



INTERSTITIAL KERATITIS

Nonsyphilitic Interstitial Keratitis (Cogan's Syndrome)

- deep nonsyphilitic interstitial keratitis
- vestibuloauditory dysfunction
- life-threatening vasculitis in 10% of patients
- younger individuals.
- autoimmune reaction to a common autoantigen in the cornea and middle ear
- most common eye finding chronic, bilateral interstitial keratitis.
- begins with an acute onset of paralimbal injection associated with diffuse or sectoral, anterior stromal, subepithelial infiltrates 0.5-1.0mm in diameter
- Topical corticosteroid treatment at this point may reduce the viral-like infiltrates and inhibit the development of the classic late interstitial keratitis.
- This nummular, anterior stromal keratitis may be misdiagnosed as a viral keratitis.
- Bullae may form within the edematous epithelium.
- Symptoms include redness, irritation, photophobia, tearing, decreased vision, and blepharospasm.
- The process usually regresses over a period of months, with the peripheral infiltration clearing. It then may remain peripheral or progress centrally.
- Late corneal findings include vascularization, scarring, and associated irregular corneal astigmatism.
- Vascular flow gradually decreases to leave characteristic ghost vessels.
- Mild conjunctivitis and anterior uveitis with fine keratic precipitates also may develop in association with the keratitis;
- 10% of cases may present with episcleritis, scleritis, or other noninterstitial keratitis anterior segment inflammatory disease, which has been termed 'atypical Cogan's syndrome'.
- Posterior signs may include vitritis, choroiditis, pars planitis, or cotton-wool spots.
- Diagnosis acute onset of vestibuloauditory findings concomitant with the ocular disease

- The former may include fluctuating sensory hearing impairment, tinnitus, vertigo, and reduced vestibular response.
- Atypical Cogan's syndrome may include posterior scleritis, episcleritis, or uveitis associated with vertigo, tinnitus, and deafness.
- The most common eye symptoms associated with the ocular inflammatory disease are pain, redness, and photophobia.
- **Immediate diagnosis** and medical intervention may provide **optimum auditory recovery and preservation**

DIFFERENTIAL DIAGNOSIS OF INTERSTITIAL KERATITIS
Infections
Congenital/acquired syphilis Lyme disease Chlamydia Viral keratitis
Vasculitis
Wegener's granulomatosis Polyarteritis nodosa
Collagen vascular disease
Rheumatoid arthritis Relapsing polychondritis Sarcoidosis

- Luetic interstitial keratitis is probably the most common infectious cause of interstitial keratitis. It differs from Cogan's syndrome in that the inflammation is typically bilateral and more progressive and severe in its course. The corneal inflammation in Cogan's is also more anterior while Luetic disease is more posterior. Finally, positive serology and other systemic clinical manifestations of congenital syphilis are present.
- systemic findings associated abnormalities of the skin, auditory, and cardiovascular systems
- Vasculitis does not appear to be the cause of the cochlear disease.
- most common ear findings include an acute onset of Meniere-like syndrome with tinnitus, nausea, vomiting, and vertigo. Common vestibuloauditory findings include nystagmus, oscillopsia, and hearing loss.
- Immediate diagnosis and systemic immunosuppressive therapy is necessary to prevent hearing loss.
- Most patients **(60-80%) become deaf if the syndrome is not treated**
- The major systemic complication is vasculitis and the **associated life-threatening aortitis**, which develops in 10% of patients and may lead to aortic insufficiency associated with exertional dyspnea and ischemic disease of other organs.
- Nonspecific weight loss, fever, headache with arthralgias and myalgias also may occur

- On pathology, cornea demonstrates a lymphocytic infiltrate, old, deep stromal vascularization, and thickened corneal epithelium.
- IgA and IgG against Chlamydia trachomatis
- Increases in HLA-B17, HLA-A9, HLA-Bw35, and HLA-Cw4 → genetic predilection?
- Leukocytosis is common and elevation in ESR and C-reactive protein
- Echocardiographic abnormalities, **cardiac catheterization, audiograms, aortic ultrasound, and angiography** should be performed if indicated.
- Calcific bony and soft tissue obliteration of the intralabyrinth fluid w\ high resolution CT/MRI.. Cochlear hydrops also may occur.
- multidisciplinary approach -- ophthalmologist and otolaryngologist, internal medicine physician
- Frequent audiologic assessment is necessary to monitor disease activity.
- **Systemic corticosteroid therapy** is indicated to manage the vestibuloauditory signs associated with most cases of typical Cogan's syndrome.
- good prognosis
- oral corticosteroids for 2-6 months.
- ESR CRP monitor subclinical disease activity
- High-dose combination therapy may be required for more severe ocular and/or systemic inflammatory disease, including large vessel aortitis.
- Aortic valve replacement may be required for severe insufficiency, which develops in 10% of patients.
- Systemic cyclophosphamide and cyclosporin have been used also for atypical Cogan's syndrome with severe, biopsy proved systemic vasculitis.
- Corticosteroids used late in the course of the disease may exacerbate the hearing loss if cochlear hydrops is present.
- The acute stage typically lasts months to years and the chronic phase lasts indefinitely.