Cogan's Syndrome: A Rare Atypical Presentation

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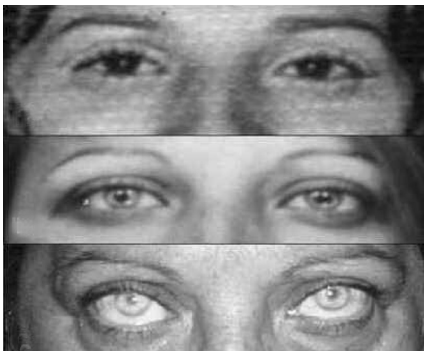
Cogan's syndrome is a rare disorder, typically defined by non-syphilitic interstitial keratitis associated with audiovestibular symptoms characterized by abrupt onset of vertigo and tinnitus reminiscent of Meniere's disease, with rapid development of bilateral deafness [1]. The interval separating ocular and audiovestibular symptoms usually ranges from 1 to 6 months, but may be as long as 2 years [1]. Cogan's syndrome becomes atypical when the eye and/or ear involvement is of a different type or when the interval separating their appearance exceeds 2 years [2,3]. The syndrome affects primarily young adults, involves an additional organ in two out of three cases, and presents a clinical picture of systemic disease reminiscent of vasculitis in one patient out of three [2,3]. The commonest symptoms are cardiovascular, musculoskeletal, neurologic, gastrointestinal, and mucocutaneous. No biological test for confirmation of the diagnosis is currently available. The etiology and pathogenesis remain obscure. Prognosis is dominated by the risk of definitive deafness and by cardiovascular complications [4,5]. Treatment consists mainly of corticosteroids. The ocular symptoms usually regress but deafness is only rarely reversible. Immunosuppressive agents have been recommended, especially when there is associated systemic vasculitis [4,5].

We describe a patient with a rare atypical presentation of Cogan's syndrome characterized by a very long interval between the ocular and the audiovestibular involvement.

Patient Description

A 50 year old woman was referred to our department because of sudden sensorineural hearing loss in her right ear that had developed over 5 days. On admission she reported that during the previous 5 years she had suffered from frequent (monthly) attacks of left high pitched tinnitus and vertigo spells with progressive fluctuating SNHL, and that in the year prior to her admission these attacks had involved the right ear as well. She had been diagnosed in the community clinic as suffering from Meniere's disease and treated accordingly with diuretics, low salt diet, and other non-steroidal medications, but with no improvement. When the patient was asked about other medical problems, she reported having suffered for 20 years from recurrent infections in both eyes, which were treated locally with various types of eyedrops with temporary relief.

Over the last few years her eyes problem subsided and she had less severe inflammations, but her eyes had developed a strange pale blue color. Examination of the head and neck, including bedside vestibular evaluation, disclosed no other remarkable findings. Audiometry showed bilateral asymmetric SNHL with pure tone audiometry values (average for frequencies of 500, 1000, 2000 and 4000 Hz) of 51 dB in the right ear and 91 dB in the left ear. Photographs taken at different ages showed that her eyes, originally dark in color, had faded over the years [Figure]. Ophthalmologic examination showed atypical interstitial keratitis with bilateral corneal stromal cloudiness without pupillary involvement. Taking into account the progressive SNHL, the vertigo and the ophthalmologic disease, oculovestibulo-auditory syndrome (Cogan's syndrome) was suspected. The change in eye color was presumed to be a result of the chronic eye inflammation with scarring of the cornea. Treatment with prednisone (1 mg/kg/day) elicited a marked clinical response. The ocular symptoms resolved immediately, and repeated audiometry showed significant improvement in her right ear with pure tone audiometry of 40 dB but little improvement in the left ear. Steroid dosages were tapered gradually and immurane treatment (100 mg/day) was added. Maintenance treatment included prednisone (5 mg/d) with methotrexate (15 mg/7 days). Follow-up for 3 years has shown stable hearing with no vestibular or ocular complaints.



Photographs of the patient taken at different ages showing progressive fading of eye color. Note the strange pale color of the eyes.

SNHL = sensorineural hearing loss

Comment

The main ophthalmologic symptoms of Cogan's syndrome reportedly include ocular redness, photophobia with tearing, ocular pain, and transitory diminution of visual acuity [1,2]. According to Vollersten et al. [2] and Chynn and Jakobiec [5], the interstitial keratitis involves mainly the anterior and middle aspects of the peripheral corneal stroma near the limbus. Vollersten et al. [2] also found that

60–80% of patients progress to irreversible bilateral deafness. Rabinovitch et al. [4] and Vollersten et al. [2] reported that in the majority of patients such progression occurs within the first 3 years after disease onset. Although the cause of Cogan's syndrome remains obscure, numerous observations support an infectious or autoimmune mechanism. Many patients have an antecedent upper respiratory tract infection or were vaccinated. It has been suggested that this may sensitize the immune system to cross-react against antigens in the cornea and audiovestibular system [5]. No treatment has demonstrated clear-cut efficacy when the auditory involvement is resistant to corticosteroids. As an alternative, and because of frequent relapses, ongoing treatment with immunosuppressants might be needed for many years.

Our patient presented a rare, atypical form of Cogan's syndrome with a very long interval (more than 15 years) between the otologic and the ocular manifestations, unusual corneal cloudiness, no pupillary involvement, and complete change of eye color. Her symptoms responded well to combined steroidal and immunosuppressive therapy. In conclusion, because of its extremely variable clinical course, diagnosis of Cogan's syndrome demands a high level of awareness and suspicion of ocular inflammation in any patient presenting with vestibuloauditory symptoms. Correct diagnosis and prompt treatment are critical for a favorable prognosis.

References

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