A Seemingly Innocuous Headache Heralding Onset of Rare Autoimmune Disorder.

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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.

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 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 360°
- 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, Guillain-Barre syndrome, & IgA nephropathy

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

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: