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Case Report

Atypical Cogan's syndrome: A case report and summary of current treatment options

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ABSTRACT

Cogan's syndrome is a rare chronic vasculitis, characterized in its typical form by progressive sensorineural bilateral hearing loss, vestibular symptoms and non-syphilitic interstitial keratitis. Only a few cases have been reported in children, most of whom have been diagnosed with the typical form. Early diagnosis and treatment are crucial to ensure a favorable prognosis. Systemic treatment usually begins with high dosage corticosteroids. In case the initial treatment fails, other immunosuppressive drugs are used (cyclophosphamide, methotrexate, cyclosporine A and azathioprine). Additional treatment possibilities, such as plasmapheresis, TNF-alpha blockers (etanercept and infliximab), rituximab, tocilizumab and mycophenolate mofetil have been described over the past few years.

1. Introduction

Cogan's syndrome (CS) was first described by the American ophthalmologist David C. Cogan in 1945. CS is a rare autoimmune vasculitis characterized by non-syphylitic interstitial keratitis and bilateral cochlear and vestibular dysfunction [1–4]. In 1980, Haynes et al. defined the atypical form of Cogan's syndrome as including audiovestibular symptoms and accompanying intraocular lesions, such as episcleritis, scleritis, iritis, conjunctivities, vitritis, retinitis, choroiditis and optical neuritis [3]. CS is a rare condition and its incidence remains unknown. It is extremely rare in children, with only a few cases of the atypical form described in the literature [2]. Without treatment, profound hearing loss develops in a matter of weeks or months in most patients [1,3]. In children, slightly better outcomes have been observed [2].

The case of a 16 year old female with the atypical form of the disease is presented and new therapeutic approaches are discussed.

2. Case report

A sixteen year old female presenting with rotational vertigo lasting for 1 h without nausea sought treatment at the ENT clinic. Clinical examination of the audiovestibular system showed no pathology, and no hearing loss was found. The patient was examined by a neurologist the same day, with a negative result. Dehydration and stress were considered to be the causes of vertigo. One week previously, the patient had been treated for episcleritis with steroid eye drops.

A week after this treatment, rotational vertigo with nausea, vomiting and bilateral hearing loss appeared. There was no sign of nystagmus during the clinical examination and audiological tests revealed bilateral sensorineural hearing loss, mild on the left side and severe on the right side (Fig. 1). Systemic corticosteroids (methylprednisolon 250 mg/day), with vasodilatant drugs (vinpocetinum 20 mg/day, pentoxifylin 100 mg/day and betahistin 48 mg/day) were administered, and a 10 day course of hyperbaric oxygen therapy was completed as well. After 10 days of receiving this therapy, there was significant improvement. Vertigo and episcleritis disappeared completely, but mild sensorineural hearing loss at high frequencies in both ears remained.

One day after completing the therapy, deterioration of hearing loss to the pretreatment level was observed and the likelihood of systemic or autoimmune disease, including Cogan's syndrome,

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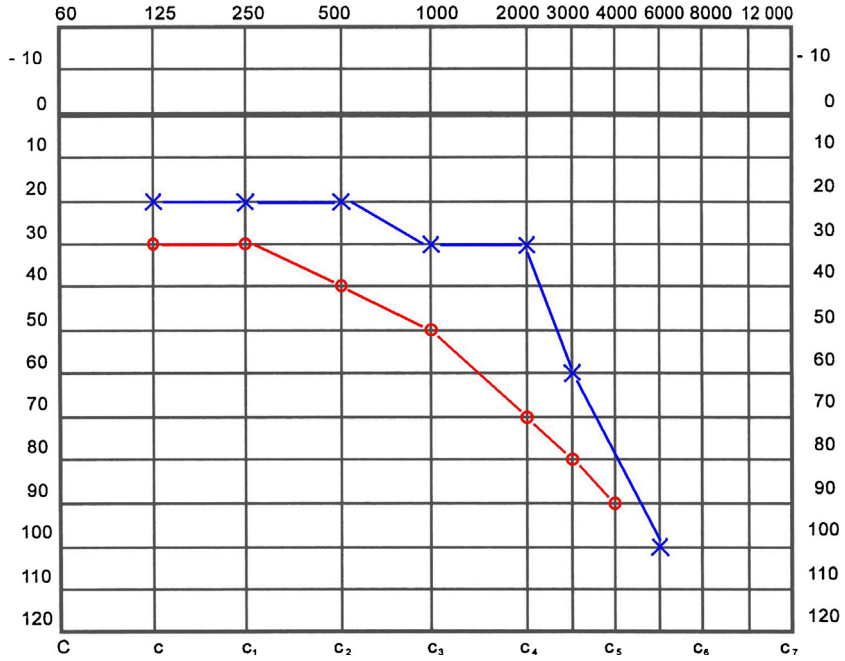

Fig. 1. Initial audiological test. Bilateral sensorineural hearing loss, mild on the left side and severe on the right side.

was considered. Detailed examination (chest X-ray, abdominal ultrasound, brain MRI, PET/CT, serology and immunology tests) revealed no pathology. The diagnosis of atypical Cogan's syndrome was arrived at the basis of clinical presentation. The treatment with systemic corticosteroids (methylprednisolon 250 mg/day) was re-administered for 3 days, with consequent progressive decrease of the dosage to 8 mg of methylprednisolon per day. Despite continuation of this treatment (8 mg of methylprednisolon per day), after initial improvement, deterioration of hearing occurred again after 3 months. Therefore azathioprine (50 mg) was added to the treatment regimen. Despite this combined immunosuppressive therapy the patient developed bilaterally severe sensorineural hearing loss (Fig. 2) 6 months after the initial presentation.

3. Discussion

The diagnosis of CS is often delayed or missed because of extensive differential diagnosis of ocular and audiovestibular manifestations. Moreover, there are still no specific tests and no diagnostic criteria. Lately, anti-Hsp70 antibodies have been suggested as potential markers of typical CS [5]. Nevertheless, their relevance remains to be confirmed and they seem to be of no significance in the diagnosis of the atypical form of CS. Thus, the

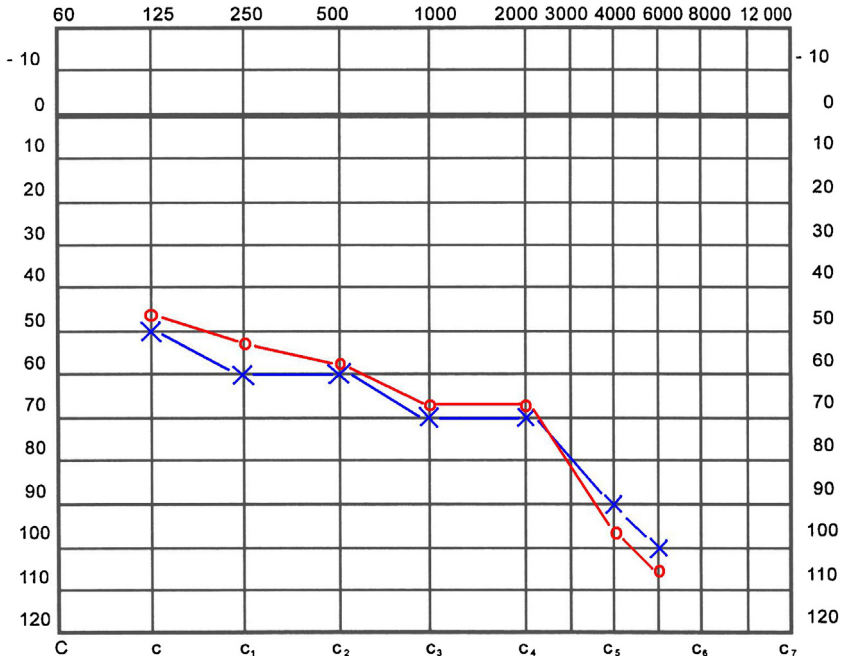

Fig. 2. Audiological test after 6 months. Bilaterally severe sensorineural hearing loss.