“The Prognosis for Life and Vision is Good, for Hearing, Poor”*

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Case

A thirty-six year-old Caucasian woman with severe myopia presented to an outside emergency room with injected eyes, blurred vision, photophobia, and headache of about a week's duration.

Her exam demonstrated interstitial keratitis with extensive vascular ingrowth (figures 1 and 2). Syphilis was ruled out and she was started on antibiotic drops. Her symptoms initially improved but a few weeks later she developed worsening blurred vision on the right, acute onset of right-sided tinnitus and decreased hearing, vertigo, abdominal pain, and vomiting. She re-presented and was admitted to a medicine service.

Audiometry demonstrated mild sensorineural hearing loss on the right (figure 3). A normal magnetic resonance imaging scan of the brain ruled out multiple sclerosis and acoustic neuroma. Tests for herpes simplex virus and Lyme disease were negative.

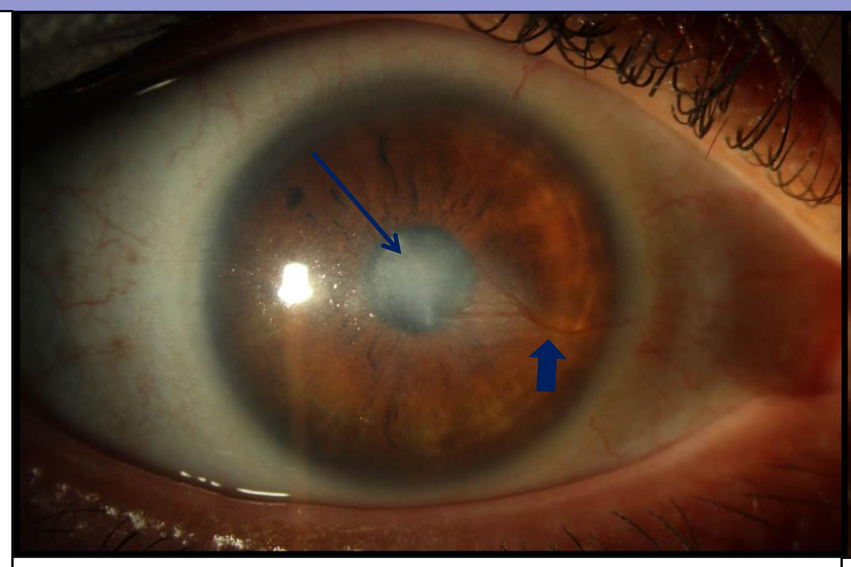

Figure 1
Right eye: Mild diffuse corneal stromal haze. 360 degrees of neovascularization. Multiple deep areas of mature neovascularization (thick arrow) extending to central corneal infiltrate (thin arrow).

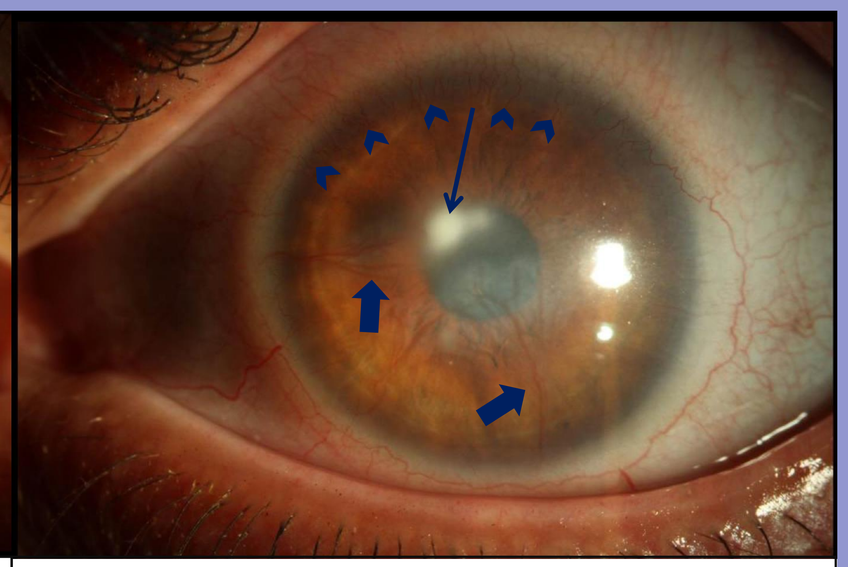

Figure 2
Left eye: Mild diffuse corneal stromal haze. 360 degrees of neovascularization (arrowheads) with multiple deep mature neovascularization extending to near central visual axis (thick arrow). Superonasal corneal infiltrate (thin arrow).

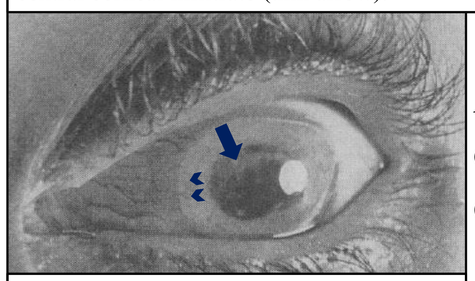

Figure 4
Sketch of the left eye of a patient in the original case series showing infiltrates (arrow) and vascularization (arrowhead).

Case (con’t)

After excluding viable alternatives the patient received a diagnosis of Cogan’s syndrome, an autoimmune condition which causes interstitial keratitis and vestibuloauditory symptoms.

Prednisone was started at 1 mg/kg. The plan was to repeat the audiometry to assess for response but the patient did not return.

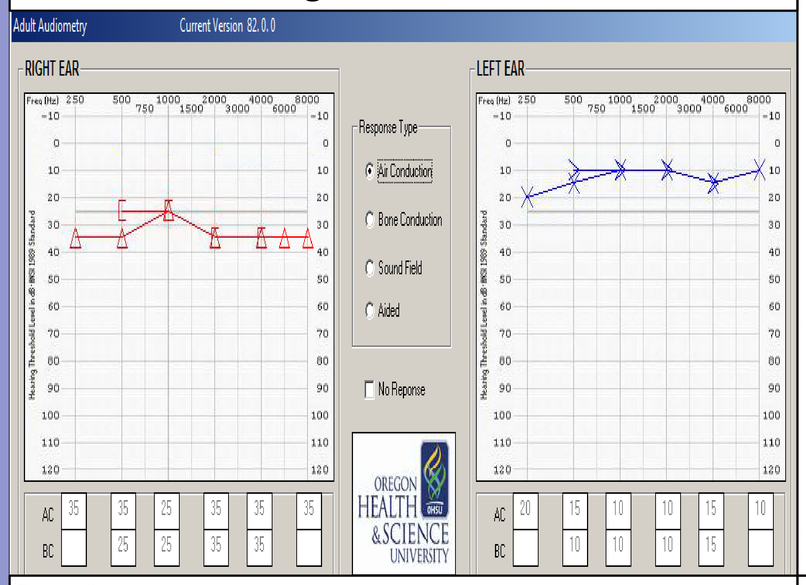

Figure 3
Right ear: Mild sensorineural hearing loss. Speech reception threshold was obtained at 25 dB HL.
Left ear: Normal hearing sensitivity. Speech reception threshold was obtained at 15 dB HL.

Discussion

Described in 1945, the initial cohort of patients with Cogan's syndrome had acute sensorineural hearing loss and non-syphilitic interstitial keratitis (figure 4), but the definition has expanded to include other types of inflammatory eye conditions. Typical presentation is with either hearing loss or visual symptoms but to meet diagnostic criteria patients must develop both within two years of onset. As in our case, Cogan’s syndrome typically affects Caucasians in their third decade.

The pathogenesis of Cogan’s syndrome is not well described. One study isolated autoantibodies from patient’s with Cogan’s syndrome specific for a peptide expressed in endothelial cells as well as in epithelial cells of the inner ear. This peptide is homologous to connexin 26, which is implicated in most cases of congenital deafness. Introducing these autoantibodies into a mouse model caused hearing loss and corneal inflammation.

Patients are usually treated empirically with steroids. Most patients improve initially and later relapse. Vestibuloauditory outcomes are poor and most patients develop severe hearing loss or deafness. In contrast, long-term visual defects are rare.

References

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