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Short Report

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A Case of Atypical Cogan's Syndrome and Review of Literature

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Cogan's Syndrome (CS) is a disease affecting young adults and consists of episodes of acute interstitial keratitis with vestibuloauditory dysfunction. This syndrome was originally described in 1945 by Cogan as a disorder of the eyes and ears, characterized by nonsyphilitic interstitial keratitis with deafness. Sporadic reports showing the development of vasculitis in patients with eye and ear disease have been published over the last 30 years. When ocular signs, in addition to interstitial keratitis, are present, the syndrome is referred to as atypical Cogan's Syndrome. Although cases of scleritis and posterior scleritis accompanying interstitial keratitis have been reported in the literature (1, 2), iridocyclitis is extremely rare (3). Presented below is a report of an atypical CS with iridocyclitis, and a review of the literature to familarize doctors with this unusual but treatable disorder.

A 17-year-old man was admitted to hospital on June 25, 1996, suffering from deafness, a burning sensation in the eyes, redness of the eyes and blurring of the vision. He had been well until a week before, when he began to experience wheezing. He was admitted to a local hospital, but deafness and blurring of the vision were added to the clinical picture and the patient was referred to our hospital.

An ophthalmologic examination revealed that visual acuity was 0.2 bilaterally. Conjunctival hyperemia and mild ciliary injection, and bilateral interstitial keratitis, as well as iridocyclitis with three positive cells in the anterior chamber were detected with a slit lamp examination. Intraocular pressures were within the normal limits. Ophthalmoscopy at the acute stage was not possible because of anterior segment reaction. There was no history of Behçet's disease or other vasculitic disorders.

On physical examination, bilateral deafness was detected. Rinne tests were negative and auditory-evoked potential was not detected in either ear. Bilateral total deafness was detected through pure tone audiometry. A complete blood count and urinanalysis were found to be normal. Serologic tests for syphilis were negative. Electroencephalography, cranial computerized tomography and visual-evoked potentials were also normal. Echocardiography revealed that the heart was hyperdynamic.

High-dose prednisolone (100mg/day) was initiated. The patient also received topical prednisolone eye drops 8 times daily and cycloplegics as well as warm compresses, 3 times daily. The iridocyclitis was completely resolved within ten days. After three months, the interstitial keratitis showed significant improvement and visual acuity increased bilaterally by 4 to 5 lines. Ophthalmoscopy did not reveal any abnormality in either eye. In general, good progress was observed. However, total deafness remained a major problem, showing no improvement despite therapy.

CS was first described by Cogan in 1945. It is characterized by nonsyphilitic interstitial keratitis and vestibuloauditory dysfunction. The average age for the onset of the disease is twenty-five, but it has been reported in patients between 4 and 63 years of age (4). Our patient was 17 years old and did not show any clinical or laboratory evidence of syphilis.

The primary ocular manifestation of typical CS is chronic bilateral interstitial keratitis. Keratitis is manifested by patchy infiltrates of the deep corneal strome. Atypical forms of CS have been described with bilateral posterior scleritis (1). However, iridocyclitis occuring as the accompanying ocular lesion is extremely

rare. Klausen et al. (3) reported such a case in 1992 and, to our knowledge, ours is the second case with iridocyclitis accompanying interstitial non-syphilitic keratitis. The visually evoked potential (VEP) may be normal, as was pointed out by Magni et al. (5). Our patient also had a normal VEP.

Vestibuloauditory involvement typically begins unilaterally with hearing loss and tinnitus, accompanied by vertigo, nausea, vomiting, ataxia and nystagmus (6). Vestibuloauditory dysfunction may occur concomitantly with the visual symptoms or they may precede or follow the onset of visual symptoms by as much as three months. Although hearing loss is complete in CS, sometimes it may be preceded by exacerbations and remissions. Similar involvement of the other ear often follows within the next few weeks or months, leaving the patient permanently deaf. Hearing tests in the early stages of the disease show cochlear involvement. Caloric tests usually produce negative results. In some patients with CS, small calcified obliterations were detected with computerized tomography and a high signal inside the membranous labyrinth was observed on T1 weighted magnetic resonance images (7). Cranial computerized tomography in this case was normal.

A review of 109 cases revealed that 72 percent of patients had clinical manifestations beyond the ophthalmic, auditory and vestibular signs (8). The most important systemic complication of CS is aortic valve involvement with acute aortitis in up to 10 percent of patients. In our case, echocardiography showed that the heart was hyperdynamic. Other systemic complications are unexplained fever (34%), heart disease (25%), gastrointestinal signs (25%), musculoskeletal involvement (23%), hypertension (9%) and cerebral artery occlusion (5%). The patient in this case did not have these complications.

The pathogenesis of CS has not yet been explored. Autopsy findings revealed aortitis with involvement of all layers of the aorta, secondary heart and kidney infarction, and necrotizing angiitis in the dura, brain, gastrointestinal tract, spleen and kidneys as well as focal degeneration of arterial walls at the extremities and lymphocyte and plasma cell infiltration in deep layers of the cornea (9). Ishii et al. reported that the ligamentum spirale cochlea was infiltrated with plasma cells and lymphocytes in CS; endolymphatic hydrops in the cochlea, collapse of the saccule and fibrosis in the posterior semicircular canal were also described by the same authors (10).

The presence of lymphocytes and plasma cells in the conjunctiva and cornea suggests immune-mediated damage. Anterior uveitis, detected in this case, may also suggest a possible immune mechanism. However, the exact mechanism has yet to be revealed. CS has been detected in a patient with antiphospholipid antibodies (11). CS associated with Crohn's disease has also been reported; immune-mediated mechanisms again were thought to play a major pathogenic role (12). An interesting clinical entity, autoimmune sensorineural hearing loss, is also believed to be closely related to Cogan's syndrome (13, 14, 15).

There is no effective therapy for CS. As for the management of keratitis, topical steroids are beneficial. The treatment of uveitis, if present, is not different from the classical uveitis management, involving topical steroids, mydriatics and cycloplegics as well as warm compresses whenever necessary. Our case also responded well to such therapy. Systemic corticosteroids are indicated in the early treatment of acute deafness in CS. Therefore, one to two mg/kg of oral prednisone may be administered for 2 to 4 weeks. If hearing is improved by this treatment, corticosteroids should be continued to be taken on alternate days. Hearing did not improve in our case and there are other similar examples of such in the literature (16).

Here a case of atypical CS has been described and the relevent literature reviewed. The importance of early diagnosis and treatment to prevent hearing loss have been emphasized once more.

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