

Cogan's Syndrome: Atypical Ophthalmic Manifestations

Case Report

Abstract

The purpose of this report is to describe a case of Cogan's syndrome (CS) with atypical ophthalmic features, such as diffuse anterior scleritis and bilateral peripheral ulcerative keratitis. The article also discusses the clinical aspects, treatment and prognosis of this rare disease. A 66-year-old woman presented with bilateral a symmetric diffuse anterior scleritis and peripheral ulcerative keratitis. Two years later, the patient complained of hearing loss and the audiometry showed sensorineural hearing impairment. The ancillary tests for infectious or autoimmune diseases were unremarkable, leading to the diagnosis of atypical CS. Therefore, the relevance of this article is to emphasize that CS should be considered in patients with recurrent eye inflammation and audiovestibular symptoms. The peripheral ulcerative keratitis is an atypical finding that has not been previously described related to CS. The diagnosis of the syndrome in the early stages is important for appropriate therapy and prevention of hearing impairment and permanent visual damage.

Keywords: Cogan's syndrome; Scleritis; Keratitis, Ulcerative; Hearing loss, Sensorineural

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Abbreviations: CS: Cogan's Syndrome; OD: Right Eye; OS: Left Eye; OU: Both Eye; PUK: Peripheral Ulcerative Keratitis

Introduction

Cogan's syndrome (CS) is a rare inflammatory disease of unknown etiology that affects young adults and is characterized by recurrent eye inflammation, sensorineural deafness and audiovestibular symptoms. It's typical form presents as non-syphilitic interstitial keratitis associated with audiovestibular dysfunction. In the atypical form, other ocular lesions have been described, such as episcleritis, scleritis, retinitis, glaucoma, optic neuritis and papilledema [1]. This case report describes a 66 year-old woman with CS and atypical ophthalmic manifestations.

Case Presentation

A 66-year-old woman presented with pain and redness in both eyes (OU) and did not report systemic symptoms. The examination revealed best corrected visual acuity of 20/25 in the right eye (OD) and 20/30 in the left eye (OS). The ocular inspection showed diffuse anterior scleritis and the biomicroscopy revealed peripheral ulcerative keratitis (PUK) with asymmetric severity (Figures 1 & 2). The intraocular pressure and the fundus examination were unremarkable in OU. Multidisciplinary evaluation was performed and the ancillary tests excluded infections and autoimmune diseases.

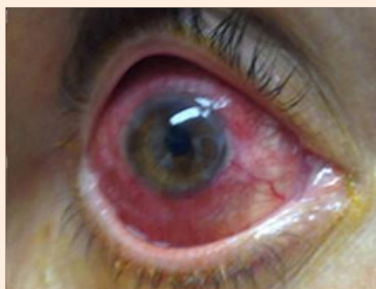


Figure 1: OD: Diffuse Anterior Scleritis

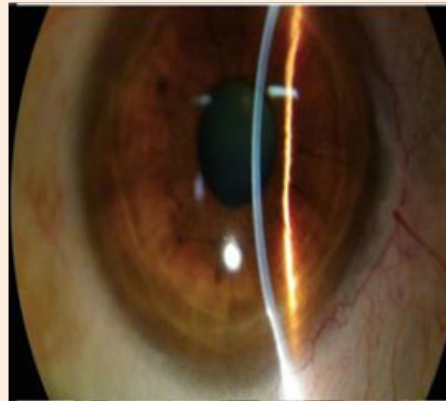


Figure 2: OD: Peripheral Ulcerative keratitis.

The patient was treated with eye drops of dexamethasone (1 mg / ml), ocular lubricants, topical and systemic non-steroidal anti-inflammatory medication with partial improvement. After recurrent episodes, pulse therapy and methotrexate were performed to control the disease. Two years later, the patient presented with hearing impairment. She did audiometry that showed bilateral sensorineural hearing loss leading to the suspicious of the diagnosis of Cogan's syndrome.

Discussion

Cogan's syndrome is a rare disease that affects mainly caucasian young adults with no sex predominance [2]. The etiology is not well defined. It is believed that Cogan's syndrome is related to an autoimmune mechanism and therefore it is possibly associated with other autoimmune diseases, such as Wegener's granulomatosis, polyarteritis nodosa and rheumatoid arthritis [3]. The typical ocular signs and symptoms are pain, redness, blurred vision and non-syphilitic interstitial keratitis [1]. However, the patient can present with atypical ophthalmic

features. Conjunctivitis, episcleritis, scleritis, uveitis, papilledema and retinal vasculitis have already been described. Ophthalmic manifestations can precede the audiovestibular symptoms by months to years, delaying the diagnosis of the disease. Atypical CS tends to be more aggressive and has a worse prognosis [1].

In the typical form, the gap between onset of ocular manifestations and the sensorineural hearing loss is usually less than 2 years, while in the atypical form manifests after more than two years [4]. The vestibular symptoms have a sudden onset, characterized by vertigo, tinnitus, imbalance, nausea and vomiting, and may be the first symptoms of the CS. The sensorineural hearing loss is usually bilateral, which progresses to severe and irreversible deafness in most of the cases, which affects the quality of life [1]. The systemic manifestations of Cogan's syndrome are fever, headache, myalgia, arthralgia, arthritis, gastrointestinal symptoms and skin lesions such as rash, vasculitis and vitiligo. Cardiovascular and genitor-urinary involvements, splenomegaly and lymphadenopathy have also been reported [1,5].

Cogan's syndrome is a diagnosis of exclusion, since there are not laboratory tests to the diagnosis of the syndrome. Therefore, it is necessary to exclude other causes of eye disease, such as infectious and autoimmune diseases [6]. In the present report, as a confounding factor, the hearing impairment could be related to the age of the patient and raise the question about whether this is in fact atypical CS; however the audiometric pattern is not characteristic of presbycusis and the patient denied exposure to harmful noise. The treatment is performed with immunosuppression. The use of prednisone, methotrexate and cyclophosphamide has already been reported and the patients should be regularly monitored for adverse effects of these drugs [7].

Summary

In summary, the multidisciplinary evaluation plays an important role for the early diagnosis of CS. Time between onset of signs and symptoms and the diagnosis directly influences the prognosis and the quality of life of the patient. The peripheral ulcerative keratitis described in this report is an atypical finding that has not been previously described related to CS.

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