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A case report of Cogan syndrome based on clinical manifestations with dramatic response to immunosuppressive therapy

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Case Report

Mohamad S*, Akhtarzai A and Zaqoot S

Department of Otolaryngology Head & Neck surgery, NMC Royal Hospital Sharjah, UAE

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***Corresponding author:** Mohamad S, Department of Otolaryngology Head & Neck surgery, NMC Royal Hospital Sharjah, UAE

Abstract

Cogan's syndrome is a rare inflammatory disorder mainly affecting ocular and audio vestibular system. The typical picture are of interstitial keratitis and sudden onset of Ménière-like attacks (nausea, vomiting, tinnitus, vertigo and hearing loss). A high index of suspicion and multispecialty input will help in the early initiation of therapy and reduce long-term morbidity.

We report a case of this rare entity where the patient had a dramatic response to steroid and immunosuppressive drugs. Considering the variable onset and development of symptoms as well as the lack of specific laboratory tests, the diagnosis of Cogan's syndrome is also a challenge and is often based on clinical features and response to immunosuppressive treatment.

Introduction

Cogan's syndrome is a rare disorder of unknown origin characterized by inflammatory eye disease and vestibulo-auditory symptoms, which primarily affects young white adults, without a hereditary pattern. Mostly affects caucasian young adults. The average age of onset is 25 years, and there is no gender-specific prevalence It was first described by an ophthalmologist, Dr David G. Cogan, in 1945, who reported on a "syndrome of non-syphilitic interstitial keratitis and vestibuloauditory symptoms even though, in 1934, Morgan[1].

Cogan's syndrome is mainly characterised by acute interstitial keratitis with audio-vestibular dysfunction, associated in close temporal proximity to flares of interstitial keratitis clinically indistinguishable from episodes of Ménière's disease[2].

Case Presentation

A 14-year-old girl of Arabic origin first presented to the Ear Nose and Throat out-patient clinic with a 3-week history of vertigo, left tinnitus and bilateral hearing loss. She initially was treated as vestibular neuritis with vestibular sedatives and small dose of short course of oral prednisolone with no response. She then developed bilateral red eyes on further follow up review. Therefore, an autoimmune disease was suspected. She was referred and evaluated by Ophthalmologist, which confirmed bilateral diffuse interstitial keratitis. Based on the clinical picture, a diagnosis of cogan syndrome was suspected.

The patient had a normal inflammatory response and her screen for anti-nuclear antibodies, extractable nuclear antigen, rheumatoid factor and anti-neutrophil cytoplasmic antibodies were all negative. A full virology screen was also negative. Having failed to respond to all

supportive treatment modalities, a trial of a tapering two-week course of prednisolone starting at 25mg daily was instigated. The patient's symptoms improved by more than 50% on glucocorticoids subsequent to which the diagnosis of Cogan syndrome was established. As the patient's symptoms relapsed when glucocorticoids were stopped, she was recommenced on prednisolone 25mg in line with azathioprine 100mg daily.

The patient's hearing, which was almost lost in the left ear, improved significantly by more than 70% initially, and then to normal hearing as stated by the patient supported by special ENT testing (Audio). Her balance problems and vertigo also improved remarkably. Her tinnitus, which was the last symptom to resolve, almost went away after 2 months of treatment. The patient has now come off glucocorticoids and is staying on Azathioprine 75mg daily.

Discussion

Cogan's syndrome is a rare clinical disease, which primarily affects young adults; however, published reports range from 2.5 to 60 years for age of onset. This disease primarily affects whites and no evidence for hereditary involvement. Typical CS is characterized by interstitial keratitis(IK) and vestibuloauditory dysfunction. The interstitial keratitis(IK) usually occurs with acute onset and characterized by photophobia, lacrimation, and ophthalmic pain.. Clinical data from a literature review of 32 patient, seventeen patients had typical Cogan's syndrome and 15 had atypical Cogan's syndrome. Apart from non-syphilitic interstitial keratitis, the ocular manifestations of patients with atypical Cogan's syndrome were mainly uveitis and episcleritis [4,5].

In 1980, Haynes et al is specified the diagnostic criteria for typical and atypical Cogan's syndrome, which include a large spectrum of clinical manifestations.

Typical Cogan's syndrome is defined using Cogan's original criteria with the following three conditions: (1) ocular symptoms, non-syphilitic interstitial keratitis (IK); (2) audiovestibular symptoms similar to those of Meniere's syndrome (sudden onset of tinnitus and vertigo, accompanied by gradual hearing loss); and (3) an interval between the onset of ocular and audiovestibular manifestations of less than 2years[3].

In addition to ocular and vestibuloauditory dysfunctions, approximately 70% of patients have underlying systemic disease for which vasculitis is considered the pathological mechanism[3].

The audio-vestibular manifestations of Cogan's syndrome are similar to those of Ménière's syndrome, with sudden onset of vertigo, instability, nausea, vomiting and tinnitus. Generally, the vestibular manifestations are secondarily associated with an often severe hearing loss, leading to deafness. The vestibuloauditory dysfunction is usually bilateral, presenting with tinnitus, sensorineural hearing loss, and acute episodes of vertigo [5,8].

The pathophysiology is not yet clearly yet, but an immune reaction directed against the constituents of the cornea is suspected. Ocular manifestations simulate retinal vasculitis[11].

The variability of the clinical presentation, the diagnosis is complex and usually late, and requires the exclusion of other diseases. It is based on the presence of characteristic inflammatory ocular disease and vestibular-auditory dysfunction, in the presence or absence of vasculitis[6].

No treatment has proven very effective in CS. Corticosteroids often have good results on ocular, vascular, and visceral signs, but the effect on audiovestibular symptoms is not so effective, especially once deafness is present. Anterior ocular inflammation, such as interstitial keratitis anterior uveitis, scleritis, and episcleritis, usually responds to topical corticosteroids.

Topical or systemic nonsteroidal anti-inflammatory drugs may be useful in patients with episcleritis and scleritis. Rarely, anterior inflammation may require systemic corticosteroid therapy. Posterior ocular involvement requires treatment with systemic corticosteroids, starting with a dose of prednisone of 1 mg/kg/day and slowly tapering.

In cases of treatment failure or corticosteroid-sparing therapy, other immunosuppressive drugs can be used, such as cyclophosphamide, azathioprine, methotrexate, cyclosporine and tumour necrosis factor-alpha blockers. [7,9]. Given the rarity of this disorder, there are no classification criteria as yet hence the need to case reporting. Moreover, there is no clear guidance on the duration of immunosuppressive treatment of this condition.

As for the biological drugs, infliximab seems to be the most effective of the anti-TNFs for the induction and maintenance therapy of the CS resistant to glucocorticoids and immunosuppressant's being more effective if it is introduced early and etanercept increases the word recognition in hearing impairment[10].

Conclusion

Cogan syndrome is a rare disorder and can be difficult to diagnose in particular in children. The diagnosis is essentially clinical including the presence of audio-vestibular symptoms and interstitial keratitis along with response to immunosuppressive medication.

The consistent clinical picture supported by the positive effect to glucocorticoids especially in the absence of an alternative diagnosis would make the diagnosis of Cogan syndrome likely.

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