VISUAL SNOW SYNDROME IN A PEDIATRIC CASE

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INTRODUCTION

Visual snow (VS) is a rare condition that is characterized by continuous dynamically flickering dots in the entire visual field that imitate the 'static' or 'snow' of an analogue television set that is not connected to the antenna [1]. VS was first described in 3 of 10 migraineurs patients who presented with a spectrum of positive visual symptoms [2]. The symptoms of VS can persist for many years. Although VS might be expressed in patients with migraine as visual aura, persistent VS has been accepted as a distinct clinical entity and termed as visual snow syndrome (VSS) independently from migraine. Schankin et al. proposed that the criteria for diagnosis of VSS consisted of visual snow as the main criterion, with some additional criteria [3]. A few cases with childhood VSS have been described in literature [4-6]. Herein, the case of a teenager was presented to emphasize the importance of differential diagnosis in persistent positive visual phenomena.

CASE REPORT

A 17-year-old male patient was referred to our clinic with the complaint of atypical visual distortions in both eyes since the age of 16. He reported seeing tiny flickering dots all around the visual field of both of his eyes, which resembled migraine aura without any pattern of headache. His visual symptoms were constantly bilateral, never unilateral, and they never improved at any time during the day. Additionally, he described persistent photophobia. A complete ophthalmological evaluation, including fundoscopy and visual field testing, together with spectral domain optic coherence tomography (SD-OCT) was performed. His best corrected visual acuity was 20/20 (Snellen chart) in both eyes. His intraocular pressures were within normal limits. His visual field analyses were normal in both eyes, with normal ocular motility examination, normal light reflexes, and normal color vision tests (Ishihara). His fundus evaluation was normal, except for a limited area of lattice degeneration on his left eye. Although the sensation of photopsia, which is observed in patients with retinal degenerations due to vitreo-retinal tractions, is seen unilaterally, it was aimed to exclude any possible effect of a subclinical vitreoretinal traction; hence, argon laser photocoagulation was performed around the area of this degenerative area in his left eye. No clinical improvement was observed after argon laser photocoagulation and his complaints were persistent throughout the following days of treatment. His SD-OCT images of the macula and optic nerve (retinal nerve fiber layer) were analyzed and found to be normal. The atypical visual distortions he defined were thought to be in the form of illusionary palinopsia and positive light phenomena. No history of migraine with aura was determined in his family history and they were found to

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be free of any complaints, including periocular pain or headache. Since he experienced tinnitus in both ears, an otoscopic examination was performed, in addition to an audiological evaluation and vestibular tests. His audiogram was normal (right 5/5 dB, left 9/5 dB). No spontaneous nystagmus was seen. His Romberg test was negative. On video-nystagmography, his positional tests were negative (Dix-Hallpike and Roll maneuvers). His cerebellar test was normal. He had minimal low amplitude nystagmus on his gaze-evoked tests on both sides. Additional blood tests and electroencephalography were normal. Cranial magnetic resonance imaging (T1, T2, fluid attenuated inversion recovery, and diffusion-weighted images) showed no specific findings. After the exclusion of other possible etiological factors through physical examination and laboratory tests, lamotrigine was prescribed with the diagnosis of VSS.

DISCUSSION

VSS is a benign neurological condition that results in persistent and disabling visual disturbances that are distinctive from migraine visual aura. VSS is a diagnosis of exclusion; therefore, it should be rule out ophthalmological problems (retinal and vitreous diseases, retinal detachment), and the use of drugs resulting in similar symptoms. Due to the high comorbidity of migraine in VSS, the differential diagnosis of visual symptoms from migraneous aura is also necessary[1,7].

Due to the different clinical features when compared to migraine aura and no response to migraine therapy, VSS has been accepted as a separate entity. Schankin et al. first proposed the diagnostic criteria of VSS [3]. As the result of newly reported cases in which the proposed criteria were not met, the criteria were modified by replacing enhanced entoptic phenomena with other persistent positive phenomena (Table I)[1,7]. Although VSS is regarded as a distinct entity, patients with VSS are reported to have associated nonvisual symptoms, including comorbid migraine (58%), tinnitus, impaired concentration, depression, balance disorder, and tremors [1]. In the case herein, tinnitus was experienced as a complaint associated to the VS symptoms. According to his medical history and laboratory including ophthalmological evaluations, the diagnosis of the patient was compatible with VSS.

VSS mainly affects young patients, and equally affects men and women. However, the prevalence of VSS is unknown [5]. Pediatric cases have rarely been reported. Simpson et al. reported VS in a 12-year-old girl who was suffering from migraine. Persistent white, bright, jagged spots and black and white flashes with sparkles and dots were described in the case as the symptoms of VS. They did not observe any response in the visual symptoms to drug treatment [4]. Santos-Bueso et al. reported an 11-year-old girl with the complaint of continuous vision of white dots in both eyes. After excluding the neurological and ophthalmological etiologies, she was diagnosed with VS [5]. Bruen et al. presented a 14-year-old girl who was experiencing a constant visual acuity of 'falling rain' with flickering white lights in both eyes. However, they accepted the symptoms as prolonged migraine visual aura instead of VSS [6]. Several drugs used for acute and prophylactic treatment of migraine have been used for the treatment of symptoms of VSS because of the thought that they share a similar pathophysiological background. However, little or no success was often observed in the

Table I. Proposed criteria for visual snow syndrome[7]

1. Visual Snow lasting longer than 3 months: dynamic, continuous, tiny dots in the entire visual field
2. Presence of at least two additional visual symptoms from the following four categories:
   a. Palinopsia
   b. Photophobia
   c. Nyctalopia
   d. Other persistent positive visual phenomena including (but not limited to): enhanced entoptic phenomena (excessive floaters or blue field entoptic phenomenon), kaleidoscope-type colors with eyes open or closed, spontaneous photopsias
3. Symptoms are not consistent with typical migraine visual aura
4. Symptoms are not better explained by another disorder

improvement of visual symptoms [1,7]. There have been few cases of patients with partial attenuation of VS symptoms with lamotrigine. Unal-Cevik et al. reported VS symptoms in a 25-year-old woman with a 10-year history of migraine with aura. They chose lamotrigine for the treatment of the VS based on reported cases with persistent migrainous visual phenomena responsive to lamotrigine. With the lamotrigine treatment, the patient reported no longer complained of visual snow [8]. In a retrospective case series consisting of 47 patients with VSS, van Dongen et al. evaluated the response to treatment. They most frequently prescribed lamotrigine (n: 26) and observed the partial remission of symptoms in 5/26 patients (19.2%). Due to adverse events, including allergic reactions and excessive daytime sleepiness, only 4 patients continued the lamotrigine treatment. Other treatments (valproate, topiramate, acetazolamide, and flunarizine) have not led to any improvement, except in 1 of 4 patients receiving topiramate [9]. Evans and Aurora reported a case of VS with partial improvement using topiramate [10]. Bou Ghannam et al recommended lamotrigine, acetazolamide or verapamil as first-line treatment options [11]. As a result, evidence for the pharmacological treatment of VS is currently insufficient (Class IV evidence).

In conclusion, VSS must be kept in mind in the differential diagnostic evaluations of patients with persistent positive visual symptoms, even though it is not a commonly known clinical entity. A detailed medical history and laboratory tests (ophthalmological and neurological evaluations) will prevent the condition from being either misdiagnosed as psychological or diagnosed as migraine with aura, and are secondary to drug use.

The authors declare no conflict of interest

Informed consent was obtained by the parents of patient

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