Polskie Towarzystwo Okulistyczne
www.pto.com.pl

KLINIKA OCZNA 2023, 125, 4: 252-256
Received: 20.12.2021 Accepted: 28.04.2022

CASE REPORT

Access this article online

Website:
www.klinikaoczna.pl

DOI:
https://doi.org/10.5114/ko.2023.133446

Atypical Cogan's syndrome case with recovery of hearing loss

Piotr Lasota, Krzysztof Kiciński

Department of Ophthalmology, Provincial Specialist Hospital, Slupsk, Poland

ABSTRACT

Aim: To report a case of Cogan's syndrome with successful recovery of hearing loss.

Case report: A 26-year-old man with interstitial keratitis was treated by a local ophthalmologist for conjunctivitis after parainfluenza. As no improvement took place and vestibular symptoms occurred, the patient was hospitalized. Cogan's syndrome with profound hearing loss was diagnosed. Long-term steroid therapy gradually reduced the inflammation. After 5 months of treatment the hearing successfully recovered and no systemic symptoms occurred until the seventh month of follow-up.

Conclusion: Early diagnosis and immunosuppressive treatment can help to prevent irreversible deafness.

KEY WORDS: atypical Cogan's syndrome, interstitial keratitis, hearing loss, steroids, autoimmune disease.

INTRODUCTION

In 1934 Morgan and Baumgartner described a patient with Ménière's disease and interstitial keratitis [1]; however, the name of the syndrome originates from the ophthalmologist Dr David G. Cogan. In 1945 he presented a series of 4 patients with non-syphilitic interstitial keratitis and vestibuloauditory symptoms [2]. This is a rare (less than one case in 5000 people), auto-immune vasculitis in relatively young patients with average age of 30 years [3], involving mainly the eye and internal ear [4]. In addition, systemic involvement can occur with serious complications. In many cases the viral infection precedes the syndrome, with the infectious process likely dysregulating the immune system. Diagnostic errors are common because the symptoms and signs are usually not specific and there is no reliable clinical laboratory test to confirm the diagnosis [5]. Commonly a delay of diagnosis leads to a late and advanced presentation. The interdisciplinary approach with immunosuppressive treatment is necessary as soon as possible. If not, bilateral deafness in the young patient can occur, with a cochlear implant being the only possible treatment. In 1981 Haynes *et al*. divided Cogan's syndrome into 2 forms: typical and atypical [6]. The typical form presents with interstitial keratitis and hypoacusis similar to Ménière's disease. The atypical form presents with hypoacusis with inflammation of any part of the eye. As the syndrome is usually a chronic disease, typical Cogan's syndrome can evolve over the years to atypical Cogan's syndrome as well.

CASE REPORT

The 26-year-old man presented with complaints of photophobia, eye pain, decreased vision, redness of the eyes and dizziness. His eyes had been red for about 2 weeks, and the condition had been treated as conjunctivitis with local antibiotics. His recent medical history also included parainfluenza infection about 2 weeks before and an emergency department visit 4 days prior to presentation. The diagnosis of sinusitis was considered by an ENT doctor due to the thickening of mucosa in sinuses on the CT examination. As the inflammatory markers slightly decreased, the patient was discharged from the emergency department on oral levofloxacin after a 1-day stay.

When he presented at the hospital again directly to the ophthalmology department his visual acuity (Snellen) was 0.1 and 0.4 for the right and left eye respectively. Signs of diffuse episcleritis, interstitial keratitis and anterior uveitis were present. All these signs were much more prominent on the right eye (Figure 1). The corneal opacifications were mainly located at the periphery and were similar to nummular keratitis. The fundus examination was normal on the left eye but was not possible on the right eye. B-scan ultrasound showed bilateral scleritis (Figure 2). The patient also suffered from vertigo with unstable walking and general fatigue. No hearing problem was mentioned. The patient was hospitalized, and a bolus of general steroids was given with two broad spectrum antibiotics. Additionally, steroid drops (dexamethasone) were prescribed. Initially CRP was 98 and

CORRESPONDING AUTHOR

Piotr Lasota, MD, Department of Ophthalmology, Provincial Specialist Hospital, 1 Hubalczykow St., 76-200 Slupsk, Poland, e-mail: piotr_lasota@yahoo.fr

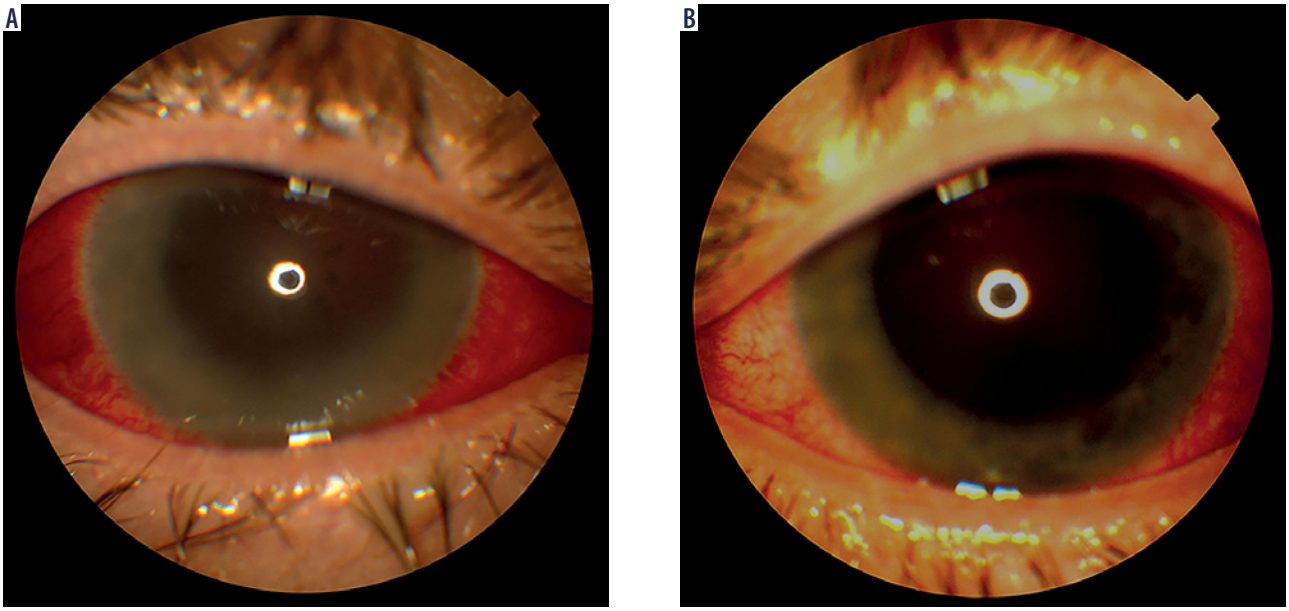

Figure 1. A) RE on the left with diffuse scleritis, edematous cornea and pigment on the surface of the lens. **B**) Left eye episcleritis

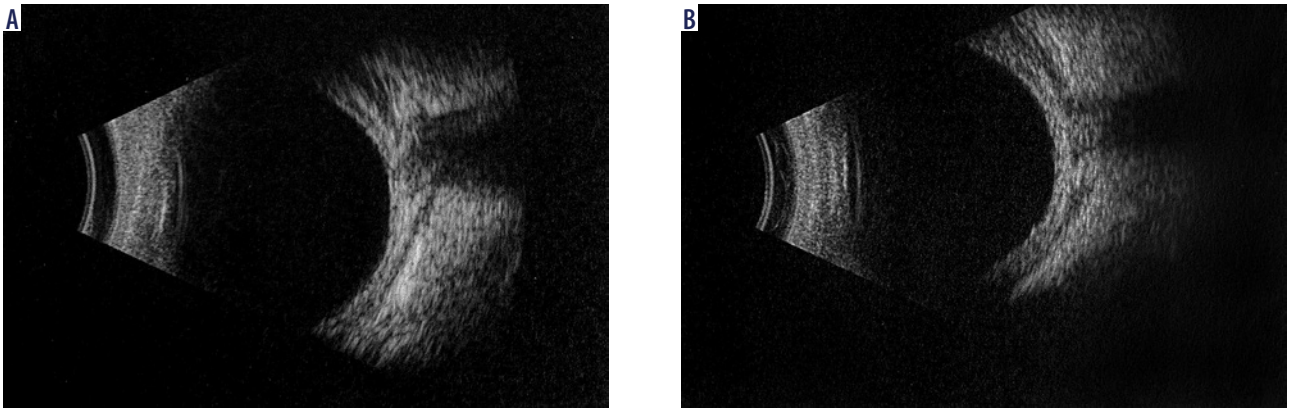

Figure 2. B-scan ultrasound: hypoechogenic band under the tenon's capsule in both eyes (T-sign)

leukocytosis was 20 810. MRI of the orbits confirmed scleritis and perineuritis of the optic nerve of both eyes. Apart from that, acoustic neurinoma was excluded by MRI. Other tests including a broad panel of antibodies (cANCA, pANCA, profile ANCA, ANA, aCL, CCP), chest radiograph, MRI and angio-MRI of the brain, cardiac echocardiogram, abdominal ultrasound, lumbar puncture, ionogram, proteinogram, rheumatoid factor, C3 and C4 complement were all negative. Cogan syndrome was suspected because of signs of interstitial keratitis with concomitant ENT symptoms (dizziness and tinnitus). Therefore an ENT consultation was requested. ENT examination revealed acute profound sensorineural hearing loss of the right ear (Figure 3A) and significantly decreased labyrinth function bilaterally (Brunings test). The therapy with intravenous steroids was prolonged and replaced with oral treatment, which gradually decreased to 5 mg of prednisone. At the 1-month follow-up visit VA was 1.0 for both eyes. Apart from mild anterior vitritis of the right eye, no other abnormalities were found including no signs of retinal vasculitis on fluorescein angiography. The hearing loss of the right ear decreased (Figure 3B) but dizziness was still present.

At the 2-month follow-up visit the RE was red again with small opacifications of the peripheral superior cornea although oral steroid treatment with 10 mg of prednisone was ongoing. Local dexamethasone was added and improvement was noted after one week. However, at the 3-month follow-up the redness reoccurred in both eyes without affecting the visual acuity. Dizziness decreased but was still present. Therefore oral steroids were increased to 15 mg and local dexamethasone maintained. At the 5-month follow-up visit the audiogram of the right ear looked normal (Figure 3C). Interestingly, now the right eye was normal but the left eye had congested conjunctiva with slight opacification of the peripheral cornea. The patient missed the 6-month follow-up visit and stopped oral and local steroids on his own at that time.

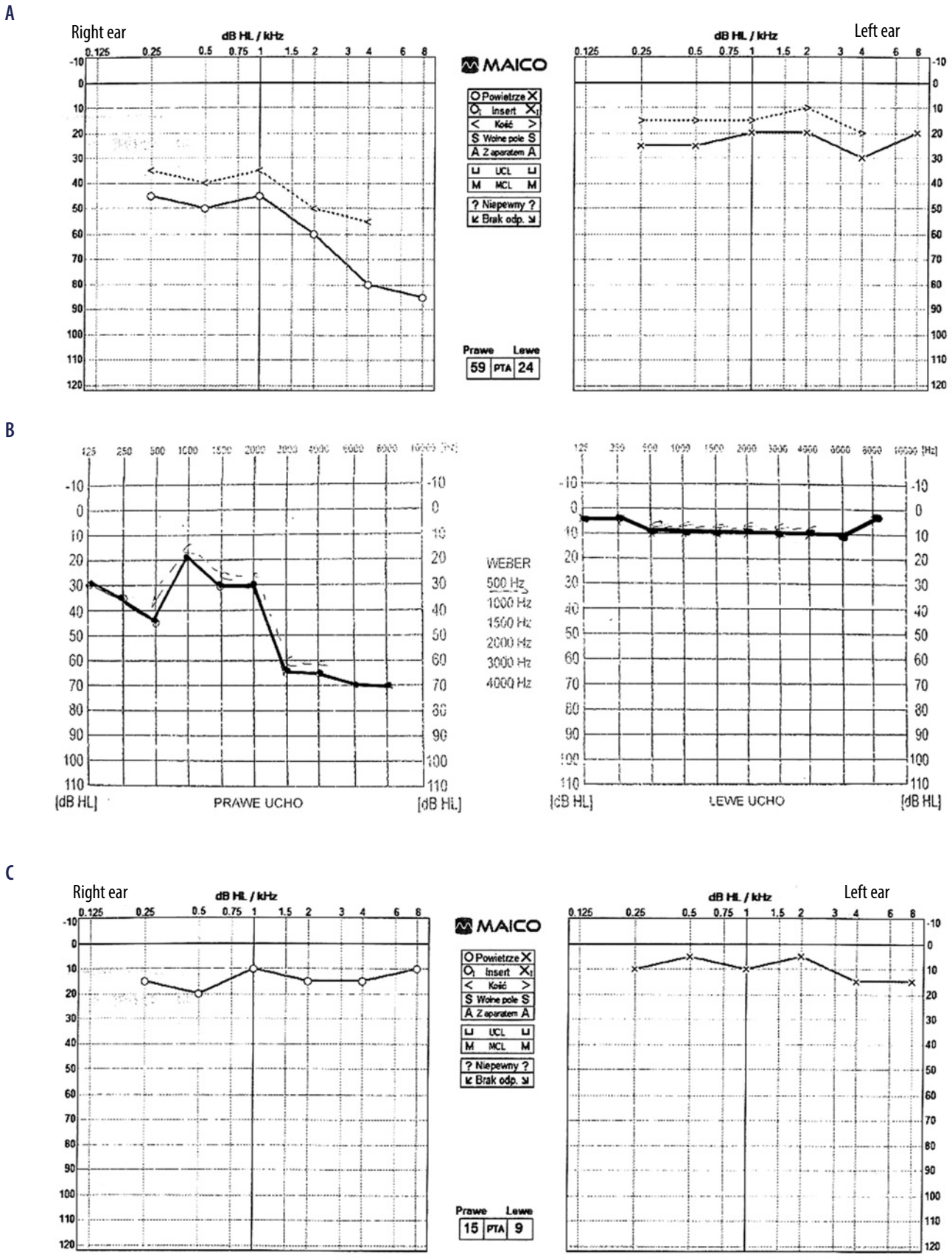

Figure 3. Audiograms: on the left – the right ear, with profound hearing loss at baseline (**A**), slight improvement of high frequencies at 1-month FU (**B**) and return to normal at 5-month FU; (**C**) on the right – the left ear, which is normal and stable

At the 7-month follow-up visit no ocular inflammation was found. According to the patient the hearing had markedly improved but its quality had not returned to the baseline.

DISCUSSION

Cogan's syndrome is a rare disease with a poor prognosis if not diagnosed early. Our patient is an example of atypical Cogan's syndrome. The conjunctiva, sclera, cornea, iris, optic nerve and internal ear were affected. Asthenia as a systemic symptom was present. Fortunately, there was a relatively short time interval (2-3 weeks) between ocular and ENT symptoms/signs, which helped us to make a proper diagnosis after just a few days. As the patient suffered from parainfluenza infection and had red eyes before hospitalization, he was diagnosed with conjunctivitis by another ophthalmologist, which is a common mistake. According to Durtette's review, the median time to the diagnosis of Cogan syndrome from the first signs is 12 months [7].

The wrong diagnosis prolongs the inflammation (at admission CRP was 98), which silently damages the internal ear. Therefore if there is no conjunctival secretion and there are some subtle cornea peripheral opacities we should avoid just simple diagnosis of conjunctivitis. In our case irregular corneal infiltrations were close to the limbus and were located in the anterior part of the cornea. The typical Cogan's syndrome keratitis does not decrease the visual acuity because the changes are not on the visual axis. However, our patient's vision decreased due to diffuse corneal edema, especially on the right eye, and optic perineuritis. The anterior chamber was difficult to assess but the pigment on the anterior capsule suggested broken posterior synechiae. The strong pain of the eyes was due to keratitis, scleritis and perineuritis. All these ocular signs/symptoms responded very quickly to the bolus of steroids (initially dexamethasone and later on methylprednisolone), and this is quite common [5-7].

There were three recurrences of interstitial keratitis but not so intense as at baseline. It is quite typical for the disease course. At different intervals keratitis reoccurs with different intensity. After an initial flare lasting a few weeks, the disease becomes chronic with remission and progression [8]. It means that the inflammatory process is long, and when steroids are weaned off too fast, it may relapse. There was no retinal vasculitis, macular edema, papilledema, or central retinal occlusion in our patient.

In contrast to the rapid ophthalmological improvement, the symptoms of internal ear disease (similar to Ménière's disease) were more resistant. They started with vestibular manifestations (vertigo and tinnitus), followed by hearing loss, which is consistent with the findings of Grasland *et al*. All these ENT symptoms are due to autoimmune inner ear disease. In the literature autoantibodies against the inner ear are reported with small vessel vasculitis in the cochlea and vestibular system [5, 9].

Regarding our patient, the auditory function of right ear improved very little after 1 month of treatment, with dizziness still being present. Fortunately a significant improvement finally took place after 5 months of steroids (Figure 3C). The hearing of the right ear became normal again. This is a very positive outcome. The early diagnosis with aggressive steroid treatment helped in auditory recovery [10]. Usually if deafness occurs, it is irreversible, although some improvements can happen. Grasland analyzed 111 patients and found 50 cases of deafness [8].

In the case of resistance to steroids, other immunosuppressive drugs should be prescribed quickly. Infliximab has an 80% response rate regarding the vestibuloauditory symptoms resistant to other therapies [7].

We can also suppose that early treatment has prevented general manifestations in our patient so far. Systemic symptoms of Cogan syndrome usually include fever, weight loss, fatigue, arthromyalgia, and abdominal pain. Coronary failure, renal stenoses, meningitis, encephalitis and cerebral stroke can also occur. Up to 68% of patients already have systemic symptoms at the time of diagnosis, but some of them may occur many years after the diagnosis. In the series of Grasland *et al*. 5 of 32 patients developed new systemic manifestations (aortic aneurysm, carotid stenosis, aortitis, nephrotic syndrome and polychondritis) between 6 months and 12 years after the diagnosis.

Therefore a long-term follow-up is necessary in Cogan's syndrome to avoid potentially lethal complications.

DISCLOSURE

The authors declare no conflicts of interest.

References

1. Morgan RF, Baumgartner CJ. Meniere's disease complicated by recurrent interstitial keratitis: excellent result following cervical ganglionectomy. West J Surg 1934; 42: 628.
2. Cogan DG. Syndrome of nonsyphilitic intrestitial keratitis and vestibuloauditory symptoms. Arch Ophthalmol 1945; 33: 143-149.
3. Oliner L, Taubenhaus M, Shapira TM, et al. Nonsyphilitic interstitial keratitis and bilateral deafness Cogan's syndrome) associated with essential polyangitis (periarteritis nodosa). A review of the syndrome with consideration of a possible pathogenic mechanism. N Engl J Med 1953; 248: 1001-1008.
4. Cheson BD, Bluming AZ, Alroy J. Cogan's syndrome: sistemic vascultis. Am J Med 1976; 60: 549-555.
5. Shamriz O, Tal Y, Gross M. Autoimmune Inner Ear Disease: Immune Biomarkers, Audiovestibular Aspects, and Therapeutic Modalities of Cogan's Syndrome. J Immunol Res 2018; 2018: 1498640.
6. Haynes BF, Pikus A, Kaiser-Kupfer M, et al. Successful treatment of sudden loss in Cogan's syndrome with corticosteroids. Arthritis Rheum 1981; 24: 501-503.
7. Durtette C, Hachulla E, Resche-Rigon M, et al. Cogan syndrome: characteristics, outcome and treatment in a French nationwide retrospective study and literature review. Autoimmun Rev 2017; 16: 1219-1223.

8. Grasland A, Pouchot J, Hachulla E, et al. Typical and atypical Cogan's syndrome: 32 cases and review of the literature. Study Group for Cogan's Syndrome. Rheumatology (Oxford) 2004; 43: 1007-1015.
9. Espinoza GM, Wheeler J, Temprano KK, Keller AP. Cogan's Syndrome: Clinical Presentations and Update on Treatment. Curr Allergy Asthma Rep 2020; 20: 46.
10. Vollersten RS, McDonald TJ, Younge BR, et al. Cogan's syndrome: 18 cases and a review of the literature. Mayo Clin Proc 1986; 61: 344-361.