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(CASE REPORT)

Sensorineural hearing loss as an initial manifestation of eosinophilic granulomatosis with polyangiitis: A diagnostic challenge

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Abstract

Background: Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg–Strauss syndrome, is a rare ANCA-associated vasculitis characterized by asthma, hypereosinophilia, and systemic involvement. Otorhinolaryngological manifestations are common, but sensorineural hearing loss as an initial presentation remains rare and poorly described.

Case presentation: We report the case of a 54-year-old woman with long-standing asthma who presented with progressive bilateral sensorineural hearing loss, leading to ENT evaluation. She also reported nasal obstruction, rhinorrhea, and peripheral facial paralysis. Clinical examination revealed bilateral nasal polyposis. Laboratory findings showed marked hypereosinophilia (1950/mm^3), elevated inflammatory markers, and positive C-ANCA. Imaging demonstrated sinonasal polyposis and peripheral pulmonary infiltrates. The diagnosis of EGPA was established based on clinical, biological, and radiological criteria. The patient was treated with systemic corticosteroids and underwent endoscopic sinus surgery, with favorable clinical evolution and stabilization of hearing.

Conclusion: This case highlights that sensorineural hearing loss may represent an early and misleading manifestation of EGPA. Recognition of this atypical presentation is essential for early diagnosis and prompt treatment, in order to prevent irreversible organ damage.

Keywords: EGPA; Churg-Strauss syndrome; Sensorineural hearing loss; Nasal polyposis; Vasculitis; ENT

1. Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg–Strauss syndrome, is a rare necrotizing vasculitis affecting small- to medium-sized vessels and classified among ANCA-associated vasculitides. It is characterized by the association of asthma, peripheral eosinophilia, and systemic organ involvement, including pulmonary, neurological, and cutaneous manifestations [1,2].

Otorhinolaryngological involvement is frequent in EGPA, occurring in up to 70–90% of cases, and typically presents as chronic rhinosinusitis with nasal polyposis [3,4]. However, auditory manifestations are less common and are usually limited to conductive hearing loss secondary to middle ear involvement. In contrast, sensorineural hearing loss is rare and remains poorly described in the literature [5].

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We report a case of EGPA revealed by progressive bilateral sensorineural hearing loss, highlighting the importance of recognizing this atypical presentation. Early identification of such manifestations is essential to avoid diagnostic delay and initiate appropriate treatment, thereby preventing irreversible systemic complications.

2. Case presentation

A 54-year-old woman was referred to the otorhinolaryngology department for evaluation of progressive bilateral hearing loss that had been evolving insidiously over several months. The hearing impairment was gradually worsening and was not associated with tinnitus or vertigo.

Her past medical history was significant for long-standing asthma diagnosed at the age of 18, which had been partially controlled with inhaled therapy. She also had autoimmune thyroiditis and type 2 diabetes mellitus managed with oral antidiabetic agents. There was no history of otologic disease, noise exposure, or ototoxic drug use.

In addition to hearing loss, the patient reported a constellation of symptoms that had progressively appeared over the preceding months. These included chronic bilateral nasal obstruction, persistent anterior rhinorrhea, and a chronic cough. She also described migratory polyarthralgia involving large joints, episodes of palpitations, and general fatigue. Notably, she developed a peripheral facial paralysis, which prompted further medical evaluation.

On physical examination, anterior rhinoscopy revealed bilateral polypoid masses occupying the nasal cavities, consistent with nasal polyposis. There was no evidence of purulent discharge. Otoscopic examination showed normal tympanic membranes without signs of middle ear effusion. Neurological examination confirmed peripheral facial nerve palsy without other cranial nerve deficits. Cardiovascular examination revealed regular tachycardia without murmurs, and the remainder of the systemic examination was unremarkable.

Audiological assessment using pure-tone audiometry demonstrated bilateral sensorineural hearing loss, predominantly affecting the higher frequencies, consistent with cochlear involvement. Speech audiometry confirmed reduced speech discrimination scores.

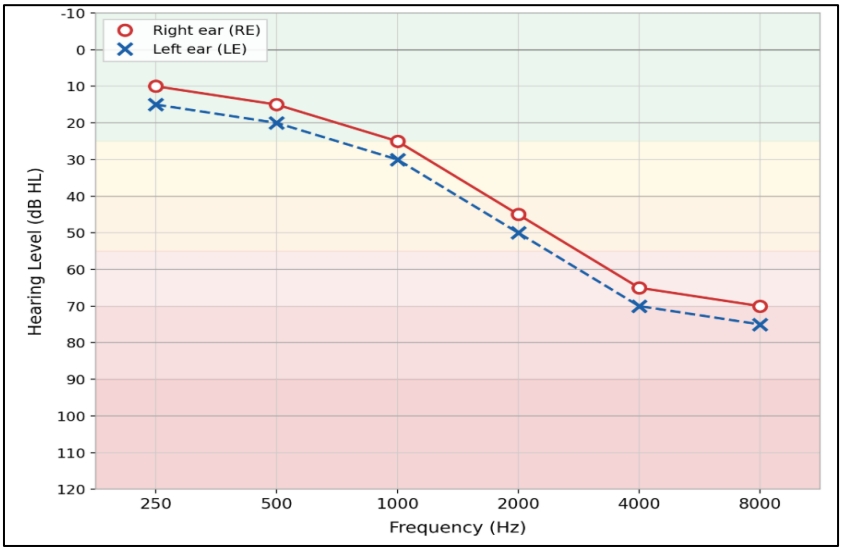

Figure 1 Pure-tone audiogram showing bilateral sensorineural hearing loss with a high-frequency sloping pattern

Radiological investigations were performed to further evaluate the extent of disease. Computed tomography (CT) of the paranasal sinuses demonstrated extensive bilateral sinonasal polyposis, associated with septal deviation and turbinate atrophy, without bone destruction. Chest imaging revealed peripheral pulmonary infiltrates suggestive of inflammatory involvement. Brain magnetic resonance imaging (MRI) did not show any central nervous system abnormalities.

Laboratory investigations revealed marked hypereosinophilia with an eosinophil count of 1950/mm^3. Inflammatory markers were elevated. Immunological testing showed positivity for cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA), while perinuclear ANCA (P-ANCA) were negative.

Based on the combination of long-standing asthma, significant hypereosinophilia, sinonasal polyposis, pulmonary infiltrates, and neurological involvement, the diagnosis of eosinophilic granulomatosis with polyangiitis (EGPA) was established according to the American College of Rheumatology classification criteria.

The patient was initiated on systemic corticosteroid therapy at a dose of 1 mg/kg/day, followed by gradual tapering. Given the significant sinonasal involvement, she underwent endoscopic sinus surgery with nasal polypectomy to improve symptoms and ventilation. Supportive treatment, including calcium and vitamin D supplementation, was also prescribed.

The clinical course was favorable, with marked improvement in nasal symptoms and general condition. Pulmonary and systemic manifestations regressed under treatment. Hearing loss remained stable without further deterioration, with no significant recovery observed, suggesting possible irreversible cochlear damage. The patient continues to be followed regularly with multidisciplinary care.

3. Discussion

EGPA is a rare and complex systemic necrotizing vasculitis affecting small- to medium-sized vessels, belonging to the spectrum of ANCA-associated vasculitides [1,2]. It is classically characterized by the association of asthma, peripheral eosinophilia, and multisystem involvement. The disease typically evolves through three phases: a prodromal allergic phase marked by asthma and rhinosinusitis, an eosinophilic phase with tissue infiltration, and a vasculitic phase responsible for systemic manifestations [3].

Otorhinolaryngological involvement is common in EGPA and is reported in up to 70–90% of cases [4]. It predominantly manifests as chronic rhinosinusitis with nasal polyposis, which was observed in our patient. However, auditory involvement remains uncommon and is rarely the presenting symptom. When present, hearing loss is most often conductive, related to middle ear effusion secondary to Eustachian tube dysfunction. In contrast, sensorineural hearing loss (SNHL), as observed in our case, is rare and suggests inner ear involvement [5].

The pathophysiology of SNHL in EGPA is not fully elucidated but is thought to involve several mechanisms. The most widely accepted hypothesis is vasculitic involvement of the cochlear microcirculation, leading to ischemia of the stria vascularis and hair cells. Eosinophilic infiltration and the release of cytotoxic proteins may also contribute to direct inner ear damage. Additionally, immune-mediated mechanisms associated with ANCA may play a role in endothelial injury [5,6]. These mechanisms explain the often progressive and sometimes irreversible nature of hearing impairment in EGPA.

Our case is particularly noteworthy because hearing loss was one of the initial manifestations that led to medical evaluation, preceding the full expression of systemic disease. Such atypical presentations may delay diagnosis, especially when respiratory symptoms are attributed solely to pre-existing asthma. This underlines the importance of considering systemic vasculitis in patients presenting with unexplained sensorineural hearing loss associated with ENT and systemic symptoms [2,5].

Neurological involvement is frequent in EGPA, most commonly presenting as peripheral neuropathy. Cranial nerve involvement, such as peripheral facial paralysis as observed in our patient, is less common but has been described. Its association with SNHL further supports a vasculitic mechanism affecting multiple neural structures [2].

The presence of ANCA in EGPA is variable, being detected in approximately 30–40% of cases [9]. ANCA-positive patients tend to exhibit more vasculitic manifestations, including neuropathy and renal involvement, whereas ANCA-negative patients more frequently present with eosinophilic tissue infiltration. In our patient, C-ANCA positivity is unusual, as EGPA is more commonly associated with P-ANCA (anti-MPO). This finding may reflect phenotypic variability and highlights the heterogeneity of the disease [2,9].

The diagnosis of EGPA is based on a combination of clinical, biological, and radiological findings. The American College of Rheumatology (ACR) classification criteria include asthma, eosinophilia >10%, neuropathy, pulmonary infiltrates, paranasal sinus abnormalities, and extravascular eosinophils [7]. Our patient fulfilled several of these criteria, supporting the diagnosis. In addition, the Chapel Hill consensus nomenclature provides a standardized definition of ANCA-associated vasculitides [8].

Differential diagnoses include other ANCA-associated vasculitides such as granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA). GPA typically presents with necrotizing granulomatous inflammation and more

frequent ENT destructive lesions, while MPA lacks granulomatous inflammation and prominent eosinophilia. The presence of asthma, marked eosinophilia, and nasal polyposis strongly favors EGPA in our case [2,3].

Early recognition and treatment of EGPA are essential to prevent irreversible organ damage. Systemic corticosteroids remain the cornerstone of therapy and are effective in controlling both eosinophilic inflammation and vasculitic activity. In cases with severe or refractory disease, immunosuppressive agents or biologic therapies such as anti–IL-5 monoclonal antibodies (e.g., mepolizumab) may be required [1,9]. In our patient, corticosteroid therapy led to clinical improvement and stabilization of hearing loss, highlighting the importance of early intervention.

This case emphasizes that sensorineural hearing loss may represent an early and atypical manifestation of EGPA. Clinicians, particularly otorhinolaryngologists, should be aware of this possibility to ensure timely diagnosis and appropriate management.

4. Conclusion

Eosinophilic granulomatosis with polyangiitis is a rare and heterogeneous systemic vasculitis with a wide spectrum of clinical manifestations. Although ENT involvement is frequent, sensorineural hearing loss remains an uncommon and underrecognized presentation, which may precede the onset of systemic features.

This case highlights the importance of maintaining a high index of suspicion for EGPA in patients presenting with unexplained sensorineural hearing loss, particularly in the presence of asthma, eosinophilia, or sinonasal disease. Early recognition and prompt initiation of appropriate therapy are crucial to prevent irreversible organ damage and improve patient outcomes.

Increased awareness among otorhinolaryngologists is essential to reduce diagnostic delay and ensure timely multidisciplinary management.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of ethical approval

The present study was conducted in accordance with ethical standards.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report.

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