



Wegener's Granulomatosis Presenting with Otological Manifestations

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Abstract

Objectives: To describe the otologic manifestations of patients presenting with a confirmed diagnosis of granulomatosis with polyangiitis (GPA).

Study design: Retrospective case series.

Methods: To describe a series of GPA patients presenting with otologic complaints.

Results: Of the 9 patients included, 3 (33%) had a previous diagnosis of GPA. The remaining 6 (67%) were diagnosed due by the neurotologist due to suspicious otologic complaints. Of the cohort, 8 (88%) patients presented with hearing loss, more than half of which was unilateral (62%). Upon audiometric examination, all but 1 patient had either mixed or conductive hearing loss. All patients presented with Eustachian tube dysfunction (ETD), otitis media with effusion (OME), or both. Nasal endoscopy showed intranasal pathology in 3 (33%) patients, including a nasal septum perforation or granulation tissue on the lateral wall of the nose. One patient developed subglottic stenosis after diagnosis.

Conclusions: GPA should be included in the differential diagnosis of patients with mixed or conductive hearing loss and new onset serous effusion or adult with acute otitis media in the absence of a previous history of ETD. ANCA test, ESR, CRP, and a urinalysis are rapid tests that can help screen these patients.

Introduction

- GPA, previously known as Wegener's granulomatosis, is a rare multisystem autoimmune disorder, characterized by necrotizing granulomatous inflammation and pauci-immune vasculitis.
- In the United States, GPA has a prevalence of 3 cases per 100,000.¹
- Common head and neck manifestations include chronic sinusitis, rhinitis, and epistaxis, among others.
- Notably, up to 88% of patient's experience ENT symptoms, with otologic symptoms present 19-61% of the time.²
- We analyzed a cohort of GPA patients who presented with otologic findings and discuss its diagnostic pitfalls.

Methods and Materials

- Retrospective review of patients presenting to an outpatient otology/neurotology clinic at a tertiary care academic center over a 6-year period.
- 6 patients presented with a primary otologic complaints with no previous history of GPA. After clinical examination, and further laboratory testing all patients included were determined to have a diagnosis of GPA, confirmed with an elevated c-ANCA.

Results

- A total of 9 patients presented with a confirmed diagnosis of GPA (**Table 1**), with an equal distribution of males and females and an average age of 52 years old.
- 3 (33%) patients were of Asian descent, the remaining six (67%) were Caucasian.
- Upon examination (**Figure 1**), 4 (44%) patients had only OME, 2 (22%) patients had only ETD, and 3 (33%) patients had both.
- Nasal endoscopy showed evidence of intranasal pathology in 3 (33%) patients, with 1 (11%) patient having a septal perforation.
- The most common otologic finding was hearing loss, which was observed in 8 (88%) patients, and 5 (55%) patients endorsed otalgia.
- Of the 8 patients with hearing loss, audiometric testing showed 4 (55%) had mixed, 3 (38%) had conductive, and 1 (13%) had sensorineural hearing loss.
- Pattern of hearing loss was unilateral in 5 (55%) patients.
- All patients received immunosuppressive therapy for the systemic disease process. Otologic treatment is outlined in the **Table 1**.
- All patients experienced improvement of otologic symptoms after treatment (immunosuppressants or tube placement), with an average of 4 in-office follow-up appointments.

Patient	Age	Gender	Otologic finding	Otalgia	Type of hearing loss	Pattern of hearing loss	Otologic treatment
Patient 1	36	F	OME/ ETD	N	Conductive	Unilateral	PE tube placement
Patient 2*	41	F	ETD	Y	Conductive	Unilateral	PE tube placement
Patient 3*	51	M	OME	Y	Mixed	Bilateral	PE tube placement
Patient 4*	52	M	OME	Y	Mixed	Unilateral	PE tube placement
Patient 5*	51	F	OME/ ETD	Y	Mixed	Bilateral	PE tube placement
Patient 6	46	F	ETD	N	Conductive	Bilateral	Observation
Patient 7	78	M	OME/ ETD	N	Sensorineural	Bilateral	Observation
Patient 8*	43	F	OME	N	None	Unilateral	Observation
Patient 9*	60	M	OME	Y	Mixed	Unilateral	Observation

* Denotes patients diagnosed first in the otology clinic based on the otologic manifestation.

Table 1. Tabulation of lesion characteristics. OME = Otitis media with effusion; ETD = Eustachian tube dysfunction

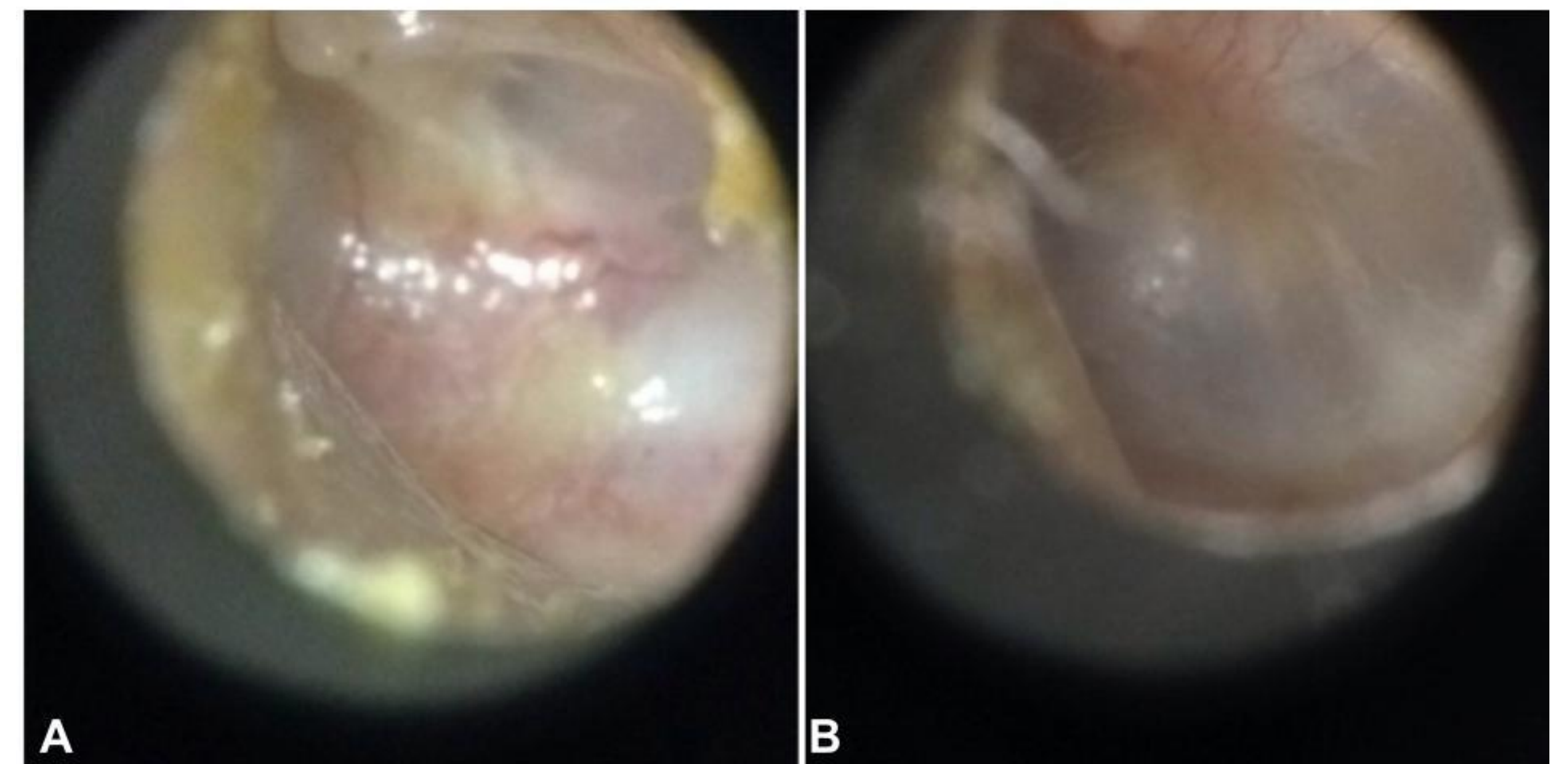


Figure 1. (A) Pale thickened mucosa in the middle ear, (B) middle ear effusion.

Discussion

- This patient cohort illustrates the significance of thoroughly assessing all otologic complaints as a possible indication for GPA.
- The otologic manifestations of GPA are commonly the only symptoms on presentation that can aid in the diagnosis of GPA.
- Others have reported that 25% of GPA patients present with OME, with hearing loss as the most common symptom found in 6% of patients.³
- We confirm the high prevalence of both OME and hearing loss, suggesting that clinicians should maintain a high index of suspicion for GPA in patients presenting with these clinical findings.
- As we demonstrated, GPA patients are responsive to treatment with near complete symptomatic resolution of all otologic symptoms.
- In the work up of patients with no diagnosis of GPA, three patients had an initial negative ANCA test which became positive on a follow up test.
- All patients with a false negative test were on steroids at the time of testing. Therefore it is imperative to have a high index of suspicion and re-test patients 2-3 weeks after steroids have been stopped. Reassessment of nasal endoscopy results and biopsy of nose granulation tissue can assist in the diagnosis of GPA when ANCA testing is negative.
- As such, in the context of otologic complaints described above, and a high ESR, CRP or an abnormal urine analysis the suspicion of GPA should increase even when ANCA testing is normal.

Conclusions

- A high index of suspicion for GPA should be applied to patients with mixed or conductive hearing loss with new onset serous effusion in the absence of previous ETD.
- All patients with a false negative tests were on steroids at the time of testing. Therefore it is imperative to have high suspicion and re-test patients 2-3 weeks after steroids have been stopped. Reassessment of nasal endoscopy results and biopsy of nose granulation tissue can assist in the diagnosis of GPA when ANCA testing is negative.
- As such, in the context of otologic complaints described above, and a high ESR, CRP or an abnormal urine analysis the suspicion of GPA should increase even when ANCA testing is normal.

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