A Seemingly Innocuous Headache Heralding Onset of Rare Autoimmune Disorder.

Thomas Quinn DO
Lehigh Valley Health Network, Thomas.Quinn@lvhn.org

Noor Salam MD
Lehigh Valley Health Network, noor.salam@lvhn.org

Published In/Presented At

Quinn, T. Salam, N. (2017, October 28). *A Seemingly Innocuous Headache Heralding Onset of Rare Autoimmune Disorder*. Poster Presented at: PA-ACP Eastern Region Abstract and Doctors Dilemma Competition, Harrisburg, PA.

A Seemingly Innocuous Headache Heralding Onset of Rare Autoimmune Disorder

TA Quinn, DO, N Salam, MD

Department of Internal Medicine, Lehigh Valley Health Network, Allentown, Pennsylvania

INTRODUCTION

Headaches and migraines are commonly encountered symptoms in the outpatient and inpatient arenas.

Management revolves around ruling out alarm symptoms, and achieving symptomatic control first with over-the-counter medications, with additional options available depending on severity.

Unfortunately, these symptoms may rarely indicate a more precarious etiology.

CASE PRESENTATION

A 29-year-old Hispanic Female with history of episodic binge-drinking presented for well-visit. Casual review of symptoms revealed mild bitemporal headaches with wave-like intensity pattern.

- Tension headaches were diagnosed, with counseling to improve sleep hygiene and decrease alcohol intake

Returned one month later for worsened headache despite lifestyle changes. Now with associated photophobia, tearing, nausea, and new onset blurry vision OU.

- Described a central arc of distortion as if “looking through a pane class with a bend in the middle” from her left eye and “a ball of blurry vision” in the center of her right eye. When focusing on objects, she described “everything in my peripheral vision is vibrating or shaking.” She also described “clicking noise in left ear like soda fizzing”
- Exam significant for pain with extraocular movement, bilateral conjunctival injection and inability to perform ophthalmoscopy due to photophobia.
- She was diagnosed with status migrainosis, prescribed oral dexamethasone, sumatriptan, topiramate, and referred to ophthalmology.

Headache resolved with dexamethasone.

However, presented twice to Emergency Department for reoccurrence of above ophthalmic symptoms.

- ED prescribed topical antibiotics twice for presumed corneal abrasions

Presented to Ophthalmology. History obtained revealed alopecia with a bald patch on the right parietal scalp about size of a half dollar, vitiligo on forearm, oral ulcers, hair whitening, and joint pain.

- Exam significant for Va 20/200 OD, 20/400 OS, and OU findings of injection, keratic precipitates, 2+ anterior chamber (AC) cell/flare, multiple iris synechiae, 2+ vitreous cell, absent retinal tears/ detachment, linear areas of macular whitening with focal atrophy, and peripheral white dots 360º
- Fundus autofluoresence showed no serous detachment but ultrasound revealed choroidal thickening

Testing revealed negative CXR, ACE, ANA, RF, anti-DS DNA, HLA-B27, syphilis, toxoplasma, histoplasma, lyme, and QuantiFERON-TB gold.

Diagnosed with Vogt-Koyanagi-Harada syndrome. She was started on atropine and prednisolone acetate ophthalmic drops, and oral prednisone.

Va improved to 20/80 OD & 20/125 OS with persistent AC reaction OS>OD requiring increased prednisolone acetate, and sub-Tenon’s triamcinolone injections. Developed steroid-induced subcapsular cataracts OU.

Rheumatology started methotrexate, with subsequent improvement and ability to significantly taper oral steroids.

DISCUSSION

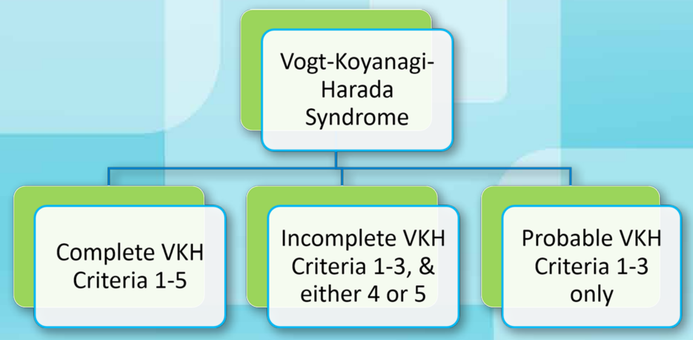
Vogt-Koyanagi-Harada (VKH) syndrome is an idiopathic multisystem autoimmune disease featuring inflammation of melanocyte containing tissues

HLA associations. Higher prevalence in Asian, Middle Eastern, Hispanic, and Native American populations

There are no confirmatory tests. Diagnosis is based on a constellation of clinical signs and symptoms

May be associated with other autoimmune disorders such as hypothyroidism, Hashimoto thyroiditis, DM, autoimmune polyglandular syndrome, Gullian-Barre syndrome, & IgA nephropathy

No associated mortality. Long term complications include vision loss secondary to cataracts, glaucoma, and choroidal neovascularization



1.	No history of penetrating ocular trauma or surgery preceding initial onset of uveitis
2.	No clinical or lab evidence suggestive of other ocular disease entities
3.	Bilateral ocular involvement
4.	Neurologic/auditory findings (one sufficient) • Mengingismus (malaise, fever, headache (not isolated), nausea, abdominaal pain, neck/back stiffness) • Tinnitus • Cerebrospinal fluid pleocytosis
5.	Integumentary findings (one sufficient) • Alopecia • Poliosis • Vitiligo

CONCLUSION

Our patient demonstrated all five of the diagnostic criteria for Complete VKH: neurological/auditory manifestations of headache & tinnitus; integumentary findings of alopecia, poliosis & vitiligo.

Recognition of this rare disorder is vital as prognosis is partly dependent on early aggressive control.

References:

1. Bowling, Brad (2015). Kanski’s Clinical Ophthalmology: A Systematic Approach. Saunders Ltd. 8th ed. p415-416
2. Walton, RC (April 15, 2016). Vogt-Koyanagi-Harada Disease. Medscape. Retrieved from http://reference.medscape.com/article/1229432-overview