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SUNCT headaches after ipsilateral ophthalmic-distribution zoster

Maria A. Nagel^a, Ted M. Burns^c, and Don Gilden^{a,b,*}

^a Department of Neurology, University of Colorado School of Medicine, Aurora, CO, USA

^b Department of Immunology and Microbiology, University of Colorado School of Medicine, Aurora, CO, USA

^c Department of Neurology, University of Virginia, Charlottesville, VA, USA

Abstract

Nine days after left ophthalmic-distribution zoster, a 47-year-old man developed SUNCT headaches (short-lasting unilateral neuralgiform headache with conjunctival injection and tearing). In contrast to two prior cases of SUNCT that developed after varicella zoster virus (VZV) meningoencephalitis without rash, this case describes an association of SUNCT with overt zoster, thus adding to the spectrum of headache and facial pain syndromes caused by VZV reactivation.

Keywords

SUNCT; VZV; Headaches

1. Introduction

Trigeminal autonomic cephalgias (TACs) are characterized by unilateral trigeminal-distribution head pain and ipsilateral autonomic symptoms. TACs include cluster headaches, paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms other than both conjunctival injection and tearing (SUNA). TACs can be differentiated by the length and frequency of recurrence of the headaches, as well as type of autonomic features, and most likely represent a spectrum of the same disease process. The estimated incidence of SUNCT is 1.2/100,000 and the prevalence is 6.6/100,000 [1]. SUNCT occurs most often in the fourth or fifth decades and is more common in men [1]. The cause of SUNCT is unknown. Treatment includes intravenous lidocaine, lamotrigine, topiramate, gabapentin, carbamazepine, botulinum toxin,

* Corresponding author: Don Gilden, MD, Department of Neurology, 12700 E. 19th Avenue, Mail Stop B182, Aurora, CO 80045, USA. Tel.: 1-303-724-4326; fax: 303-724-4329. don.gilden@ucdenver.edu.

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Conflict of interest statement

All authors report no conflicts of interest.

nerve blocks, deep brain stimulation and greater occipital nerve stimulation, all targeted at symptomatic relief. Herein, we present a case of SUNCT following ipsilateral ophthalmic-distribution zoster, suggesting that VZV reactivation precipitates SUNCT.

2. Case report

A 47-year-old man developed left ophthalmic-distribution zoster treated the same day with oral valacyclovir, one gram 3 times daily for 7 days. Five days after zoster onset, pain resolved and rash was crusted. Nine days after zoster onset and two days after discontinuing valacyclovir, he experienced the first of 20-25 ipsilateral SUNCT headaches. Headaches were stereotypical: severe (10/10), lasting 15-30 seconds and always associated with left eye lacrimation and redness, occurring up to 3 times daily. Many headaches were precipitated by scratching or touching the left forehead. During attacks, even the brief pain was so severe that he had to discontinue current activities, e.g., driving and sitting at a hotel dinner table, until pain predictably resolved. Neurological examination was normal. Brain imaging was not performed. When he developed SUNCT, he was re-treated with valacyclovir, one gm tid for 10 days. By the fourth week, the SUNCT headaches occurred only twice weekly and then resolved completely. He has been headache-free for over 2 years.

One year before the onset of zoster and SUNCT, the patient was treated for squamous cell carcinoma of the left frontal sinus with X-irradiation for 4 weeks followed by frontal sinus surgery and chemotherapy with cisplatin, 5-FU and docetaxol for 5 months. Treatments were completed 4 months before the onset of zoster. Brain MRIs every 6 months to the present time have been negative. Past medical history was also remarkable for varicella (chickenpox) at 5 years of age.

3. Discussion

Herein, we present a novel case of left-sided SUNCT that developed 9 days after ipsilateral zoster and resolved within 4 weeks in association with antiviral therapy. Furthermore, unlike many cases of SUNCT which are refractory to treatment, the patient has been headache-free for 2 years since antiviral treatment. This case is unique in that it is the first association of SUNCT that developed after overt zoster rash, adding to the spectrum of headache and facial pain syndromes associated with VZV reactivation.

Two earlier cases of SUNCT were associated with VZV infection but without rash. The first was a 46-year-old woman who developed meningoencephalitis followed by right-sided SUNCT [2]. Neurological examination, CBC and head CT were normal. The CSF contained 40 leukocytes, 88% mononuclear, and 20 red cells; CSF protein and glucose were normal; PCR for VZV DNA and herpes simplex virus (HSV) was not performed. She was treated with intravenous acyclovir for 10 days for possible HSV meningoencephalitis and on day 2, she developed right trigeminal neuralgia. At day 3, MRI revealed two hyperintense lesions at cortical-white matter junctions bilaterally and mild enhancement of a right occipital lobe lesion; at day 5, she developed partial motor seizures. Additional studies revealed elevated serum anti-VZV IgM, but not anti-HSV IgM antibodies, supporting the diagnosis of VZV meningoencephalitis; VZV serology on CSF was not performed. Right trigeminal neuralgia

resolved 2 weeks after presentation, and the right-sided SUNCT gradually improved and resolved completely

by 6 months. In the second case, a 72-year-old man with diabetes and hypertension presented with acute fever and right temporal headache without autonomic features and an elevated C-reactive protein of 42.6 (normal 5-10 mg/L) [3]. A few days later, he developed right-sided SUNCT followed one week later by high fever, nausea and right facial nerve palsy. Serum contained anti-VZV IgM antibodies. The CSF contained 80 leukocytes, 100% mononuclear, and elevated CSF protein of 78 and elevated glucose of 113; PCR revealed amplifiable VZV DNA. He was then treated with intravenous acyclovir. Two weeks later, he developed right paramedian pontomesencephalic and left hippocampal ischemic lesions and a right ponto-bulbar meningeal thickening. Ten days later, he developed myocarditis and died of an arrhythmia. Finally, in a study of 43 patients with SUNCT, 13 (30%) were found to have other preceding diagnoses (stress, psychiatric conditions, herpes zoster, seizures or Bell's Palsy); however, among these 13, the number of patients with preceding zoster and details regarding the period from zoster to development of SUNCT were not provided [4].

After primary VZV infection typically manifesting as varicella, virus becomes latent in human trigeminal ganglionic neurons [5,6] in >90% of humans [7-9], as well as in other cranial nerve ganglia, including the geniculate ganglia, dorsal root ganglia and autonomic ganglia. Autonomic ganglia in which VZV becomes latent in the head and neck include the pterygopalatine (sphenopalatine) ganglion [10] which provides parasympathetic innervation to the ipsilateral lacrimal gland. Thus, reactivation of VZV from trigeminal ganglia and/or autonomic ganglia could readily account for unilateral headache and tearing seen in SUNCT.

Although our patient was treated with antiviral therapy, it is yet unknown whether treatment helped, since he still developed SUNCT after first-course treatment and retreatment did not alleviate headaches for another 4 weeks. Overall, the value of antivirals in cases of SUNCT associated with VZV reactivation awaits further investigation.

4. Conclusion

Unlike two prior cases of SUNCT that developed after VZV meningoencephalitis without rash, we report the first association of SUNCT headaches with overt zoster rash. This case adds to the spectrum of headache and facial pain syndromes associated with VZV reactivation.

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Abbreviations

SUNCT	short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing
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VZV	varicella zoster virus
HSV	herpes simplex virus

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HIGHLIGHTS

- SUNCT headaches developed after ipsilateral ophthalmic-distribution zoster
- SUNCT headaches may follow VZV reactivation without rash
- The value of antiviral treatment for SUNCT is yet unknown