CLINICAL VIGNETTE

The Eyes Have It: A 51-Year-Old Male with Vogt-Koyanagi-Harada Syndrome

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Case Report

A 51-year-old male with type 2 diabetes was admitted with vision changes, headache, and right ear pain. He reported progressive blurring of vision in his left eye one week prior to admission followed by blurring of vision in his right eye three days prior to admission. His headache started 2 weeks prior with a gradual onset associated with photophobia with response to non-steroidal anti-inflammatories. Right ear pain started 3 weeks prior to admission, without hearing loss or tinnitus. He was diagnosed with right otitis externa and started on ear drops. He also reported bilateral eyelid swelling. He denied any trauma, fevers, chills, neck stiffness, painful eye movements or double vision. He was seen by ophthalmology who noted painless vision loss with bilateral papilledema and sent him to the emergency room for additional evaluation.

On presentation to the emergency room, his exam was notable for decreased visual acuity, conjunctival injection, and mild periorbital edema. Neurologic exam was non-focal and without meningismus. Routine labs were unremarkable. Head CT scan showed no acute intracranial process but noted right external auditory canal thickening suggestive of otitis externa and maxillary mucosal thickening indicating a possible fungal infection. MRI brain showed no acute intracranial abnormalities. MRI orbit showed thickening of the posterior aspect of the bilateral globes. Due to concern for early meningitis, a lumbar puncture was performed, which revealed a normal opening pressure, a lymphocytic pleocytosis, and mildly elevated glucose and protein. His CSF testing was negative for bacterial, viral, and fungal infections and he was started on empiric antimicrobials.

Neurology, Infectious Disease, ENT, and Ophthalmology consulted. Tests for TB, sarcoid, VZV, HSV, leptospirosis, West Nile Virus, endemic fungi, and malignancy were negative. Based on imaging findings, a right ear culture was obtained, which revealed aspergillus. On re-examination, Ophthalmology noted he now had evidence of anterior segment inflammation and posterior synechiae, which, in the setting of lymphocytic pleocytosis, was consistent with Vogt-Koyanagi-Harada (VKH) syndrome. He was treated with high-dose prednisone, topical prednisolone, and topical cycloplegics with near-resolution of symptoms. Repeat orbital MRI following treatment showed resolution of the posterior globe bilateral thickening seen early in presentation. The aspergillus was felt to be unrelated and was treated with voriconazole.

Discussion

This 51-year-old male presented with symptoms concerning for meningitis and was ultimately diagnosed with VKH syndrome. Bacterial meningitis presents similarly to the often-self-limited aseptic meningitis. Aseptic meningitis can be related to viral, fungal or bacterial infections or may be drug-induced. In this patient with ear infection and type 2 diabetes, invasive fungal infections were initially considered. The lymphocytic pleocytosis broadened the differential to include potential noninfectious causes, including autoimmune and malignant etiologies.

VKH syndrome is a rare autoimmune disorder that affects pigmented tissue of the eyes, skin, ears, and nervous system. As with many autoimmune diseases, the etiology has not been fully identified. Although viruses including Cytomegalovirus and Epstein Barr virus are thought to play a role, some research suggests a genetic component.[1] Females and males are affected at similar rates, but the disorder is more prevalent in Asian, Hispanic, and Native American populations. Diagnostic criteria include no known ocular injury, as well as at least three of the following four criteria: (1) posterior uveitis; (2) neurologic signs including stiffness in the neck, tinnitus, CNS symptoms, or CSF pleocytosis; (3) skin findings including alopecia, vitiligo, and/or poliosis; and (4) bilateral iridocyclitis.[2] Additionally, MR can show choroidal thickening as in our patient.[3]

Presentation is variable, and clinical features have been divided into four phases consisting of the prodromal, acute uveitic, convalescent, and recurrent phases.[2] The prodromal stage is similar to a non-specific viral infection and lasts a few days. The acute uveitic phase starts within days of the prodromal phase and can last several weeks. In this phase, vision loss occurs in both eyes, often starting in one eye and progressing to the contralateral eye a few days later, similar to our patient. There is no preference for the right versus the left eye.[4] The convalescent phase presents months after the acute uveitic stage and consists of depigmentation including vitiligo, alopecia, and poliosis.[2] The recurrent phase, which can overlap with the convalescent phase, has been associated with rapid corticosteroid tapering with complications including cataracts, glaucoma, and choroidal neovascularization.[2] Our patient presented in the acute uveitic phase with blurry vision and a severe headache.

Prompt treatment is recommended and usually involves high dose systemic corticosteroids. Prednisone should be given at 1

to 1.5mg/kg per day or a pulse dose course of 1g methylprednisolone per day for 3 days followed by a slow taper over a minimum of six months. Immunosuppressive therapy should be considered in corticosteroid refractory cases. Some experts recommend immunosuppressants be considered as first-line therapy.[2,5] Dermatology, Ophthalmology, and Neurology can assist with management. Treatment can improve vision and hearing problems, but dermatologic changes are likely permanent.

Conclusion

Lymphocytic pleocytosis may not indicate viral meningitis, and a broad differential including autoimmune causes should be considered. Maintain a high index of suspicion for VKH when unexplained ocular, neurologic, and/or dermatologic findings are present. Consider prompt initiation of corticosteroid therapy as this improves the likelihood of symptom resolution.

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