Fever of unknown origin, bilateral sensorineural hearing loss with canal paresis and uveitis with iridocyclitis and episcleritis: a case of Cogan's syndrome

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DESCRIPTION

A 69-year-old Japanese woman presented with 1-month history of continuous spiking fever, proximal myalgia with weakness, weight loss, intermittent abdominal pain and lower back pain. Except for the use of oral hypoglycaemic drugs to control her diabetes mellitus over the last 7 years, she had been otherwise healthy until she developed these symptoms. Additionally, she had a history of recurrent tinnitus and vertigo for the last 2 years and was suspected to have Ménière's disease or benign paroxysmal positional vertigo by an otolaryngologist. Seven days prior to her first visit, she developed progressively worsening bilateral deafness. She was admitted to the surgical ward to assess intermittent abdominal pain and subsequently underwent gynaecological examination; however, no diagnosis was made. Five days after the admission, she developed progressive bilateral blurred visions and an ophthalmologist diagnosed her with uveitis with iridocyclitis and episcleritis (figure 1A). She was then referred to the Department of Medicine since no unifying diagnosis was achieved. At that time, she began experiencing rotational vertigo with nausea, vomiting and exacerbation of her bilateral hearing loss. The otolaryngologist provided a diagnosis of bilateral sensorineural hearing loss with canal paresis (figure 1B). Laboratory findings revealed elevated C reactive protein levels and erythrocyte sedimentation rate (86 mm/hour). Serological tests for infectious agents including syphilis yielded negative results. Additionally, both serology for autoimmune diseases and coagulation test yielded negative results. She underwent temporal artery biopsy to preclude the possibility of ischaemic arteritis, which revealed diffuse lymphocytic infiltrates without multinucleated cells (figure 1C). Based on all these findings, a diagnosis of Cogan's syndrome was considered. She began glucocorticoid therapy, which resulted in the rapid resolution of the high fever, intermittent abdominal pain and proximal myalgia and the gradual improvement in her vestibular symptoms. However, her visual and hearing losses exhibited little improvement.

Cogan's syndrome is characterised by inflammatory ocular and vestibuloauditory abnormalities. Additionally, approximately 30%–50% of patients with the condition exhibit features of various systemic manifestations including fever, arthralgia, anaemia, neurological disease and gastrointestinal tract disease. The mechanism of the disease is considered as vasculitis involving vessels of all sizes.[1 2] However, due to the variable onset of symptoms and the lack of specific laboratory tests, diagnosing Cogan's syndrome is challenging. Indeed, it is clinical diagnosis chiefly based on audiovestibular symptoms, ocular inflammation and negative serological tests for infectious diseases, including syphilis in the presence of histologically proven vasculitis.[3] Patients with Cogan's syndrome may visit ophthalmologists for ocular symptoms and otolaryngologists for vestibular and auditory symptoms. However, diagnostic delay may occur if vestibular-auditory or ocular symptoms are not carefully observed, and variable systemic manifestations are prominent.[1–3] For this

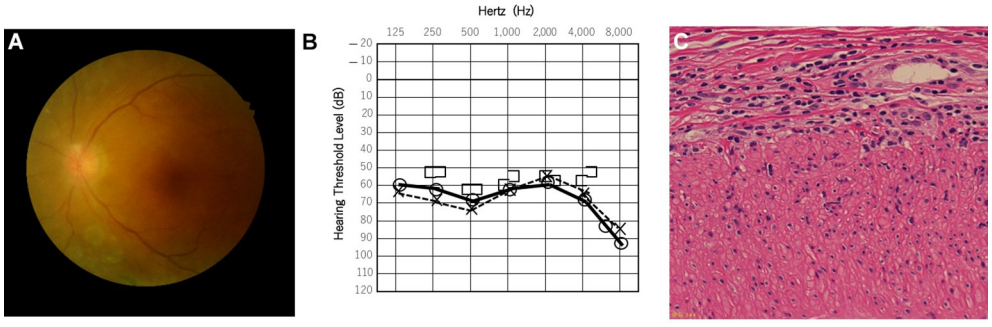

Figure 1 (A and B) Fundoscopic examination and audiogram revealing bilateral severe uveitis with iridocyclitis, episcleritis and bilateral sensorineural hearing loss with canal paresis. (C) Biopsy specimens (H&E staining) from the left temporal artery showing irregularly thickened intima, severe diffuse lymphocytic infiltrates in the arterial wall and focal degenerative change of elastic fibres. No multinucleated giant cells were observed.

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reason, high index of suspicion is considered for a diagnosis of Cogan's syndrome if patients present with fever of unknown origin and inflammatory ocular and bilateral audiovestibular symptoms. Its diagnosis is based on considering all features of patients. Clinicians need to be always mindful of 'not seeing the forest for the trees'.

Patient's perspective

I agree to report this rare disease for medical and clinical improvement. I hope if it is possible to establish a method of treatment in this rare disease in nearly future. (The author's translation)

Learning points

- Cogan's syndrome is an autoimmune inflammatory disease of unknown origin that presents with Ménière's disease-like vestibular symptoms, bilateral sensorineural hearing loss and inflammatory ocular manifestations.
- Due to the variable onset of symptoms and the lack of specific laboratory tests, diagnosing Cogan's syndrome is challenging.
- Clinicians should suspect Cogan's syndrome when patients with fever of unknown origin have inflammatory ocular symptoms and bilateral audiovestibular symptoms.

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