Cogan's syndrome, which primarily affects children and young adults, causes eye inflammation, vasculitis, and hearing loss.

What is Cogan’s Syndrome?

Cogan’s syndrome is a rare autoimmune-mediated rheumatic disorder of interstitial keratitis (inflammation of the cornea) that develops in children and young adults shortly after their recovery from an otherwise unremarkable respiratory infection. First reported in 1945, Cogan’s syndrome causes symptoms that are similar to those of syphilitic keratitis although there is no evidence of syphilis infection in Cogan’s syndrome.

Who is Affected?

Cogan’s syndrome primarily targets children and young adults in their 20’s and 30’s. Cogan’s syndrome frequently occurs after an influenza-like infection. Although no definitive infectious agent has been established as a precursor of Cogan's syndrome, several studies show an association with a previous Chlamydia infection.

Symptoms

Cogan’s syndrome causes eye inflammation, including conditions such as keratitis, iritis, scleritis, or conjunctivitis, associated with balance problems, dizziness and ear ringing (tinnitus) and other symptoms that resemble Meniere’s disease. These vestibuloauditory symptoms can progress to rapid bilateral deafness due to inflammation affecting the 8th cranial nerve.

Other symptoms include vasculitis, nausea, vertigo, poor balance, fever, fatigue, weight loss, vomiting, light sensitivity, and a bilateral blurring of vision. Rarely, patients may exhibit enlarged lymph nodes, rash, chest pain, night sweats, arm pain, cardiac involvement and shortness of breath.

Systemic cardiac manifestations of Cogan’s syndrome include aortitis, aortic valve insufficiency, pleuritis, pericardial effusion, coronary arteritis, and possibly myocardial infarction.

Vasculitis in Cogan’s syndrome can cause major organ damage, and in rare instances it can be fatal. Hearing loss is often progressive and can lead to permanent deafness within two years without early diagnosis and treatment intervention.

Diagnosis
The erythrocyte sedimentation rate and the C-reactive protein tests show mild to marked elevations. Vasculitis, if present, can be detected with imaging tests and confirmed with tissue studies. While there are no formal diagnostic criteria for Cogan’s syndrome, it is diagnosed in patients recovering from respiratory infection with rapid-onset sensorineural hearing loss, eye inflammation, and symptoms of vasculitis. In a few cases, Cogan’s syndrome has been associated with Chlamydia infection, with patients showing high titers of antibodies to Chlamydia.

**Treatment**

Anti-inflammatory eye drops are used in cases of mild eye symptoms. If signs of infection occur, antibiotic eye drops can be used. In severe eye disease, oral corticosteroids or immunosuppressive medications such as cyclosporine or cyclophosphamide may be used. In cases where the eye’s blood vessels are damaged, surgical intervention and corneal transplant may be required.

If excess fluid in the inner ear affects balance, diuretics may be used to reduce fluid. Antihistamines such as diphenhydramine (Benadryl) are also used for problems with imbalance. Cochlear implants are used in cases of hearing loss.

**Resources:**


Cogan’s Syndrome, National Association for Rare Diseases

S Van Doornum, G McColl, Prolonged prodrome, systemic vasculitis and deafness in Cogan’s Syndrome, Annals Rheumatic Disease, 2001,

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