

Fig. 2—Multifocal electroretinogram: a reduced central retinal function at the first presentation (March 2018) in (A) right eye and (B) left eye; normal multifocal electroretinogram at the last follow-up visit (August 2020) in the right eye (C) and borderline multifocal electroretinogram in the left eye (D).

steric properties of the alkyl group might influence the binding affinity of poppers to a currently unknown binding site within a cone photoreceptor, resulting in differing toxicity of various alkyl nitrites.

Rok Sega,
Mojca Urbancic

Eye Hospital, University Medical Centre Ljubljana, Ljubljana, Slovenia.

Correspondence to:

Mojca Urbancic, MD, PhD; mojca.urbancic@kclj.si.

- Davies AJ, Kelly SP, Naylor SG, Bhatt PR, Mathews JP, Sahni J, et al. Adverse ophthalmic reaction in poppers users: case series of poppers maculopathy. *Eye* 2012;26:1479–86.
- Davies AJ, Borschmann R, Kelly SP, Ramsey J, Ferris J, Winstock AR. The prevalence of visual symptoms in poppers users: a global survey. *BMJ Open Ophthalmol* 2017;1 1–1.
- Audo I, El Sanharawi M, Vignal-Clermont C, Villa A, Morin A, Conrath J, et al. Foveal damage in habitual poppers users. *Arch Ophthalmol* 2011;129:703–8.
- Rewbury R, Hughes E, Purbrick R, Prior S, Baron M. Poppers: legal highs with questionable contents? A case series of poppers maculopathy. *Br J Ophthalmol* 2017;101:1530–4.

References

- Sigell LT, Kapp FT, Fusaro GA, Nelson ED, Falck RS. Popping and snorting volatile nitrites: a current fad for getting high. *Am J Psychiatry* 1978;135:1216–8.

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Central retinal artery occlusion associated with Sweet syndrome



A 64-year-old Caucasian man presented with a 1-day history of acute painless vision loss in the right eye. On examination, his visual acuity was hand motions OD and 20/30 OS. His intraocular pressures were normal OU and he had a 3+ relative afferent pupillary defect OD. The anterior segment examination was unremarkable OU. Fundoscopic examination of the right eye revealed a cherry red spot (Fig. 1A). No vitreous cell, vasculitis, or chorioretinitis was noted OU. A fluorescein angiogram of the right eye showed an arterial filling line (Fig. 1B) with no leakage or staining in later frames. Optical coherence tomography of the right eye showed inner layer hyper-reflectivity indicative of acute ischemia (Fig. 1C). A bilateral carotid doppler ultrasound and computed tomography angiography of the head and neck was negative for stenotic disease. Given the patient's age and

elevated inflammatory markers, a temporal artery biopsy was done, which did not show evidence of giant cell arteritis.

Before presentation, the patient returned to Canada from a 1-month trip to Mexico where he developed upper respiratory tract symptoms. Following self-isolation for 14 days as a result of coronavirus disease (COVID-19) precautions, his symptoms resolved. Three weeks later, the patient developed a progressive, painful, diffuse, nodular rash to all 4 extremities (Fig. 1D), polyarthritides, fever, and difficulty ambulating. He was admitted to the general internal medicine ward where initial testing revealed neutrophilia, an elevated C-reactive protein, and strongly positive anti-cyclic citrullinated peptide titers (>500 U/mL). The patient underwent a skin punch biopsy of the right arm, which showed a dense neutrophilic infiltrate and confirmed a diagnosis of acute febrile neutrophilic dermatosis, also known as Sweet syndrome. A full body positron emission tomography

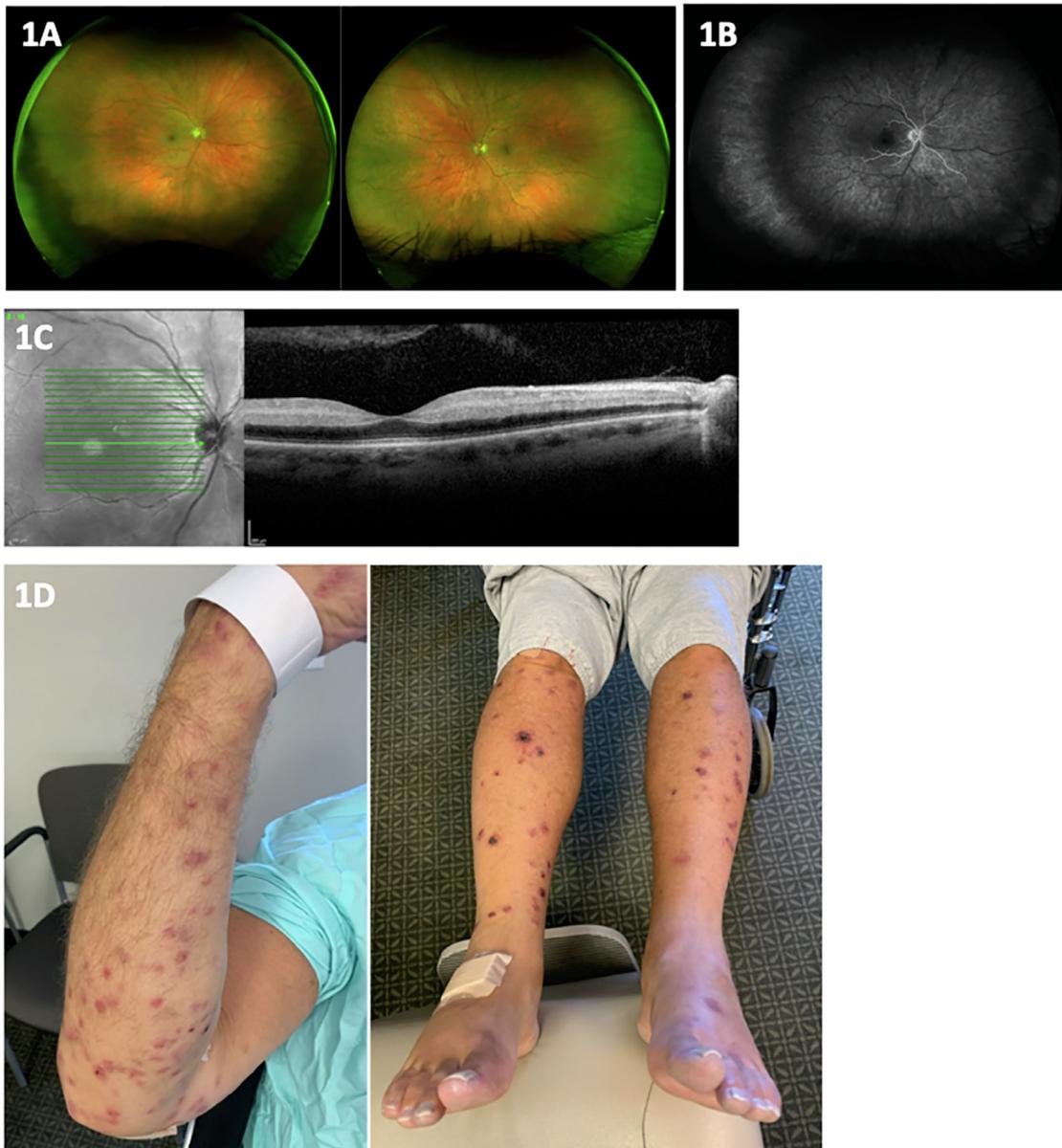


Fig. 1—(A) Color photograph of the right and left eye exhibiting a right cherry red spot. (B) Fluorescein angiogram of the right eye at 50 seconds demonstrating an arterial filling line. (C) Optical coherence tomography of the right eye demonstrating hyperreflectivity of the inner retina indicative of ischemia. (D) External photograph of nodular skin lesions on arms and legs.

scan was negative for malignancy. The rheumatology service was consulted, and the patient was treated with intravenous Solumedrol 1 g for 3 days, followed by 1 week of oral Prednisone 100 mg.

Discussion

Sweet syndrome is a dermatologic disorder first described by Dr. Robert Douglas Sweet in 1964. This disorder is generally characterized by tender cutaneous papules and nodules, pyrexia, elevated neutrophil count, and diffuse infiltration of mature neutrophils in the upper dermis on skin biopsy.¹ Sweet syndrome can be divided into classical/idiopathic,

malignancy-associated, and drug-induced. The diagnostic criteria for Sweet syndrome, originally proposed by Su et al.² and later modified by von den Driesch,³ are listed in Table 1. Both major criteria and 2 of 4 minor criteria must be present for the diagnosis of Sweet syndrome.

Ophthalmic involvement is thought to occur in about 33% of patients diagnosed with Sweet syndrome.³ Reported ocular manifestations include blepharitis, conjunctivitis, episcleritis, scleritis, peripheral ulcerative keratitis, iritis, panuveitis, glaucoma, choroiditis, retinal vasculitis, dacryoadenitis, and periorbital inflammation.¹ These inflammatory ophthalmic manifestations have been described as occurring at the same time or a few days after the onset of the tender

Table 1—Diagnostic criteria for Sweet syndrome

Major criteria	1) Abrupt onset of tender or painful cutaneous erythematous plaques or nodules, occasionally with pustules, vesicles, or bullae 2) Histopathologic evidence of dense neutrophilic infiltrate in the absence of leukocytoclastic vasculitis
Minor criteria	1) Fever >38°C 2) Preceded by a nonspecific upper respiratory tract or gastrointestinal infection or vaccination or associated with an inflammatory disorder or pregnancy or hematologic or other malignancy 3) Good response to treatment with systemic corticosteroids or potassium iodide 4) Three of 4 of the following abnormal laboratory values at the time of presentation: a. Erythrocyte sedimentation rate >20 mm/h b. Elevated C-reactive protein c. Leukocyte count >8000 d. White blood cell differential count >70% neutrophils

erythematous skin lesions.¹ The majority of ocular manifestations associated with Sweet syndrome are mild and carry a favorable prognosis. Associated cases of choroiditis and retinal vasculitis, however, can present a diagnostic challenge as these manifestations may resemble Behcet's disease, which require further investigation to differentiate between.^{4,5} In the present case, vision loss occurred 2 days after starting IV Solumedrol. Both a fluorescein angiogram and computed tomography angiography of the head and neck failed to identify active vasculitis in the retinal or larger vessels; however, given 2 days of high-dose systemic steroid treatment, the absence of vasculitis could have been a false negative. To the best of our knowledge, this was the first case report of a central retinal artery occlusion as an ocular manifestation of Sweet syndrome.

Although rare, this case highlights the importance of including Sweet syndrome in the differential diagnosis for patients presenting with severe vision loss and tender cutaneous erythematous lesions.

Helya Aghazadeh, David Sia, David Ehmann
University of Alberta, Edmonton, Alb.

Originally received Dec. 22, 2020. Accepted Dec. 31, 2020.

Correspondence to:

David Ehmann, MD: dehmann@ualberta.ca.

References

- Gottlieb CC, Mishra A, Belliveau D, Green P, Heathcote JG. Ocular involvement in acute febrile neutrophilic dermatosis (Sweet syndrome): new cases and review of the literature. *Surv Ophthalmol* 2008;53:219–26.
- Su WPD, Liu HNH. Diagnostic criteria for Sweet's syndrome. *Cutis* 1986;37:167–74.
- von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol* 1994;31:535–56.
- Salvador-Osuna C, Fernandez-Mosteirin N, Mayayo P, Delgado P, Giral M. Choroiditis as systemic manifestation of a Sweet's syndrome associated to myelodysplasia: a case report. *Haematologica* 2002;87:ECR07.
- Sato M, Kawamura T, Hase S, Katsumata S, Oshika T. A case of bilateral retinal vasculitis associated with Sweet syndrome. *Retina* 2005;25:800–2.

Footnotes and Disclosure

The authors do not have any conflicts of interest or proprietary interests to declare.