The clinical behavior of orbital SFT is usually benign, although some of these tumors have exhibited aggressive features such as infiltration of the orbital roof and optic nerve, as well as extraorbital extension. The treatment for SFT is en bloc excision because incomplete removal results in a higher recurrence rate and carries a potential for malignant transformation. Careful long-term follow-up after complete excision seems to be the standard of care.

To our knowledge, this case represents the first published report of SFT arising from the sclera. Although rare, SFT should be included in the differential diagnosis of intraocular and epibulbar masses.

**Grant W. Su, MD**
**Noel Perez, MD**
**Kenneth B. Simons, MD**
**Gerald J. Harris, MD**

**Correspondence:** Dr Harris, The Eye Institute, 925 N 87th St, Milwaukee, WI 53226 (gjharris@mcw.edu).

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**Uveitis Associated With Atypical Cogan Syndrome in Children**

Atypical Cogan syndrome involves nonsyphilitic interstitial keratitis associated with audiovestibular involvement that progresses to complete deafness within 2 years. Atypical Cogan syndrome occurs when sensorineural hearing loss is associated with ocular inflammation distinct from interstitial keratitis, such as uveitis, scleritis, conjunctivitis, or retinal vasculitis. We describe 3 cases of atypical Cogan syndrome that were undiagnosed for a long time. Awareness of this syndrome will aid in the diagnosis and subsequent treatment of this unusual entity. Cogan syndrome is described as typical when it appears in young adults with flare-ups of interstitial keratitis and sudden onset of Ménière disease–like attacks, which progress to sensorineural deafness within 2 years. The keratitis is usually bilateral peripheral subepithelial keratitis, which can progress to nummular lesions. A deep stromal keratitis can occur and vascularization of the cornea usually occurs. Early diagnosis is most important, as early treatment of cochlear symptoms with corticosteroids may prevent or lessen the extent of deafness.

Atypical Cogan syndrome occurs when types of ocular inflammation other than interstitial keratitis happens in conjunction with hearing loss. We describe 3 adolescents with atypical Cogan syndrome who lost all of their hearing owing to sensorineural involvement.

**Report of Cases.** Case 1. A 16-year-old African American adolescent boy was seen 4 months after an acute loss of all of his hearing, which developed during 1 week. For the past 4 months, his eyes were constantly red. His pediatrician had prescribed him systemic antibiotics to subsequent treatment of this uncommon entity. Cogan syndrome involves nonsyphilitic interstitial keratitis associated with audiovestibular involvement that progresses to complete deafness within 2 years. Atypical Cogan syndrome occurs when sensorineural hearing loss is associated with ocular inflammation distinct from interstitial keratitis, such as uveitis, scleritis, conjunctivitis, or retinal vasculitis. We describe 3 cases of atypical Cogan syndrome that were undiagnosed for a long time. Awareness of this syndrome will aid in the diagnosis and subsequent treatment of this unusual entity. Cogan syndrome is described as typical when it appears in young adults with flare-ups of interstitial keratitis and sudden onset of Ménière disease–like attacks, which progress to sensorineural deafness within 2 years. The keratitis is usually bilateral peripheral subepithelial keratitis, which can progress to nummular lesions. A deep stromal keratitis can occur and vascularization of the cornea usually occurs. Early diagnosis is most important, as early treatment of cochlear symptoms with corticosteroids may prevent or lessen the extent of deafness.

Atypical Cogan syndrome occurs when types of ocular inflammation other than interstitial keratitis happens in conjunction with hearing loss. We describe 3 adolescents with atypical Cogan syndrome who lost all of their hearing owing to sensorineural involvement.
There were no corneal lesions. The dilated fundus examination findings were normal. Blood studies were negative for antinuclear antibodies and rheumatoid factors and showed a normal complete blood count and negative angiotensin-converting enzyme levels.

We diagnosed bilateral atypical Cogan syndrome in the patient. The bilateral iritis resolved with cycloplegics and topical steroid drops. The patient has been referred for a cochlear implant. He had another episode of bilateral iritis 3 months later, which also resolved with topical cycloplegic and steroid eye drops.

Case 2. A 10-year-old white boy had a history of bilateral red eyes for 1 month at initial examination. One year prior to this eye examination, he had acutely lost all of his hearing and had undergone a cochlear implant in his right ear. Visual acuity was 20/30 OD and 20/20 OS. He had bilateral iritis with posterior synechiae in the left eye. There were no corneal lesions and the dilated fundus examination findings were normal. Blood studies were performed and levels of rheumatoid factor, fluorescent treponemal antibody absorption, angiotensin-converting enzyme, and complete blood count were all normal. The iritis resolved quickly with topical dilating and corticosteroid drops. Six months later, the patient had another episode of bilateral iritis without corneal inflammation, which also resolved with topical drops. At the 2-year follow-up, there was no recurrence.

Case 3. A 13-year-old white boy had a history of red eyes off and on for the past 2 years, which were treated with topical antibiotic drops. He had lost all hearing acutely during a 2-week period 18 months previously. He had received a successful left cochlear implant. Visual acuity was 20/30 OU and bilateral iritis was present. There were no corneal lesions and the dilated fundus examination findings were also normal. Blood tests showed that levels of fluorescent treponemal antibody absorption, rheumatoid factor, complete blood cell count, and angiotensin-converting enzyme were all normal. The iritis resolved with topical cycloplegic and steroid drops.

The patient had 1 recurrent episode of bilateral iritis 4 months later and now has been clear for 2 years.

Comment. Cogan syndrome mainly affects young adults but can appear from ages 3 to 40 years. Slightly more males are affected than females. The etiology is unknown, but in approximately 20% of cases, the onset is preceded by an upper respiratory infection. In 41% of cases, the eye is affected first; in 43%, the ear is affected first; and in 16% of cases, both organs are affected at the same time. The interval between ocular and ear involvement can be as short as 3 months in the typical syndrome or as long as 11 years in the atypical syndrome. Making the diagnosis of Cogan syndrome is important, not only because vision can be lost, but also because 10% of cases are complicated with aortic insufficiency, which can be life threatening.4 Polyarthralgias or arthritis can also accompany the disease.

Neurologic findings, such as epilepsy or encephalitis, can also be seen. Almost 90% of patients have severe hearing loss or total bilateral deafness. When deafness is not complete, high-dose corticosteroids can lead to hearing improvement in 50% of cases, but if deafness is already total, the corticosteroids do not help.3

The atypical form of Cogan syndrome can be an overlooked cause of uveitis in children; the disease had been undiagnosed in our patients until their initial visit to our office. One patient had seen 9 physicians before we made the association between uveitis and sensorineural hearing loss. Many articles on pediatric uveitis do not even mention atypical Cogan syndrome as a cause of uveitis. While most cases of uveitis will not involve sensorineural hearing loss, the small percentage that will be seen with this serious complication warrants a recommendation to evaluate the patient’s hearing status when no etiology for the uveitis is found.

Zane F. Pollard, MD
Marc Greenberg, MD
Alice Bashinsky, MD
Mark Bordenca, MD

Correspondence: Dr Pollard, Department of Ophthalmology, Children’s Healthcare of Atlanta at Scottish Rite Children’s Hospital, James H. Hall Eye Center, 5445 Meridian Mark Rd, Ste 200, Atlanta, GA 30342 (zanepollard@bellsouth.net).

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IgG4-Related Chronic Sclerosing Dacryoadenitis

Recent evidence suggests that Mikulicz disease is distinct from Sjögren syndrome and is an IgG4-related systemic disease.1 Herein we report 4 cases of Mikulicz disease in which the serum IgG4 concentrations were elevated and infiltration of IgG4-stained plasma cells with sclerosing fibrosis was pathologically observed in the lacrimal gland.

Report of Cases. Three women (aged 46, 47, and 64 years) and 1 man (aged 66 years) were referred with swelling of the lacrimal gland region (Table). Every patient had experienced a chronic episode (3 months to 5 years) of progressive eyelid swelling. The 64-year-old woman (patient 3) had a history of surgical resection of a subcutaneous mass in the upper eyelid, which was pathologically diagnosed as lymphoid hyperplasia at a different hospital 2 years prior to the initial visit. Magnetic resonance imaging depicted well-circumscribed masses including bilateral lacrimal glands in the lacrimal fossa (Figure 1). In ev-