CASE REPORT - OPEN ACCESS

ISSN: 3067-2368 American Journal of Case Reports and Clinical Images DOI: http://dx.doi.org/10.51521/AJCRCI.2025.e22.127

Contents lists available at https://amejcaserepclinimag.com/

American Journal of Case Reports and Clinical Images

Granulomatosis with Polyangiitis: A Rare Case Report

Kamran Mateen[1], Aisha[2], Touqeer Rehman[3], Zain Mustafa[4], Muhammad Azhar Uddin[5]

[1]*Northwest School of Medicine*

[2]*Islamic International Medical College, Riphah International University*

[3]*Ayub Medical and Dental College, Abbottabad*

[4]*Bakhtawar Amin Medical and Dental College, Multan*

[5]*The Queen Elizabeth hospital NHS foundation trust King's Lynn United Kingdom*

ARTICLE INFO

Article history:
Received: 24-09-2025
Revised: 15-10-2025
Accepted: 17-10-2025
Published: 21-10-2025

ABSTRACT

Wegener's granulomatosis (WG) is a rare autoimmune disorder that causes destructive inflammation in the respiratory tract, glomerulonephritis and vasculitis typically affecting adults aged 40-60 yrs. Surprisingly,we present an unusual case of localized GPA in an 18-year-old South Asian female with nasal obstruction, epistaxis, and headaches. Imaging revealed nasal polyps and sinus involvement, while histopathology confirmed granulomatous inflammation with positive c-ANCA (PR3). Treated with corticosteroids and methotrexate, she achieved remission without systemic progression. This case highlights the importance of early biopsy and ANCA testing in young patients with atypical presentations to prevent irreversible organ damage. A multidisciplinary approach ensured timely diagnosis and effective management.

Introduction

Granulomatosis with polyangiitis (GPA), previously called Wegener's granulomatosis, is a severe and potentially fatal inflammatory disease affecting small to medium-sized blood vessels, mainly in the lungs and kidneys. It is marked by granuloma formation, vasculitis, and a type of kidney inflammation known as pauci-immune necrotizing glomerulonephritis. GPA is typically linked to antineutrophil cytoplasmic antibodies (ANCA). Without treatment, the condition carries a mortality rate of up to 80%. However, aggressive immunosuppressive therapies-such as glucocorticoids, cyclophosphamide, rituximab, methotrexate, and azathioprine-have reduced this rate by nearly 70%. Despite improved survival, patients often face significant long-term complications from both the disease and its treatment, including relapses, organ damage, and ongoing low-grade inflammation, all of which contribute to reduced quality of life and substantial healthcare costs [1].

Although generally rare, this disease shows notable geographical variation, with its prevalence increasing along a latitudinal gradient-that is, it becomes more common the farther one moves away from the Equator [2]. The peak incidence is observed in mid-sixties to mid-seventies, depending upon the patient's geographical placement [3]. We report a rare case of GPA in a South Asian eighteen-year-old female with respiratory complications.

Case Presentation:

An 18-year-old previously healthy female presented to the otolaryngology department with a one-week history of bilateral nasal congestion, more pronounced on the right side. She also reported intermittent frontal headaches, postnasal drip, and episodes of epistaxis. In addition, she experienced a dry, non-productive cough and mild dyspnea. There was no sputum production, hemoptysis, or night sweats and she denied fever or weight loss. On physical examination, there was marked nasal inflammation with what appeared to be Grade IV nasal polyps causing near-complete obstruction of the nasal passages. She had no known allergies, chronic medical illness, or autoimmune disease. She was initially managed with oral antihistamines and intranasal decongestants, and was advised imaging for further evaluation.

Computerized tomography of the paranasal sinuses showed soft tissue density lesions occupying the anterior part of the right nasal cavity, bulging into the right ethmoid air cells and extending into the contralateral nasal cavity. The right inferior turbinates were markedly hypertrophied (Figure 1). Laboratory investigations revealed elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Anti-neutrophil cytoplasmic antibody (ANCA) testing returned positive for cytoplasmic ANCA (c-ANCA), while perinuclear ANCA(p-ANCA) was negative. Although a chest radiograph was unremarkable, high-resolution computerised tomography (HRCT) of the chest revealed bilateral diffused mild bronchodilation with no zonal predominance and scattered focal patches of circumferential bronchial wall thickening (Figure 2) Given the persistent and progressive nature of her symptoms and the imaging findings, the patient was admitted for diagnostic nasal endoscopy and biopsy. Intraoperatively, inflamed tissue was observed involving the nasal septum and bilateral inferior turbinates, which bled easily on manipulation. The inferior turbinates were partially resected and the hypertrophied septal mucosa was trimmed to relieve obstruction; and the samples were sent for histopathological examination and culture. Microscopy revealed granulomatous inflammation composed of histocytes, focally surrounded by blood vessels. No fungal elements or acid-fast bacilli were detected, and cultures were negative. These histopathological findings, elevated inflammatory markers and positive c-ANCA, confirmed the diagnosis of limited granulomatosis with polyangitis.

Following confirmation of the diagnosis, the patient was referred to the

* Corresponding author.
Aisha, Northwest School of Medicine; Islamic International Medical College, Riphah International University.

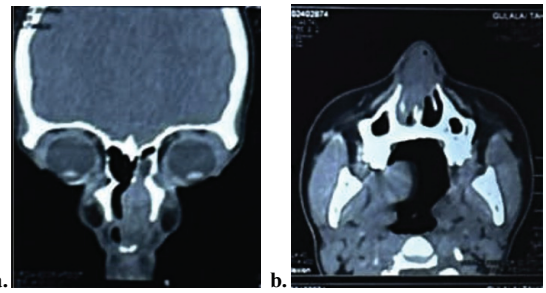

Figure 1: Plain Computed Tomography Scan of Paranasal Sinuses A. (Coronal) and B. (Axial) shows hypertrophied right inferior turbinate and soft tissue in the anterior part of the nasal cavity on the right side bulging into right ethmoidal air cells and contralateral nasal cavity.

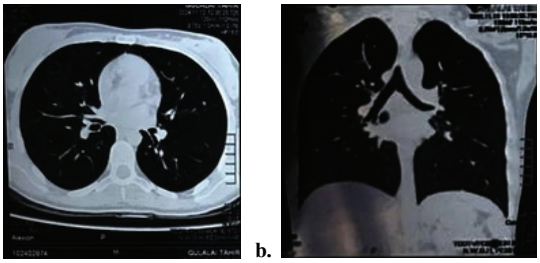

Figure 2: High-Resolution Computed Tomography of chest A. (Axial) and B. (Coronal) showing bilateral diffuse mild bronchodilation.

Rheumatology department for systemic management. Given the absence of renal involvement and the localized nature of the disease, she was initiated on oral corticosteroid therapy with prednisolone (Deltacortil) at a dose of 25 mg daily. The steroid regimen was gradually tapered over several weeks to 15 mg, then to 10 mg and subsequently maintained at 5 mg daily. Concurrently, she was prescribed methotrexate starting at 7.5 mg per week, titered to 10 mg per week, along with folic acid supplementation to prevent methotrexate-associated toxicity. Serial monitoring of complete blood count, liver function tests, renal profile, erythrocyte sedimentation rate (ESR), and c-reactive protein (CRP) was conducted every 2 weeks to track disease activity and treatment response. The patient also followed up with the ENT team for local sinonasal care, which included regular nasal saline irrigations and debridement of crusting that had developed from both the disease and surgical intervention.

The patient showed significant improvement within weeks of initiating immunosuppressive therapy. Her nasal obstruction and post nasal symptoms gradually resolved, and there was no development of systemic features such as hematuria, skin lesions or peripheral neuropathy. The patient remains on low-dose maintenance therapy and continues regular follow-up with both ENT and rheumatology departments. To date, there has been no evidence of disease extension to other organ systems.

Discussion

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare ANCA-associated vasculitis involving small- to medium-sized blood vessels, characterized by granulomatous inflammation [5]. It typically affects individuals between 40 and 60 years of age and has a slight male predominance. The presentation in our case—an 18-year-old female—is relatively uncommon, highlighting the importance of considering GPA even in younger, less typical populations.

The disease classically presents with a triad of upper respiratory tract involvement, pulmonary manifestations, and renal disease. Our patient exhibited localized upper respiratory symptoms, including nasal obstruction, epistaxis, headaches, and dyspnea due to nasal blockage, without any pulmonary or renal involvement. Although limited GPA occurs in up to 29% of cases, it is often under-recognized in the early stages due to nonspecific symptoms and absence of systemic features [4]. Nasal endoscopy revealed mucosal crusting and clots, and CT paranasal sinuses showed mass-like obstruction of the nasal cavity. Differential diagnoses at this point included chronic rhinosinusitis, fungal infections (e.g., mucormycosis), tuberculosis, sarcoidosis, and nasal NK/T-cell lymphoma. The decision to proceed with functional endoscopic sinus surgery (FESS) provided both therapeutic relief and critical diagnostic tissue. Histopathology confirmed granulomatous inflammation, while elevated c-ANCA (PR3), ESR, and CRP supported the diagnosis.

Pathophysiologically, GPA is driven by autoantibody-mediated neutrophil activation (predominantly c-ANCA/PR3), leading to necrotizing granulomatous vasculitis that damages affected tissues. In the localized form, inflammation remains confined to the upper airway mucosa, but it can evolve into systemic disease if not treated Promptly.

The patient was started on oral corticosteroids (Deltacortil 25 mg daily) and methotrexate 7.5 mg weekly. Over time, based on clinical improvement and normalization of inflammatory markers, the steroid dose was gradually tapered to minimize long-term side effects. In parallel, the methotrexate dose was increased to 10 mg weekly to maintain disease control and reduce the risk of relapse. Serial monitoring with CBC, renal function, CRP, and urinalysis showed stable results and no signs of systemic involvement. Currently, the patient is maintained on Deltacortil 5 mg daily and methotrexate 10 mg weekly, with good disease control but residual nasal crusting and occasional facial discomfort—common in chronic GPA due to mucosal damage.

This case highlights the importance of a multidisciplinary approach—involving ENT surgeons, rheumatologists, and pathologists—in the timely diagnosis and management of GPA. Early biopsy and ANCA testing were essential in confirming the diagnosis and initiating treatment before irreversible tissue or organ damage occurred. If left untreated, GPA carries a high risk of morbidity and mortality due to progressive organ involvement. Fortunately, with prompt recognition and immunosuppressive therapy, remission rates are high, even in younger patients. Long-term follow-up is crucial due to the chronic relapsing nature of the disease. In summary, this case emphasizes the need to consider GPA in young patients with persistent sinonasal symptoms. Early diagnostic biopsy and ANCA testing can be life-saving, and treatment should be initiated promptly to prevent systemic progression and improve quality of life.

Conclusion:

Our case highlights the importance of maintaining a high index of suspicion for ANCA-associated vasculitis even in atypical age groups and demographic populations. The patient's localized upper respiratory symptoms, histopathological confirmation of granulomatous inflammation, and positive c-ANCA (PR3) were decisive in establishing the diagnosis. Prompt initiation of corticosteroids and methotrexate, combined with multidisciplinary care, resulted in significant clinical improvement without systemic progression.

This case highlights that (1) GPA should be considered in young patients with refractory sinonasal symptoms, (2) early tissue biopsy and ANCA testing are critical for timely diagnosis, and (3) localized disease requires tailored immunosuppression to prevent organ damage while minimizing treatment related morbidity. Long-term follow-up remains essential given the relapsing nature of GPA.

Funding: This research received no external funding.

Institutional Review Board Statement: Not applicable.

Informed Consent Statement: Not applicable.

Data Availability Statement: No new data were created or analyzed in this study. Data sharing is not applicable to this article.

Acknowledgments: Not applicable.

Conflicts of Interest: The authors declare no conflict of interest.

References

1. Panupattanapong S, Stwalley DL, White AJ, Olsen MA, French AR, Hartman ME. Epidemiology and Outcomes of Granulomatosis With Polyangiitis in Pediatric and Working-Age Adult Populations In the United States: Analysis of a Large National Claims Database. Arthritis Rheumatol. 2018 Dec;70(12):2067-2076. doi: 10.1002/ art.40577. PMID: 29806148; PMCID: PMC6258356.
2. O'Donnell JL, Stevanovic VR, Frampton C, Stamp LK, Chapman PT. Wegener's granulomatosis in New Zealand: evidence for a latitude-dependent incidence gradient. Intern Med J. 2007 Apr;37(4):242-6. doi: 10.1111/ j.1445-5994.2006.01297.x. PMID: 17388864.
3. Kubaisi B, Abu Samra K, Foster CS. Granulomatosis with polyangiitis (Wegener's disease): An updated review of ocular disease manifestations. Intractable Rare Dis Res. 2016 May;5(2):61-9. doi: 10.5582/irdr.2016.01014. PMID: 27195187; PMCID: PMC4869584.

Kamran Mateen, Aisha, Touqeer Rehman, Zain Mustafa, Muhammad Azhar Uddin (2025). Granulomatosis with Polyangiitis: A Rare Case Report. American J Case Rep Clin Imag. 2025; Oct, 2(2), 01-03.

4. Holle JU, et al. ' Long-term outcome in patients with localized Wegener's granulomatosis.' Rheumatology (Oxford). 2010.

5. Jennette JC, et al. '2012 revised International Chapel Hill Consensus Conference nomenclature of vasculitides.' Arthritis Rheum. 2013;65(1):1–11.