Cogan-I-Syndrome
Interstitial Keratitis with Sudden Deafness

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Summary. The present paper records the case of a young man, aged 19, with influenza followed suddenly by a Cogan-I-syndrome. The corneal infiltrations recede after local treatment with prednisolone, an effect which cannot be observed at the beginning of the disease under the same therapy. At the end of the illness only a few corneal alterations are detectable. Both vestibulae are inactive, the patient remains deaf except for residual hearing on the left side.

Possible causes of the syndrome are discussed with the aid of the available literature, and diseases with similar symptomatology are mentioned.

In 1945 David G. Cogan describes a syndrome, a combination of non-syphilitic interstitial keratitis and vestibulo-auditory symptoms with subsequent inner ear deafness and loss of the vestibular functions. In 1934 Mogan and Baumgartner report a similar case, they interpret as "Menière's disease complicated by recurrent interstitial keratitis". The present literature shows that this disease can arise in the eye or in the auditory and vestibular apparatus; mostly the other organ is affected, too, within a short time. In the stroma of both corneas you can find deep infiltrations, which are localised in the posterior half; these alterations are patchy or globular, opaque and mostly placed in peripheral regions of the corneas. In a short time profound, branching vascularisations grow into the corneal stroma. The conjunctiva is injected, we see photophobia, lacrimation and blepharospasm. Fundus, tension and visual fields are generally normal. The labyrinthine symptoms include vertigo, nausea,
tinnitus and diminution of hearing with subsequent bilateral deafness in most cases.

**Case Report**

Our patient is a 19 year old man. Except for German measles, pyelitis, otitis and pneumonia he did not go through any other diseases. In summer 1970 he had an exanthema after touching Greek peaches.

His parents and his brother are healthy. Veneral disease has not existed in the family.

*Present Illness.* In the middle of August 1970 the patient has an influenza with collapses, vertigo and vomiting; these symptoms appear in a more severe form two weeks later. He is hospitalized in a bad condition and with high fever; there he notices redness of both eyes with swelling of his lids, corneal opacities, strong photophobia, epiphora, and ocular pains.

The serious condition and the eye-symptoms improve, but in November tinnitus with disturbance of hearing and a staggering gait is observed. — After 6 weeks, however, he is discharged without any appreciable troubles of hearing and vision; just only an imbalance of gait persists.

Two months later a total loss of hearing suddenly follows. An inner ear deafness with keratitis of unknown cause is diagnosed. Treatment with antibiotics, gammaglobulins and several vitamins is ineffective.

In March 1970, the patient is transferred to Essen E.N.T. Clinic; he is examined regularly in our clinic from March to August 1971. In Essen Neurological Clinic he undergoes a complete check-up.

**Ophthalmological Examination.** Visual acuity of the right eye—0.6 p, of the left eye—0.5 p which does not improve with glasses. Tension (Schiotz): both eyes 14.5 mm Hg. Both visual fields are regular. The bulbi have normal motility, no nystagmus. The corneal sensibility is of the same quality on both sides, but slightly increased. Blepharospasm with epiphora and strong photophobia are present. The conjunctivae are badly injected. The pupils are normal, they react promptly to light and convergence.

Examination with the slit-lamp shows that the anterior chambers are optically empty except for a slight Tyndall phenomenon. In the posterior half of the corneal parenchyma there are many patchy, partly globular, whitish-yellowish, opaque infiltrates, which converge mutually. The alterations are predominantly confined to the middle periphery and the areas near the limbus; the corneal center remains free. Some of the spotty infiltrations dissolve to fine granules in contrast to the homogeneous shape of the spherical lesions (Fig. 1). On the left eye a vertical fold of Descement's membrane is found. From the limbus branching, deep vessels sprout into the cornea. The epithelium is clear and shining; there are neither opacities of the posterior surface nor precipitates. The keratitic infiltrates are more pronounced than on the right side. On the anterior capsule of the lenses pigmented stars are found.

Otologically a strong labyrinthine impairment of hearing exists with complete inactivity of both vestibula.

**Neurological-Internal Examination.** The reflexes, the electro-encephalogram, and the spinal-fluid tests are normal. In the beginning there is a non-compensated unsteadiness of gait. The Wassermann's reactions as well as additional reactions in blood and liquor cerebrospinalis are negative. Investigations for sarcoidosis, tuberculosis, rheumatic diseases, especially periarteritis nodosa, for cardiovascular diseases, for toxoplasmosis, listeriosis, Morbus Bang, leptospirosis and different viruses and bacteria do not show any pathological result. All the laboratory tests