A case of Bilateral Scleritis and Vestibuloauditory Dysfunction: Cogan Syndrome

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Intoduction

Cogan's Syndrome is a rare autoimmune vasculitis with only 250–300 reported cases worldwide.

It is characterized by fluctuating sensorineural hearing loss due to eighth cranial nerve involvement, accompanied by inflammatory ocular manifestations mainly scleritis and keratitis.

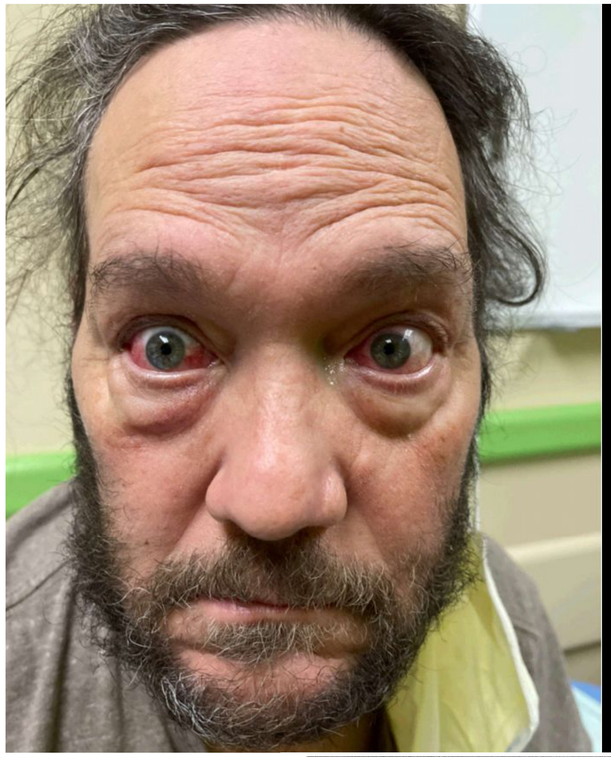
Although its pathophysiology is not fully elucidated, a post-infectious mechanism via molecular mimicry has been proposed.

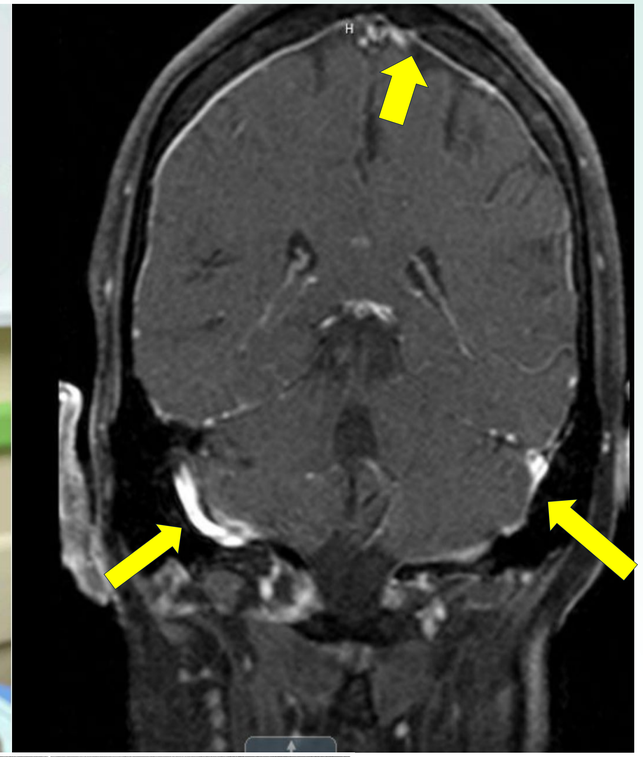
It was described in 1945 by David Cogan, who reported "non-syphilitic interstitial keratitis and vestibuloauditory symptoms."

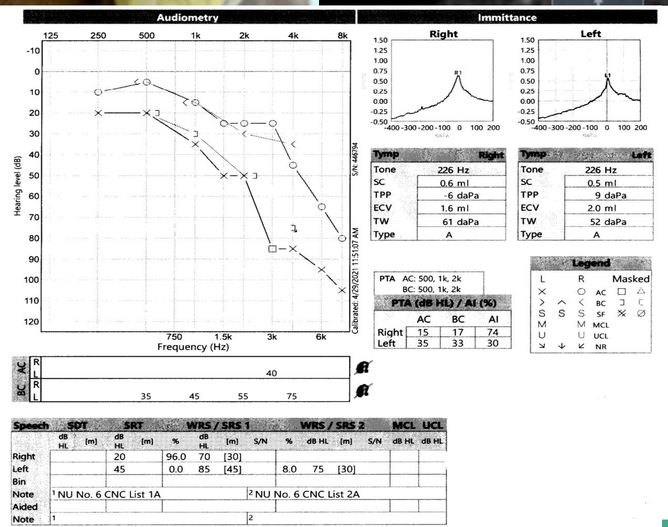
Case Presentation

A 62-year-old man who was initially treated for presumed otitis media and periorbital cellulitis, then presented with headache, vertigo, ataxia, and worsening hearing loss. Computed tomography of the head was unremarkable. Initial brain MRI demonstrated diffuse dural and meningeal enhancement, consistent with pachymeningitis. CSF fluid had WBC of 12 with 36% monocytes; TB,cytology and flow-cytometry were unremarkable. Repeat head MRI/MRV showed diffuse pachymeningeal enhancement, with dural involvement of both internal auditory canals more at the right side. He had rapidly progressive bilateral facial palsy. Audiometry showed sensorineural hearing loss, left more than right. He had non-necrotizing diffuse scleritis on ophthalmology evaluation for conjunctival injection. neurosurgical meningeal biopsy revealed acute and chronic inflammation without granulomas. An initial working diagnosis of neurosarcoidosis was made, and he received high-dose corticosteroids with modest improvement. He was discharged on oral prednisone, later initiated on methotrexate, then adalimumab. Due to hepatotoxicity, Methotrexate was switched to mycophenolate, which resulted in marked clinical improvement. Given the absence of granulomatous inflammation and marked response to mycophenolate, the diagnosis was refined to Cogan's Syndrome. Follow-up MRI demonstrated resolution of dural thickening. As of 09/2025, patient remains stable on adalimumab and mycophenolate, without new flare-ups.

Imaging/Results







Discussion

- This case highlights the difficulty in diagnosing Cogan's syndrome (CS). Like previous reports (1) (2), the absence of specific biomarkers and the overlap with other autoimmune and infectious conditions could delay the diagnosis. Our patient was initially thought to have neurosarcoidosis, reflecting the diagnostic dilemma as in many reported cases where sarcoidosis mimicked CS (4)

- The presence of pachymeningitis and rapidly progressive bilateral cranial nerve involvement is unusual but consistent with systemic and neurological manifestations reported in atypical CS. (1) (5) While ocular and audiovestibular disease are hallmarks, CNS involvement underscores the broad spectrum of the disease.

- Corticosteroids remain the mainstay of therapy, but responses are often incomplete (3) (6). In our patient, methotrexate was limited by hepatotoxicity, yet the introduction of **mycophenolate and adalimumab** resulted in marked improvement. This aligns with recent literature supporting the role of **biological agents** (TNF-α inhibitors, rituximab, mycophenolate, JAK inhibitors) in refractory disease (6).

- Early recognition and aggressive immunosuppression are essential to prevent irreversible damage. Our patient's improvement and sustained stability with biologic and steroid-sparing therapy highlight the value of individualized treatment and long-term follow-up. The resolution of pachymeningeal enhancement on MRI further emphasizes that neurologic involvement in CS, although rare, may be reversible with appropriate therapy.

- This case expands on prior literature by demonstrating pachymeningitis as a manifestation of CS and the effectiveness of **adalimumab plus mycophenolate** in long-term disease control. It underscores the need for multidisciplinary management and careful exclusion of mimicking conditions such as neurosarcoidosis, tuberculosis, and other systemic vasculitides. (4) (5)

References

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