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Atypical Cogan's Syndrome: Case Report of an Oculoaudiovestibular Disease

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Abstract Cogan's syndrome (CS) is a rare autoimmune vasculitis. Clinical hallmarks are bilateral interstitial keratitis and vestibuloauditory dysfunction. Association between Cogan's syndrome and systemic vasculitis as well as aortitis also exists. We report a case of cogan's syndrome in 12 year old girl who intialy present with bilateral interstitial keratitis with audiovestibular and cardiac symptoms. We discuss the clinical aspect, the pathogenic mechanisms, the laboratory investigations, the differential diagnosis and traitement of cogan syndrome.

Keywords: *cogan's syndrome, interstitial keratitis, deafness*

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1. Introduction

Cogan's syndrome (CS) is a rare autoimmune vasculitis. Clinical hallmarks are bilateral interstitial keratitis and vestibuloauditory dysfunction. Association between CS and systemic vasculitis also exists. Its etiology includes infection and autoimmunity. Vasculitis is considered the pathological mechanism. We report a case of a atypical Cogan's syndrome, the workup of the diagnosis, and treatment results.

2. Case Report

We report a case of a 12-year-old girl with medical history of vestibular symptoms including vertigo and imbalance associated with nausea, vomiting and musculoskeletal pains, one year ago. The patient complained redness in the left eye with diplopia, photophobia and developed acute onset of right severe sensorineural hearing loss with tinnitus. She was evaluated in the emergency department and admitted to rule out any cerebral vascular accident.

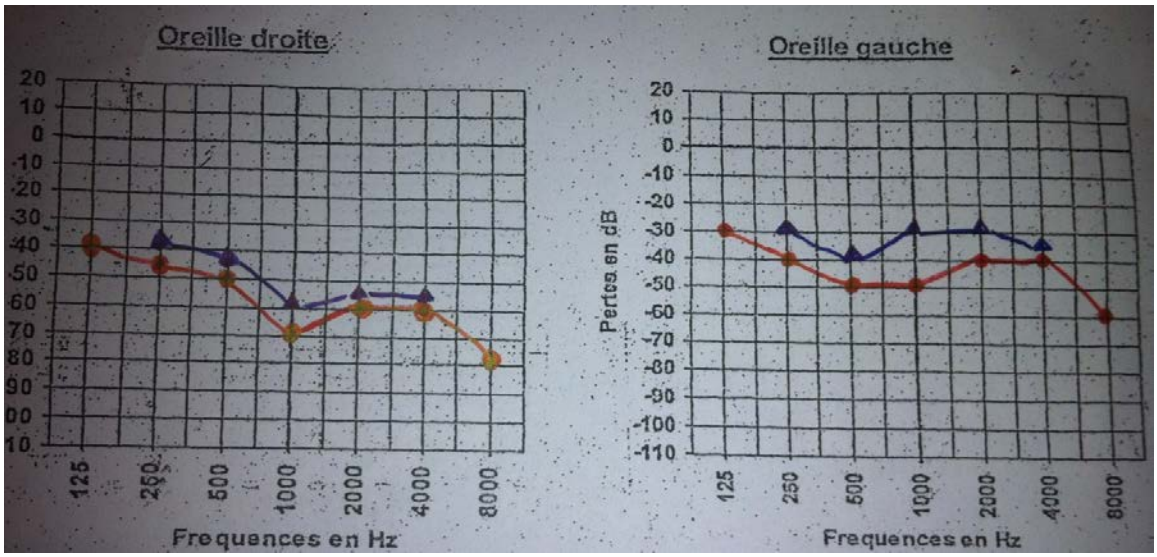

Figure 1. Audiogram showing a severe right sensorineural hearing loss, and moderate left sensorineural hearing loss

Otolaryngology consultation was obtained identifying a normal otoscopic examination. Her audiogram (Figure 1) showed a severe right sensorineural hearing loss, and moderate left sensorineural hearing loss. Ophthalmology consultation showed a bilateral interstitial keratitis, with ghost ships and ferritin deposits. (Figure 2). On physical examination, his blood pressure is 138/66 mm Hg. Cardiac auscultation revealed a murmur of mitral regurgitation. Standard electrocardiography (ECG) was performed, followed shortly by transthoracic echocardiography, which confirms moderate mitral regurgitation (Figure 3). Which has required tracking cardiology.

Figure 2. interstitial keratitis, with ghost ships and ferritin deposits

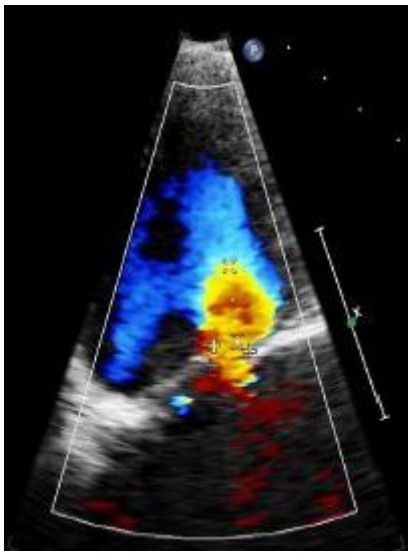

Figure 3. Assessment of te severity of mitral insufficiency

The patient was referred to a rheumatologist for additional autoimmune workup. Blood test results were negative for antinuclear antibody, antinuclear cytoplasmic antibody, rheumatoid factor, rapid plasma reagin, and HLA-B27. The hepatitis serology finding was negative. Erythrocytes sedimentation rate was elevated at 50, and C-reactive protein was mildly elevated at 6.

Computed tomography (CT) of petrous bone was normal; the chest computed tomography finding was negative for any lymphadenopathy or other lesions and the cerebral magnetic resonance imaging (MRI) was normal.

Based on audiovestibular, ocular and cardiac findings and given the patient's clinical history with negative laboratory and imaging test results, which allows the exclusion of infectious, neoplastic, granulomatous,and autoimmune etiologies, the diagnosis of atypical Cogan's syndrome was established.

It was decided to start a pulse of intravenous methylprednisolone (0,5 g/day for 3 days) followed by oral prednisolone 1mg/kg/day in a tapering regimen. 2 weeks later, her general condition stabilized, anterior eye segment changes improved.

Four months later, systemic prednisone was gradually reduced to 0,5mg/kg/day, but clinical follow-up showed a reccurence of audiovestibular symptom which has required the introduction of methotrexate for 7 months, during the following six months, systemic prednisone was gradually reduced, methotrexate was discontinued. In this case, immunosuppressive therapy failed to improve the patient’s hearing of the left ear.

Clinical follow-up showed a total right deafness at 3 years after the onset of her symptoms, hearing in her left ear remains normal. Mitral insufficiency was controlled with surgical replacement of the mitral valve. Currently the patient is candidate for corneal transplant as soon as possible in order to start the treatment of amblyopia.

3. Discussion

Cogan's syndrome is a rare autoimmune vasculitis characterised by ocular and vestibuloauditory dysfunctions, and often by systemic disease as well. It was first described in 1945 by an ophthalmologist, Dr David G. Cogan, who reported on a “syndrome of nonsyphilitic interstitial keratitis (IK) and vestibuloauditory symptoms” that resembled Meniere's disease [1,2]. Cogan's syndrome occurs equally in both sexes, in patients from 2 to 60 years of age. The average age of onset is 25 years of age and there is no gender-specific prevalence [1,2].

The variability of ocular and audiovestibular clinical manifestations complicates its diagnosis, which should be suspected whenever there is a close temporal association between ocular abnormalities and cochleovestibular

symptoms. Signs of inner ear dysfunction include nausea, vomiting, tinnitus, vertigo, and bilateral hearing loss. Deafness generally progresses rapidly during a 1- to 3-month period. Because the early auditory symptoms of Cogan's syndrome vary, differential diagnosis can be difficult [1,3,4].

Cogan's syndrome can be confused with idiopathic Meniere's disease or neurosyphylis. The audiogram is always abnormal but involvement is often asymmetrical. Ocular disease presents with redness, photophobia, watering, and reduced vision. Examination reveals ciliary injection with mild iritis and discrete fluffy opacities in the deep portion of the corneal stroma. Corneal ulceration does not occur and vascularisation is now unusual. A histological study of the ocular lesions showed lymphocytic and plasma cell infiltration in the cornea with neovascularisation but no vasculitis [1,4,5].

In addition to ocular and vestibuloauditory dysfunctions, approximately 70% of patients have underlying systemic disease for which vasculitis is considered the pathological mechanism [1,4]. Numerous systemic manifestations were reported in 1960 by Cody and Williams in patients with Cogan's syndrome [3]. The most common symptoms are cardiovascular, neurological and gastrointestinal [1,4]. The most characteristic cardiovascular manifestation of Cogan's syndrome is aortitis with aortic insufficiency [1,4,5].

Neurological manifestations may include hemiparesis or hemiplegia due to a cerebral vascular accident and aphasia due to a transient ischaemic event. Various gastrointestinal manifestations have been reported, including diarrhoea, melena and abdominal pain, sometimes related to mesenteric arteritis [1,4,6].

The clinical diagnosis is based on audiovestibular symptoms, ocular inflammation and nonreactive serologic tests for syphilis in the presence of histologically proven vasculitis [1].

Due to the variable onset of symptoms and the lack of specific laboratory tests, the diagnosis of Cogan's syndrome is a challenge and is often based upon a good response to corticosteroid treatment [1,2,4].

According to the criteria of Haynes et al. [6], patients with any of the following symptoms are classified as having atypical Cogan's syndrome: (1) inflammatory ocular manifestations, with or without IK; (2) typical ocular manifestations associated with audiovestibular symptoms different from Meniere-like episodes; or (3) a delay of more than 2 years between the onset of typical ocular and audiovestibular manifestations.

Radiographic studies, such as cranial computed tomography (CT) and magnetic resonance imaging (MRI) are often normal [7], though some authors have reported the presence of labyrinthine aspecific radiological abnormalities [1,4,6]. MRI scans (gadolinium) show calcification or narrowing and soft tissue obliteration of the vestibular labyrinth and the cochlea [8]. Its etiology includes infection and autoimmunity, Chlamydia psittaci has been isolated from a patient with Cogan's syndrome and serological evidence of a recent Chlamydia trachomatis infection was reported in 4 of 13 patients [6]. A decade ago, antibodies directed against a corneal antigen or constituents of the inner ear were detected by multiple groups [9,10]. Recent evidence strongly suggests that Cogan's syndrome is an auto immune disease [1,11]. Histopathological examination of corneal tissue and cochlea shows lymphocytic and plasma cell infiltration, suggesting a cell-mediated reaction. [1,6]. Vasculitis is considered the pathological mechanism.

The histopathologic manifestations appear to explain the audiovestibular dysfunction that has been reported in Cogan's syndrome, including bilateral fluctuating hearing loss, tinnitus, and severe vertigo. [2,4]. Fischer and Hellstrom reported primarily infiltration of the inner ear with lymphocytes, thickening of membranous tissues, endolymphatic hydrops, degeneration of organ of Corti, and atrophic of eighth nerve in temporal bones from a 32-year-old man who was diagnosed with Cogan's syndrome four years prior to death [13].

Zechner also reported a case of Cogan's syndrome in a 54-year-old individual whose hearing loss was not sudden and bilateral. Endolymphatic hydrops, degenerated organ of Corti, fibrosis and ectopic bone formation in the perilymphatic space (primarily in the scala vestibule of the basal cochlear turn were observed [14].

Medical treatment of Cogan's syndrome depends on how extensive the disease is at the time of diagnosis. Corticosteroids are the first line of treatment, and it is argued that they can aid in the recovery of hearing if given early in the disease course. Hearing improves significantly after treatment with intravenous methylprednisolone.

Immunosupressive drugs, such as methotrexate, azathioprine, cyclosporine and cyclophosphamide, have all been used with varying degrees of success. However, in the absence of controlled trials, there are no definitive therapeutic recommendations [1,12]. Patients without systemic disease generally have a good prognosis and an average life expectancy. The corneal disease may resolve without or with therapy; the deafness is often irreversible, although systemic steroids initiated within 2 weeks of the onset of symptoms may limit progression. Patients who develop serious vasculitis, such as aortitis, have an increased risk of death due to complications. Therefore, early assessment and treatment for systemic inflammation are needed to prevent life threatening complications.

4. Conclusion

Cogan's syndrome is a rare presumed autoimmune disorder characterized by nonsyphilitic interstitial keratitis and progressive audiovestibular symptoms and audiovestibular symptoms similar to those of Meniere's syndrome. Atypical CS presents with significant inflammatory eye disease with or without IK; Corticosteroids are the first line of treatment and multiple immunosuppressive drugs have been tried with varying degrees of success. Therefore, early assessment and treatment for systemic inflammation are needed to prevent life threatening complications.

References

[1] Greco A, et al, Cogan's syndrome: An autoimmune inner ear disease, Autoimmun Rev (2012).

[2] Ahad Azami, Nasrollah Maleki, Mohammadreza Kalantar Hormozi, and Zahra Tavosi, “Interstitial Keratitis, Vertigo, and Vasculitis: Typical Cogan’s Syndrome,” Case Reports in Medicine, vol. 2014, Article ID 830831, 4 pages, 2014.

[3] Cody DTR, Williams HL. Cogan's syndrome. Laryngoscope 1960; 70: 447-75.

[4] Kessel A1, Vadasz Z1, Toubi E2. Cogan syndrome-pathogenesis, clinical variants and treatment approaches. Autoimmun Rev. 2014 Apr-May; 13(4-5):351-4.

[5] Ho AC, Roat MI, Venbrux A, Hellmann DB. Cogan's syndrome with refractory aortitis and mesenteric vasculitis. J Rheumatol 1999; 26: 1404-7.

[6] Haynes BF, Kaiser-Kupfer MI,Mason P, Fauci AS. Cogan syndrome: studies in thirteen patients, long-term follow-up, and a review of the literature. Medicine 1980; 59: 426-41.

[7] Gluth MB, Baratz KH, Matteson EL, Driscoll CL. Cogan syndrome: a retrospective review of 60 patients throughout a half century. Mayo Clin Proc 2006; 81: 483-8.

[8] Casselman JW, Majoor MH, Albers FW. MR of the inner ear in patients with Cogan syndrome. AJNR Am J Neuroradiol 1994; 15: 131-8.

[9] Majoor MHJM, Albers FWJ, Van Der Gaag R, Gmelig-Meyling F, Huizing EH. Corneal autoimmunity in Cogan's syndrome? Report of two cases. Ann Otol Rhinol Laryngol 1992; 101: 679-84.

[10] Froehlich F, Fried M, Gonvers JJ, Saraga E, Thorens J, Pecoud A. Association of Crohn's disease and Cogan's syndrome. Dig Dis Sci 1994; 39: 1134-7.

[11] Schuknecht HF. Ear pathology in autoimmune diseases. Adv Otorhinolaryngol 1991; 46: 50-70.

[12] Van Doornum S, McColl G, Walter M, Jennens I, Bhathal P, Wicks IP. Prolonged prodrome, systemic vasculitis, and deafness in Cogan's syndrome. Ann Rheum Dis 2001; 60: 69-71.

[13] Fischer ER, Hellstrom HR: Cogan's syndrome and systemicsystemic vascular disease: analysis of pathologic features with reference to its relationship to thromboangiitis obliterans (Buerger). Arch Pathal 72: 572-592, 1961 6.

[14] Zechner G: Zum Cogan Syndrom. Acta Otolaryngol 89:310-316, 198089: 310-316, 1980.