



Atypical presentation of Cogan's syndrome with granulomatous anterior uveitis - A case report

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General Note

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ABSTRACT

Introduction: Cogan's syndrome is a scarce inflammatory disorder that mostly impacts the visual and audiovestibular (AV) function. There are two common classifications of Cogan's syndrome (CS), typical and atypical CS. The ordinary ocular presentation of atypical CS is non-syphilitic interstitial keratitis that may be associated with other ocular inflammation which includes non-granulomatous anterior uveitis (NAU). *Case report:* A case of atypical CS with bilateral granulomatous anterior uveitis (GAU) was described. Eighteen years old male patient was referred to the Eye Clinic, University Hospital Bretonneau, Tours, France, complaining of blurred vision in the last 3 weeks in both eyes. For the last 5 months, the patient suffered a bilateral severe progressive hearing loss, tinnitus, and vertigo in addition to repeated episodes of ocular redness. The visual acuity was 0.20 and 0.10 Log MAR in the right and left eye, respectively. Biomicroscopic examination showed GAU, the anterior chamber had +3 inflammatory cell and granulomatous infiltration of the iridocorneal angle with granulomatous keratic precipitate in both eyes. The intraocular pressure and dilated fundus examination were normal in the right and left eyes. The patient was diagnosed as an atypical CS case with GAU. The patient was

treated with intravenous methylprednisolone followed by oral prednisolone in addition to infliximab, topical steroid, and cycloplegic eye drops. The redness had relieved and the anterior uveitis resolved. However, there was no improvement in hearing, cochlear implants planned in a month. *Conclusion:* To the best of our knowledge, this is the first case of atypical CS with GAU without any systemic involvement.

Keywords: Cogan's syndrome, granulomatous anterior uveitis, audiovestibular disturbances

1. INTRODUCTION

Cogan's syndrome (CS) is an idiopathic scarce autoimmune chronic inflammatory disorder (Adriana *et al.*, 2015; Karpecki, 2015). Earlier, the syndrome was described as an association of non-syphilitic interstitial keratitis (NSIK) and the occurrence of vestibuloauditory (VA) disturbances (Morgan and Baumgartner, 1934; Espinoza *et al.*, 2020). It was later defined by David Cogan as an NSIK and Meniere-like VA dysfunction (Cogan, 1945; Espinoza *et al.*, 2020). Haynes *et al.*, (1980) suggested further categorization of CS as typical and atypical subtypes. Typical CS is distinguished by NSIK and Meniere-like VA dysfunction. The interval between the onset of AV dysfunction and ocular symptoms occurs in almost 2 years. Atypical CS is characterized by episcleritis, scleritis, uveitis, conjunctivitis, retinal vasculitis, optic neuritis and papilledema. These symptoms may be associated with NSIK. The AV dysfunction associated with atypical CS does not resemble typical Meniere's disease. The interval between the onset of AV dysfunction and ocular manifestations occurs within more than 2 years (Haynes *et al.*, 1980; Vinceneux, 2005). In this report, a case of atypical CS with bilateral granulomatous anterior uveitis (GAU) was recorded.

2. CASE REPORT

Eighteen years old male patient was referred to the Eye Clinic, University Hospital Bretonneau, Tours, France, with 3 weeks history of reduced visual acuity, pain, and redness in both eyes. In the last five months, he also complained of severe progressive hearing loss, vertigo, and tinnitus. The patient suffered repeated attacks of ocular redness in the last year which had been treated with topical corticosteroids, with the improvement of ocular symptoms. On biomicroscopic examination, the best-corrected visual acuity was 0.20 LogMAR in the right eye and 0.10 LogMAR in the left eye. The anterior segment examination revealed GAU; the anterior chamber had +3 inflammatory cells with granulomatous infiltration of the iridocorneal angle in both eyes. The examination also showed the appearance of granulomatous keratic precipitate. Posterior synechia, intraocular pressure was 16 mmHg in the left eye and 15 mmHg in the right eye. The dilated fundus examination was unremarkable in both eyes. Optical Coherence Tomography, Fundus Fluorescein Angiography, and Indocyanine Green Angiography were normal.

Multidisciplinary evaluation and ancillary tests were performed to exclude infections, granulomatous diseases, and autoimmune diseases. The baseline blood investigations were also normal. The infectious and immunological panel revealed cytomegalovirus IgG-positive and antinuclear antibody positive. The cerebrospinal fluid analysis was negative for all microbial and serologic tests. Whole-body positron emission tomography scan was performed and was normal as well. The audiogram revealed a severe sensorineural hearing loss in both ears, while cerebral magnetic resonance imaging showed bilateral labyrinthitis with progressive extinction of the fluid in the inner ears. The patient was diagnosed as an atypical CS case with GUA. He was hospitalized, and treatment modality was initiated with a daily loading dose of intravenous cortisone (1 g) for 3 days, followed by oral daily cortisone therapy (1 mg/kg) in a tapering regimen (figure 1). Additionally, the patient was treated with intravenous (5mg/kg) infliximab at weeks 0, 2, and 4 followed by a single dose every 8 weeks. Topical steroid and cycloplegic drops were included in the treatment regimen. Four weeks later, the visual acuity improved to 0.00 LogMAR and the anterior uveitis resolved. However, there was no improvement in hearing, cochlear implants planned in a month. Informed consent from the patient was assigned.

3. DISCUSSION

Cogan Syndrome is a scarce disorder of unknown etiology. The diagnosis of CS is established by the clinical associations of VA dysfunction and ocular inflammation as there are no laboratory assays to confirm CS diagnosis. Consequently, the other etiology and comorbidities must be excluded (Barbosa *et al.*, 2014). The following ocular involvement is classified as atypical CS: conjunctivitis, subconjunctival hemorrhage, choroiditis, episcleritis, iritis, scleritis, retinal artery occlusion, exophthalmos, retinal hemorrhage, papilloedema, retinal vasculitis and serpiginous choroidopathy (Johari *et al.*, 2020). Atypical CS is speculated if the ocular inflammation was associated with AV disturbance (not Meniere disease) within more than 2 years (Johari *et al.*, 2020). Atypical

CSis known to be more aggressive and can carry worse prognosis (Barbosa *et al.*, 2014). Seventy percent of Cogan's patients suffer from comorbidity with systemic diseases usually, vasculitis (Adriana *et al.*, 2015).

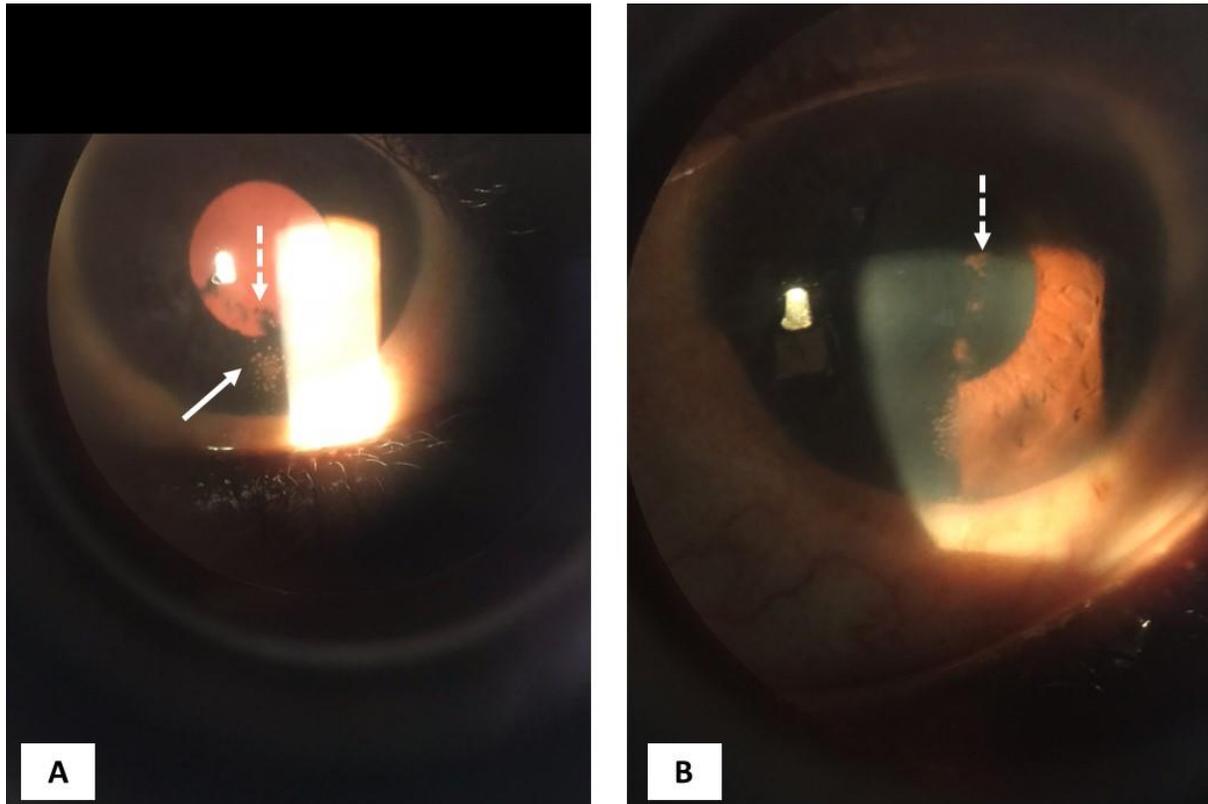


Figure 1 Anterior segment examination of both eyes revealed the presence of granulomatous keratic precipitate (solid arrow) and anterior capsular pigment following posterior synechiolysis (dotted arrows). A: Right eye; B: Left eye.

Early clinical diagnosis and application of proper treatment modality are crucial to prevent severe irreversible hearing loss (Queirós *et al.*, 2013). No clear guidelines are available for the treatment of CS, nevertheless, treatments based on disease incidence and intensity. Anterior segment inflammations such as NSIK, anterior uveitis, and episcleritis may respond well to corticosteroid eye drops, while posterior segment inflammations would require intravenous steroid therapy (1mg /kg/day). The indication of adding an immunosuppressive drug to treatment regimen includes failure of treatment within 15-21days, failure to reducing the prednisolone to 10 mg/day, and relapse of ocular inflammation and/or auditory symptoms when decreasing the dose of steroids. Many immunosuppressive agents can be used including, cyclosporine, methotrexate and azathioprine. Infliximab-tumor necrosis factor alpha-blocker produced a significant effect on the induction and maintenance of remission in atypical CS patients (Queirós *et al.*, 2013; Adriana *et al.*, 2015). (Vollertsen *et al.*, 1986; Gluth *et al.*, 2006) reported that granulomatous uveitis is linked with atypical CS, but they did not specify its type. Whereas the French Society of Ophthalmology 2010 confirmed that atypical CS is associated with NGAU. In contrary to all these previous reports, our case report specified that atypical CS was correlated to GAU.

4. CONCLUSION

As far as we know, this is the first recorded case of atypical CS with GAU without any systemic involvement. The diagnosis of CS is very challenging and should be suspected whenever there are clinical associations of VA dysfunction and ocular inflammation as there are no specific serological tests to confirm its diagnosis.

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Author Contributions

Heba Joharjy: Data curation, and writing—original draft preparation.

Pierre-Jean Pisella:	Supervision.
David Bakhos:	ENT examination.
Alexandra Audemard:	Systemic evaluation.
Marie Laure Le Lez:	Conceptualization, diagnosis, writing—review and editing.

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Conflict of interest

The authors report no conflict of interest.

List of Abbreviations

AV: Audiovestibular; CS: Cogan's syndrome; NAU: Non-granulomatous anterior uveitis; GAU: granulomatous anterior uveitis; NSIK: Non-syphilitic interstitial keratitis, VA: vestibulo auditory disturbances.

Data and materials availability

All data associated with this study are present in the paper.

Peer-review

External peer-review was done through double-blind method.

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