

Cogan and Behcet Syndromes



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KEYWORDS

- Behcet syndrome • Cogan syndrome • Large vessel vasculitis

KEY POINTS

- Cogan syndrome (CS) is a triad of inflammatory eye disease, vestibuloauditory dysfunction, and vasculitis.
- Consider the diagnosis of CS when inflammatory eye and ear disease present together.
- The hallmark of Behcet syndrome (BS) is mucocutaneous oral and genital ulcers, but BS can have heterogeneous manifestations.
- The possibility of large vessel involvement must be considered in all patients with CS and BS.
- Treatment of CS and BS is tailored to the disease manifestations and severity.

COGAN SYNDROME

Cogan syndrome (CS) is a rare autoimmune systemic disease, which is characterized by the combination of inflammatory eye disease and vestibuloauditory dysfunction, but it can have varied clinical manifestations. Some patients also develop a large vessel vasculitis.^{1,2} Morgan and Baumgartner³ first described the syndrome in 1934 in their case study of a patient with interstitial keratitis (IK) and Ménière disease. However, the disease is named after David G. Cogan, an ophthalmologist, who subsequently described 4 cases of “non-syphilitic IK and vestibuloauditory syndromes.”⁴ Seventy years after its initial description, we continue to have a limited understanding of this uncommon and pleomorphic syndrome. CS is a clinically challenging syndrome to diagnose and treat.

Epidemiology

Little is known of the prevalence and incidence of this rare disease. Approximately 250 cases have been described in the literature in all the large case series combined.^{1,2,5,6}

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Disease onset is in early adulthood (20–30 years old).^{1,2} Cases of children and older patients (>50 years old) have been reported. Men and women are affected equally. There is no racial or ethnic predilection. CS may be more prevalent within the population of patients with inflammatory bowel disease (IBD). Review of the literature shows 33 cases reported of overlap CS and IBD.^{1,6–9}

Pathogenesis

The pathogenesis of CS is unknown. Little is learned from autopsy of temporal bone and histopathology of corneal tissue. Specimens show nonspecific lymphocytic and plasma cell infiltration.^{10,11} There has not been a description of vasculitis seen on pathology in the eye or ear. However, most specimens described in the literature have been examined post mortem and are from patients who were treated with immunosuppressive agents, perhaps masking the true disease. In cases with systemic vasculitis, vessel wall histology shows acute and chronic inflammation.^{12–14} One case showed a dense mononuclear infiltrate, with multiple microabscesses.¹² There have been no reports of giant cells or granulomatous inflammation.

There is increasing evidence to support autoimmunity in CS. Autoantibodies directed against antigens found in the inner ear have been identified in sera of patients with the syndrome.^{15,16} Lunardi and colleagues¹⁵ reported antibodies against a peptide that they called Cogan peptide, found in pooled sera from 8 patients with CS. This peptide shares sequence homology with CD148 and connexin 26, which are expressed both in the endothelial cell and in inner ear cells. Compelling evidence of a pathologic role for Cogan peptide comes from animal models. Passive transfer of antibodies to the peptide reproduced symptoms of CS in mice.

Another proposed target is heat shock protein 70 (HSP-70). Antibodies to HSP-70 have been reported in patients with autoimmune sensorineural hearing loss (SNHL).^{16–18} In a study of patients with CS, 92% had anti-HSP-70 antibodies compared with only 5% of controls.¹⁷ No definite role for these antibodies has been proved, and more research is needed to examine their pathogenic role.

Traditional rheumatologic antibodies such as antinuclear antibodies and rheumatoid factor have not been consistently found in CS. Although there are several case reports of antineutrophilic cytoplasmic antibodies (ANCA) associated with CS,^{19,20} in larger series, ANCAs were found in a small percentage of patients with CS.^{1,2}

Symptoms

CS is a multisystem disease, with heterogeneous presentations. The classic presentation as outlined in Cogan's original criteria is a triad of (1) nonsyphilitic IK, (2) vestibuloauditory symptoms, and (3) an interval between ophthalmologic and auditory symptoms of less than 2 years. A summary of the most common manifestations is presented in **Box 1**. The term atypical CS has been used to describe patients who present with a variation of these features. This presentation can include inflammatory ocular manifestations other than IK or a longer delay between eye and ear symptoms. Ocular and audiovestibular symptoms typically have a rapid onset. Either organ can present first, or they may present simultaneously. Ten percent to 15% of patients have vasculitis, which rarely is the initial presentation. IK, which results in inflammation of the cornea, is the most common eye disease. Symptoms include photophobia, pain, redness, tearing, and blurring of vision. The inner ear disease in CS is part of a group of immune-mediated inner ear diseases (IMIED), also known as autoimmune inner ear diseases (AIED). Deafness occurs in 30% to 50% of patients.^{1,2,6} Long-term disease and repeated episodes may also result in the sequela of cochlear

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