHY TEMPORAL ARTERITIS

Sudden visual loss is the most dreaded ophthalmic complication of giant cell arteritis (GCA)[1-2]. It was considered to have no warning signs, but a recent study reported that in 65% of the cases the acute blindness was preceded by premonitory symptoms such as blurry vision or amaurosis fugax[3].

Hallucinations are spontaneous, unwilled sense perceptions, experienced as arising outside the self, for which there are no external causes. They differ from illusions, which are defined as misinterpretation of actual sensory experiences. Visual hallucinations (VH) may occur in the context of a psychiatric illness, but may also be experienced by sane individuals with irritative brain lesions, or with the use of certain drugs[4]. They have also been described in elderly individuals, mostly in association with decreased visual acuity, and in such cases have been termed Charles Bonnet syndrome[5-8]. These patients are aware of the unreal nature of the visions, and describe them as vivid, pleasant, and superimposed on the actual visual environment. These VH are classified as complex (i.e., have shapes of certain figures such as flowers or animals) or simple (colored lines, flashes, etc.).

Reports of VH in GCA are rare: only 4 cases have been described in detail in the English literature[9-11]: all were associated with visual loss. One of these patients was described by us previously[10]. Since this patient was initially quite reluctant to disclose her experience, fearing she would be labeled as insane, we assumed that VH may be underdiagnosed in GCA. Therefore we prospectively studied patients newly diagnosed with GCA for the occurrence of VH.

MATERIALS AND METHODS

Patients. Between 1992 and 1998, all patients diagnosed with GCA by the authors according to the criteria of the American College of Rheumatology[12] were asked upon presentation, and at each followup visit thereafter, whether they experienced any visual phenomena. Patients who reported visual symptoms underwent a detailed ophthalmologic examination. Patients reporting unusual visual experiences were asked to describe their nature, duration and frequency. They underwent psychiatric and neurological evaluations. Following the diagnosis of GCA, all patients were treated with steroids, and followed at 1-2 month intervals for 2-8 years.

RESULTS

Thirty-one patients, 19 women and 12 men, were diagnosed with GCA; 26 were biopsy positive. Six presented with visual symptoms: amaurosis fugax in one case, and irreversible visual loss in 5. The characteristics of these 5 patients are shown in Table 1.

From the Rheumatology Service, Departments of Ophthalmology and Geriatric Medicine, Shaare-Zedek Medical Center, and the Hebrew University Medical School, Jerusalem, the Department of Ophthalmology, Sapir Medical Center, Kfar-Saba, and the Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel.

G. Nesher, MD, Adjunct Associate Professor, St. Louis University School of Medicine, and Senior Lecturer, The Hebrew University Medical School, and Rheumatology Service, Shaare-Zedek Medical Center; R. Nesher, MD, Lecturer, Sackler Faculty of Medicine, Tel Aviv University, Department of Ophthalmology, Sapir Medical Center; Y. Rozenman, MD, Department of Ophthalmology; M. Sonnenblick, MD, Associate Clinical Professor, The Hebrew University Medical School, Department of Geriatric Medicine, Shaare-Zedek Medical Center.

Address reprint requests to Dr. G. Nesher, Rheumatology Service, Shaare-Zedek Medical Center, P.O. Box 3235, Jerusalem 91031, Israel.

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Table 1. Features of the 5 patients with GCA experiencing irreversible visual loss.

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age, gender	82, F	75, M	73, F	71, F	78, F
Other GCA symptoms	HA, PMR	HA, PMR	HA, JC	AION in fellow eye	HA, JC
ESR, mm/h	135	102	76	60	104
Hallucination	Carriage and horses, flowers	Cats and mice	Flowers in vases	Colorful rays	None
VH to VL (days)	3	10	1	2	
Vision, eye pathology	6/60, AION	HM, CRAO	HM, AION	HM, AION	6/60, AION
Fellow eye vision	6/24 (Cataract)	6/60 (Amblyopia)	6/18 (Cataract)	6/60 (AION)	6/9

AION: anterior ischemic optic neuropathy; CRAO: central retinal artery occlusion; ESR: erythrocyte sedimentation rate (normal < 40); HA: headache, HM: hand movement; JC: jaw claudication: PMR: polymyalgia rheumatica; VH to VL: duration of visual hallucinations prior to visual loss.

In 4 of the 5 patients with permanent visual loss, the event was preceded by 1-10 days of VH. The VH recurred 1-4 times a day, lasting less than a minute, but did not occur in clusters. Patients did not experience migraine-type headaches, dizziness or nausea at the time of VH. Three patients had one repeatable VH. Patient 1 had 2 different sights: one form of VH prior to her visual loss (horses pulling a fancy carriage), and a different form of VH (a field with flowers) 6 months later, in association with GCA activation and further visual deterioration in the other eye. Three patients described complex VH, and one (Patient 4) had simple VH. The figures and objects were moving. Patients were alone at the time of VH, which occurred during daytime while they were awake and had their eyes open. All patients were certain they did not misinterpret a real image. All were aware of the unreal nature of the sights, but 3 reported them only after being specifically questioned about them. Only Patient 2 voluntarily described the visions to his family physician 2 days prior to the occurrence of visual loss, but was referred to a psychiatrist.

All 4 patients had decreased visual acuity in the fellow eye also, due to various reasons (Table 1). Interestingly, Patient 5, who had visual loss but no VH, had near-normal vision in the other eye. The pattern of visual loss in the 4 patients with VH varied: 2 patients noted gradual deterioration of vision over a period of 3-4 days, and in the other 2 visual loss was sudden, upon waking in the morning. None had auditory or olfactory hallucinations, psychiatric or personality disorders, or had been taking medications with hallucinatory potential. Fluorescein angiography showed abnormal choroidal filling in their eyes. Brain computerized tomographic scans showed no space occupying lesions or infarctions.

Hallucinations disappeared within 2 weeks of starting steroid therapy. In Patient 1, VH recurred 6 months later during tapering of the steroid dose, in association with reactivation of the GCA and visual deterioration in the fellow eye. The GCA activation, visual deterioration and hallucinations all responded to an increase in the steroid dose.

DISCUSSION

Our data suggest that the occurrence of VH in patients with GCA-associated visual loss is more common than previously appreciated. The pathogenesis in unknown, but thought to be related to visual deprivation: a reaction of the visual cortex to lack of visual stimulation, resulting in release phenomena in the form of VH [6,13].

The occurrence of VH in these patients may be consistent with such a mechanism. All 4 patients had coexistent decreased visual acuity in the fellow eye. Under those circumstances, even a slight reduction in visual acuity in the affected eye, perceived by the patient as blurry vision, can still affect the visual cortex and be sufficient to produce release hallucinations preceding the visual loss[4].

Indeed, mild visual deterioration, described by patients as blurry vision, has been reported in almost a third of GCA patients prior to acute irreversible visual loss[3]. Such blurred vision was also noted by 2 of our patients. The mechanism of this slight deterioration of vision prior to visual loss is unclear. It has been reported that choroidal ischemia is a common finding in patients with GCA developing anterior ischemic optic neuropathy or central retinal artery occlusion, where it probably precedes the irreversible visual loss[1,2,14]. Abnormal filling of the choroidal vessels was also documented in our patients. Choroidal ischemia affects the outer part of the retina, and may lead to some degree of visual deterioration[15] preceding the visual loss. Such visual deterioration may trigger the phenomenon of VH prior to the loss of vision. VH ceased after several days in all patients. This was possibly due to re-conditioning of the visual perception in these cases.

Ischemic brain damage due to cerebral vasculitis may also result in the development of VH. However, there were neither clinical nor radiological signs of cerebro-vascular accidents, and VH were not reported by any of the 25 patients with GCA but without visual loss. A visual deprivation mechanism seems a more likely explanation of VH in these cases.

VH was the earliest visual symptom in 4 of the 5 patients

with permanent visual loss, preceding the loss of vision by several days. Thus, if reported when they first occur, these visual phenomena could serve as harbingers of imminent visual loss in patients with established GCA (as in Patients 1 and 4), or in patients in whom GCA was undiagnosed previously. Physicians treating patients with GCA should be aware of the various premonitory symptoms of visual loss, since early diagnosis and treatment may save their vision.

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