

Bilateral Sensorineural Hearing Loss as the Sentinel Presentation of Granulomatosis With Polyangiitis

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Abstract

Granulomatosis with polyangiitis (GPA) is a systemic necrotizing granulomatous vasculitis classically involving the upper and lower respiratory tracts and kidneys. Timely diagnosis requires a high index of suspicion for the presenting symptoms. Otolaryngologic manifestations are commonly reported but most often involve the nasal and paranasal sinuses. Sensorineural hearing loss (SNHL) as the sole initial manifestation of GPA is uncommon and may contribute to diagnostic delay.

We report a case of a 67-year-old male who presented with rapidly progressive bilateral SNHL that preceded pulmonary and renal manifestations by several months, during which the patient was treated for otitis externa and Eustachian tube dysfunction without improvement. Development of pulmonary symptoms prompted additional workup, which demonstrated markedly elevated cytoplasmic anti-neutrophil cytoplasmic antibody (c-ANCA) and proteinase 3-anti-neutrophil cytoplasmic antibody (PR3-ANCA), raising concern for GPA. Histopathology then confirmed the diagnosis. The patient was treated with high-dose corticosteroids and rituximab with serologic remission; however, his hearing loss was largely irreversible.

This case highlights bilateral SNHL as an uncommon sentinel manifestation of GPA and emphasizes the importance of considering GPA in patients with unexplained rapidly progressive bilateral SNHL. Early recognition and prompt treatment may reduce irreversible injury.

Categories: Otolaryngology, Rheumatology, Internal Medicine

Keywords: anca-associated vasculitis, bilateral sensorineural hearing loss, diagnostic delay, granulomatosis with polyangiitis (gpa), uncommon presentation

Introduction

Granulomatosis with polyangiitis (GPA) is a rare anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis with an estimated prevalence of three per 100,000 persons in the United States [1]. The disease is characterized by necrotizing granulomatous inflammation and immune-mediated small-to-medium vessel vasculitis, classically involving the upper and lower respiratory tracts and kidneys [2]. Otolaryngologic manifestations involving the sinonasal tract, oral cavity, pharynx, larynx, and/or ear appear in 70-90% of patients, with 16-36% having otologic symptoms at presentation [3-6]. The most common otologic manifestation is serous otitis media, resulting in conductive hearing loss; in contrast, sensorineural hearing loss (SNHL) is less common, reported in only 6-12% of patients with GPA [6,7]. Cases in which rapidly progressive SNHL is the predominant or sole initial manifestation of disease have only been reported sporadically [8,9]. Diagnosis of GPA in patients who present with isolated otolaryngologic symptoms remains challenging, with the interval from symptom onset to diagnosis often exceeding six months [4]. Delayed recognition may result in irreversible organ damage, including permanent hearing loss.

We report a case of GPA in which rapidly progressive bilateral SNHL preceded pulmonary and renal involvement by three months, resulting in diagnostic delay and irreversible auditory injury.

Case Presentation

A 67-year-old male with a history of hypertension, hyperlipidemia, and a prior diagnosis of Eustachian tube dysfunction presented with a one-to-two month history of fevers, chills, night sweats, generalized weakness, and intermittent hemoptysis preceded by progressive bilateral hearing loss.

Approximately three months prior to presentation, the patient developed left-sided hearing loss, which was initially treated as otitis externa without improvement. Over the following weeks, he developed fatigue, intermittent fevers, night sweats, and vertigo, prompting referral to otolaryngology. Audiometric testing demonstrated asymmetric SNHL, worse in the left ear (Table 1). A course of corticosteroids produced only transient mild improvement before he developed worsening hearing loss in the right ear with continued

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intermittent vertigo. Repeat audiogram revealed worsening of bilateral SNHL (Table 1). Brain MRI showed features concerning for Eustachian tube dysfunction, and the patient underwent Eustachian tube dilation without benefit.

Timepoint	Measure	Right ear	Left ear
Two months before admission	Audiogram pattern	Normal sloping to moderately severe SNHL	Moderately severe to severe SNHL
	Speech reception threshold	10 dB	70 dB
	Word recognition score	100%	36%
One month before admission	Audiogram pattern	Mild to severe SNHL	Severe SNHL
	Speech reception threshold	40 dB	85 dB
	Word recognition score	85%	Could not evaluate
During admission	Audiogram pattern	Moderate to severe SNHL	Profound SNHL
	Speech reception threshold	N/A	N/A
	Word recognition score	90%	Could not evaluate
One month after discharge	Audiogram pattern	Normal sloping to moderately severe SNHL	Severe to profound SNHL
	Speech reception threshold	20 dB	No response at 105 dB
	Word recognition score	100%	8%

TABLE 1: Longitudinal audiologic progression.

SNHL: sensorineural hearing loss; N/A: not available.

Over the next few weeks, the patient noted an unintentional 10-pound weight loss and developed new progressive hemoptysis, which eventually prompted evaluation at an outside emergency department where chest CT revealed a 3.4 × 3.8 cm left apical consolidation, mediastinal lymphadenopathy, and tree-in-bud opacities. Pulmonary coccidioidomycosis was suspected, and empiric fluconazole was initiated. However, his symptoms continued to worsen, leading to his presentation to our institution (Figure 1).

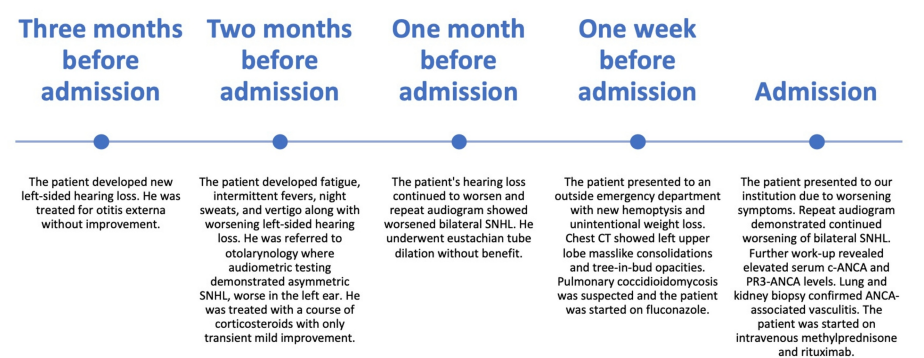


FIGURE 1: Timeline of symptoms and evaluation.

SNHL: sensorineural hearing loss; c-ANCA: cytoplasmic anti-neutrophil cytoplasmic antibody; PR3-ANCA: proteinase 3-anti-neutrophil cytoplasmic antibody.

On presentation, vital signs were notable for a heart rate of 104 beats/minute and a respiratory rate of 28 breaths/minute, with oxygen saturation of 95% on room air. Physical examination was significant only for marked hearing impairment. Repeat audiogram demonstrated progression to moderate-to-severe SNHL in the right ear and profound SNHL in the left ear (Table 1). Re-review of the outside chest CT was notable for a left apical pulmonary mass with several additional small lung nodules and associated mediastinal lymphadenopathy. The differential diagnosis included malignancy, infection, vasculitis, and paraneoplastic processes.

Initial laboratory evaluation demonstrated elevated inflammatory markers, including erythrocyte sedimentation rate (ESR) of 123 mm/hr and C-reactive protein (CRP) of 22.0 mg/dL (Table 2). An extensive infectious workup for bacteria, fungi, and tuberculosis was unrevealing. Further evaluation revealed a cytoplasmic (c)-ANCA titer of 1:640 and a proteinase 3 (PR3)-ANCA level of 1,078.3 CU (reference range <20.0 CU) (Table 2). Temporal bone MRI showed marked left otomastoid effusion with intrinsic T1-hyperintense signal and associated inflammatory changes, including abnormal enhancement of the left mastoid air cells and Eustachian tube as well as adjacent subtle pachymeningeal enhancement of the left middle cranial fossa (Figure 2). A moderate right otomastoid effusion was also present without significant inflammatory changes. These findings were interpreted as suspicious for inflammatory otomastoiditis and, together with the positive ANCA serologies, raised concern for GPA. Rheumatology was consulted, and intravenous methylprednisolone equivalent to 1 mg/kg of prednisone daily was initiated empirically.

Lab test	Result	Reference range
ESR	123 mm/hr	<= 12 mm/hr
CRP	22.0 mg/dL	< 0.8 mg/dL
Cytoplasmic (c)-ANCA	1:640 titer	< 1:20 titer
Proteinase 3 (PR3)-ANCA	1078.3 CU	< 20.0 CU
Creatinine	1.8 mg/dL	0.70 – 1.35 mg/dL (patient baseline: 1.1 mg/dL)

TABLE 2: Notable laboratory findings during admission.

ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; ANCA: anti-neutrophil cytoplasmic antibody.

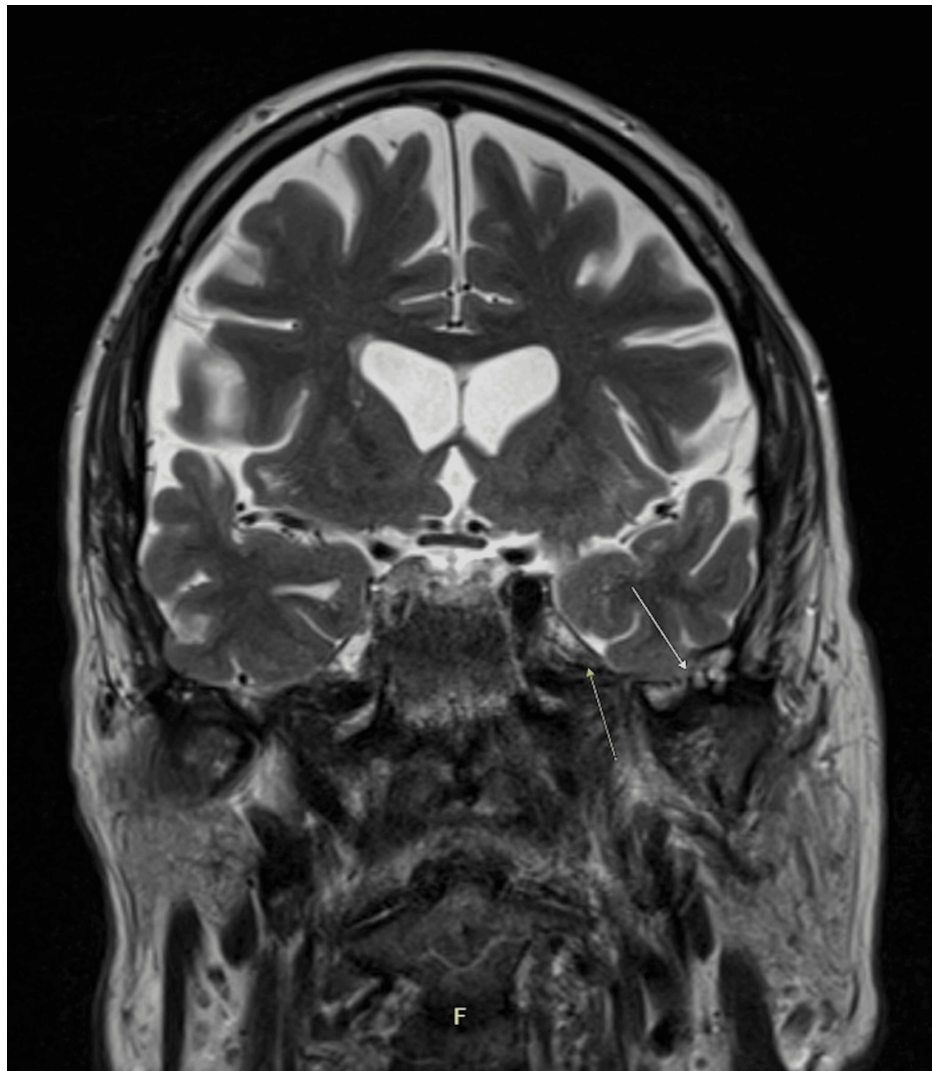


FIGURE 2: MRI of the temporal bones with gadolinium.

Coronal post-contrast T1-weighted sequence demonstrating marked left otomastoid effusion with intrinsic T1-hyperintense signal and associated inflammatory changes (white arrow), including abnormal enhancement of left mastoid air cell walls and the Eustachian tube. There is also adjacent pachymeningeal enhancement along the left middle cranial fossa with extension to the inferior margin of Meckel's cave (yellow arrow). Moderate right otomastoid effusion without inflammatory signal changes is also present.

The patient subsequently underwent bronchoscopic biopsy of the lung lesion. Pathology demonstrated focal neutrophilic capillaritis, a finding compatible with ANCA-associated vasculitis. During hospitalization, the patient developed progressive acute kidney injury with creatinine increasing from a baseline of approximately 1.1 mg/dL to 1.8 mg/dL (Table 2). Urinalysis demonstrated hematuria and trace proteinuria. Consequently, a renal biopsy was performed; pathology revealed diffuse necrotizing and crescentic glomerulonephritis with active crescents involving approximately 65% of glomeruli and rare segmental glomerular scars (<5%) (Figure 3). Pauci-immune immunofluorescence and absent immune complex deposits on electron microscopy were consistent with a diagnosis of pauci-immune PR3-ANCA-associated glomerulonephritis.

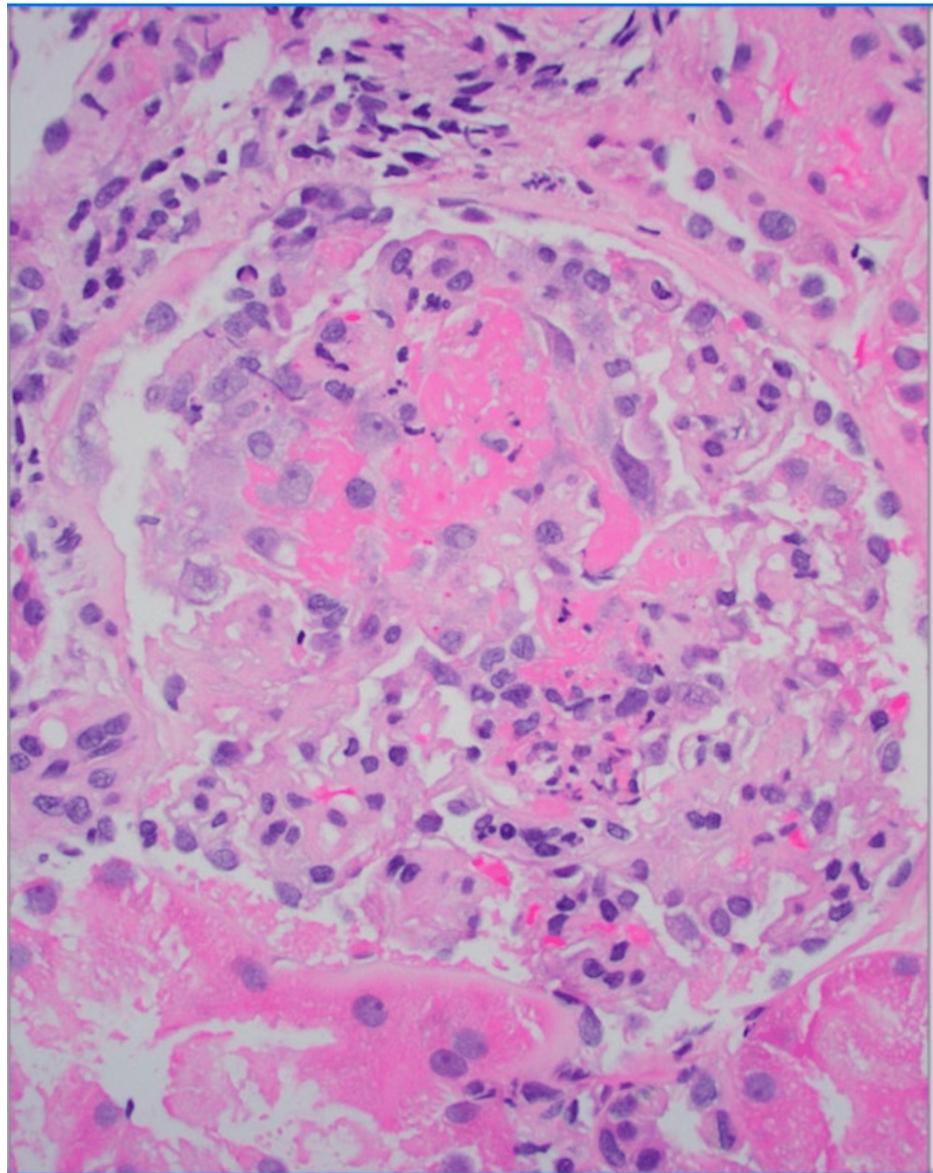


FIGURE 3: Renal biopsy (light microscopy).

Periodic acid–Schiff stain demonstrating diffuse necrotizing and crescentic glomerulonephritis with active cellular crescents involving approximately 65% of glomeruli.

Given biopsy-proven renal involvement, the patient's glucocorticoid regimen was increased to pulse-dose intravenous methylprednisolone for three days, followed by a prolonged oral prednisone taper. Weekly rituximab was also initiated (Figure 1). The patient was then discharged to outpatient care, where he completed a four-dose rituximab course, began avacopan therapy, and continued gradual glucocorticoid tapering. Serial PR3-ANCA titers demonstrated a sustained serologic response (Table 3) with improvement of his constitutional, pulmonary, and renal manifestations; however, follow-up audiologic evaluation showed persistent severe-to-profound SNHL in the left ear with only partial recovery in the right ear (Table 1).

Clinical timepoint	PR3-ANCA (CU)
Day of admission	1,078.3
Several days after admission	1,079.1
1 week post-discharge	358.4
~2 months post-discharge	87.1
~3 months post-discharge	46.7
~7 months post-discharge	<20.0

TABLE 3: Proteinase 3-anti-neutrophil cytoplasmic antibody (PR3-ANCA) trend.

Discussion

GPA is a systemic necrotizing vasculitis that classically involves the upper and lower respiratory tracts and kidneys, often presenting with rhinosinusitis, otitis media, pulmonary lesions, and/or pauci-immune glomerulonephritis along with constitutional symptoms. In contrast, our patient developed rapidly progressive bilateral SNHL approximately three months before systemic manifestations became apparent, resulting in delayed diagnosis and irreversible auditory injury.

Otolaryngologic involvement occurs in 70-90% of patients with GPA, but despite this high prevalence, patients presenting with isolated otologic symptoms often experience greater diagnostic delays [4]. Serous otitis media is the most common otologic manifestation and typically results from nasopharyngeal inflammation and/or obstruction that causes Eustachian tube dysfunction, primarily producing conductive hearing loss [6,10]. In contrast, SNHL is less common and has only been described as the presenting feature in case reports [8-10]. Its mechanism in GPA is incompletely understood but is thought to involve vasculitis of the cochlear vessels, immune complex deposition in the cochlea, granulomatous infiltration of the cochlear nerve, and/or toxic effects of inflammatory products in the middle ear [6,11].

Bilateral SNHL is rare, with cohort studies suggesting bilateral involvement in fewer than 5% of individuals with SNHL, and should prompt evaluation for systemic etiologies, including autoimmune, infectious, toxic-metabolic, vascular, and neoplastic disorders [10,12-14]. Timely diagnosis is critical, as hearing recovery appears to be influenced by early immunosuppressive therapy. In a retrospective series of eight patients presenting with mixed conductive and SNHL as the initial manifestation of GPA, individuals with hearing thresholds better than 95 dB demonstrated improvement within nine months of immunosuppressive therapy, whereas those with complete hearing loss across all frequencies showed little to no recovery [15]. This pattern mirrors our patient's course: moderate-to-severe right-sided SNHL partially improved with treatment, while profound left-sided SNHL remained irreversible despite aggressive immunosuppression. These findings underscore the importance of considering ANCA-associated vasculitis in patients presenting with rapidly progressive or bilateral SNHL.

In our patient, neuroimaging demonstrated complementary evidence of localized vasculitic injury to the auditory apparatus. Temporal bone MRI revealed a constellation of findings (T1-hyperintense otomastoid effusion, enhancement of the mastoid wall and Eustachian tube, and pachymeningeal involvement extending to Meckel's cave) that should raise suspicion for GPA in the appropriate clinical context [16,17]. Pachymeningeal GPA is a recognized entity capable of causing SNHL through direct cochlear vasculitis, ischemic injury, or neural compression along the skull base [17,18]. Its presence on the side of dominant auditory loss in this patient provides a possible anatomic correlate for the severity and irreversibility of left-sided cochlear damage.

Rituximab in combination with high-dose glucocorticoids achieved sustained serologic remission, although audiologic recovery was limited. Cochlear outcomes are often constrained by the extent of pre-existing injury: vasculitic damage to the cochlea can result in irreversible hair cell loss, and most patients with severe or prolonged GPA-associated SNHL experience only partial or no meaningful improvement despite adequate immunosuppression [19]. Isolated reports of reversible cochlear dysfunction when therapy is initiated early, such as the series described by Yamazaki et al., suggest a narrow therapeutic window [9]. In our patient, the shorter duration of right-sided disease at treatment initiation may permit further modest improvement, whereas substantial recovery of the profoundly affected left ear is unlikely.

Conclusions

This case highlights an uncommon presentation of GPA where rapidly progressive bilateral SNHL preceded the onset of pulmonary and renal manifestations by several months, contributing to diagnostic

delay. Patients presenting with bilateral SNHL should prompt evaluation for systemic etiologies, including ANCA-associated vasculitis. Early recognition is critical as hearing recovery may depend on early initiation of immunosuppressive therapy. Although glucocorticoids and rituximab led to serologic remission and partially improved the less severely affected right ear, our patient continued to have severe left-sided hearing loss, underscoring the narrow therapeutic window for preserving auditory function. Greater recognition of rapidly progressive bilateral SNHL as a potential early manifestation of GPA may facilitate earlier diagnosis, timely treatment, and improved hearing outcomes for patients.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Wen-Ching Tran, Chun Hin Chan, Samuel Margolis

Drafting of the manuscript: Wen-Ching Tran, Chun Hin Chan, Samuel Margolis

Critical review of the manuscript for important intellectual content: Wen-Ching Tran, Chun Hin Chan

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