Case Report

Granulomatosis With Polyangiitis Presenting as Hemoptysis Initially Suspected as Pulmonary Tuberculosis: A Case Report from a Tertiary Center in Nepal

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Keywords: *Granulomatosis with polyangiitis, C-ANCA, Hemoptysis, Pulmonary hemorrhage, Acute kidney injury, Vasculitis*

Abstract

Granulomatosis with polyangiitis (GPA), previously called Wegener's granulomatosis, is a rare type of small-vessel vasculitis that causes necrotizing granulomatous inflammation, mainly affecting the respiratory tract and kidneys. In areas where tuberculosis is common, it can easily be mistaken for an infection like pulmonary TB, which often leads to delayed diagnosis.

We report a case of a 66-year-old man who came in with hemoptysis and was initially thought to have pulmonary tuberculosis. His condition quickly deteriorated, developing massive hemoptysis, difficult-to-control hypertension, and acute kidney injury, with his serum creatinine rising sharply from 1.4 mg/dL to 5.3 mg/dL within a short time. Chest X-ray showed a homogeneous mass in the middle zone of the right lung, while CT scan findings were consistent with diffuse alveolar hemorrhage. Procalcitonin was normal, helping rule out bacterial infection. On further questioning, he mentioned a history of recurrent sinusitis. With involvement of multiple organ systems, we suspected vasculitis. Serology confirmed the diagnosis when cytoplasmic ANCA (c-ANCA) came back positive.

He was started on high-dose methylprednisolone and rituximab, but the response was limited. We then added plasmapheresis. After three sessions, there was a clear improvement in his hemoptysis and kidney function.

This case illustrates how challenging it can be to diagnose GPA in tuberculosis-endemic regions and emphasizes the need to keep vasculitis in mind when patients present atypically and do not respond as expected to standard treatment.

Introduction

Granulomatosis with polyangiitis (GPA) is a necrotizing granulomatous vasculitis predominantly affecting small- to medium-sized vessels and is strongly associated with cytoplasmic-antineutrophil cytoplasmic antibodies (C-ANCA) directed against proteinase 3 (PR3)[1,2]. The classic clinical triad includes upper-airway involvement (chronic sinusitis, otitis, nasal crusting), lower-respiratory-tract disease (hemoptysis, pulmonary nodules or hemorrhage) and glomerulonephritis[3]. Pulmonary involvement occurs in up to 90 % of cases and can be life-threatening when diffuse alveolar hemorrhage (DAH) develops[4]. In tuberculosis-endemic countries such as Nepal, GPA is frequently misdiagnosed as pulmonary tuberculosis because of overlapping radiographic findings and nonspecific respiratory symptoms[5,6]. Delayed diagnosis can lead to irreversible organ damage[7]. We present a rare case of GPA that initially masqueraded as pulmonary tuberculosis, progressed to severe DAH and Acute kidney injury, and responded dramatically to plasmapheresis after failure of first-line immunosuppressive therapy.

Case Presentation

A 66-year-old previously healthy Nepali male presented to the emergency department of Sukraraj Tropical and Infectious Disease Hospital,Kathmandu,Nepal with a two-week history of moderate hemoptysis. He reported a several-year history of recurrent sinusitis treated symptomatically with antibiotics and nasal sprays. Initial evaluation revealed moderate anemia, leukocytosis with Neutrophilia(Table 1) and a chest radiograph showing a homogeneous opacity in the middle zone of right lung suggestive of consolidation or mass. Sputum acid-fast bacilli smear was negative on two occasions ,Sputum GeneXpert MTB did not detect MTB ,however, given the endemic setting and radiographic appearance,

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the patient was empirically started on IV antibiotics with a presumptive diagnosis of Community Acquired Pneumonia to rule out Pulmonary Tuberculosis.

Over the next 10 days, hemoptysis worsened (now frank and voluminous), and he developed hypertensive emergency(blood pressure 200/110 mmHg) that was difficult to control with multiple agents. Laboratory evaluation revealed rising serum creatinine from a baseline of 1.4 mg/dL to 5.3 mg/dL, active urinary sediment with dysmorphic red blood cells and proteinuria, and normal procalcitonin level.(Table 2) Repeat chest radiograph demonstrated persistence and slight enlargement of the right-lung homogeneous mass and similar mass in left lung (Figure 1). Contrast-enhanced CT chest confirmed bilateral patchy ground-glass opacities and consolidation consistent with alveolar hemorrhage, without evidence of cavitary lesions or tree-in-bud opacities typical of active tuberculosis (Figure 2). Bronchoscopy was performed after stabilization of BP which did not reveal mass.Bronchoalveolar lavage did not show AFB or fungus on culture . Given the constellation of recurrent sinusitis, alveolar hemorrhage, rapidly progressive acute kidney injury alongwith hematuria, and negative infectious work-up, a clinical diagnosis of ANCA-associated vasculitis was suspected. Serum C-ANCA (PR3) returned strongly positive (titer 1:123), while P-ANCA and anti-GBM antibodies were negative.(Table 3) Native kidney biopsy showed rapidly progressive glomerulonephritis.(Figure 3)

Date	White blood cells	Neutrophil	Hemoglobin	Platelets	Others
Day 1	22,310	93%	9.32	7,54,000	Lympho-cytes 3%
Day 7	21,460	89%	8.23	6,79,000	Lympho-cytes 4%
Day 30	15,400	90%	9.2	3,74,000	Down–trending Neutrophil count
Reference range	4.0-11.0 $\times 10^3/\mu L$	40-70%	12-18 gm/dL	150-450 $\times 10^3/\mu L$	

Table 1: Hematologic profile of the patient tracking key parameters over a 30-day period

Day	Creati-nine	Urea	Sodium	Potassi-um	Cor-rected Calcium	Uric acid	Others
Day 1	5.3	73	108	4.3	9.2	5	CRP = 169
Day 7	4.7	89	110	4.5			D dimer = <0.1
Day 30	1.7	70	138	3.27			TFT = Normal
Day 90	1.4	35	135	3.5			FBS = 77 mg/dl
Reference range	0.6-1.4 mg/dL	15-45 mg/dl	135-145meq/dl	3.5-5.5 meq/dl	8-11 mg/dl	<7 mg/dl	ESR = 30 mm/1sthr

Table 2: Metabolic and Inflammatory profile monitoring renal function

Test name	Result
C-ANCA	Positive
P-ANCA	Negative
GBM antigen	Negative
ANA	Negative
Sputum AFB	Negative
Sputum GeneXpert MTB	Not detected
BAL AFB	Negative
HIV serology	Negative
IGRA test	Negative
Influenza test	Negative
SARS COV 2 antigen	Negative
Procalcitonin	Normal

Table 3: Summary of Laboratory test at presentation

ANA: antinuclear antibodies, ANCA: antineutrophil cytoplasmic antibodies,GBM:Glomerular basement membrane,AFB:Acid bacillus bacillus,MTB:mycobacterium tuberculosis, HIV: human immunodeficiency virus, IGRA: interferon-gamma release assay,

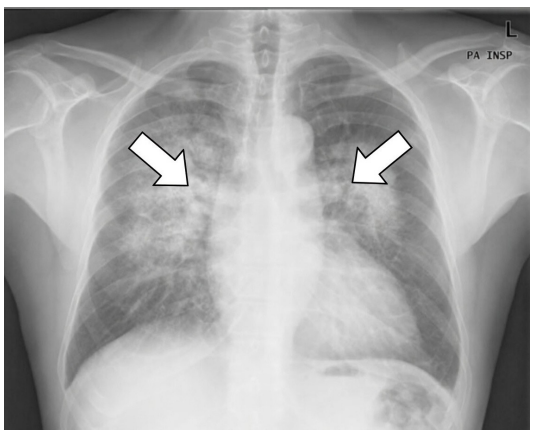

Figure 1: Posterior–anterior chest radiograph demonstrating homogenous opacity in middle zone of both lung

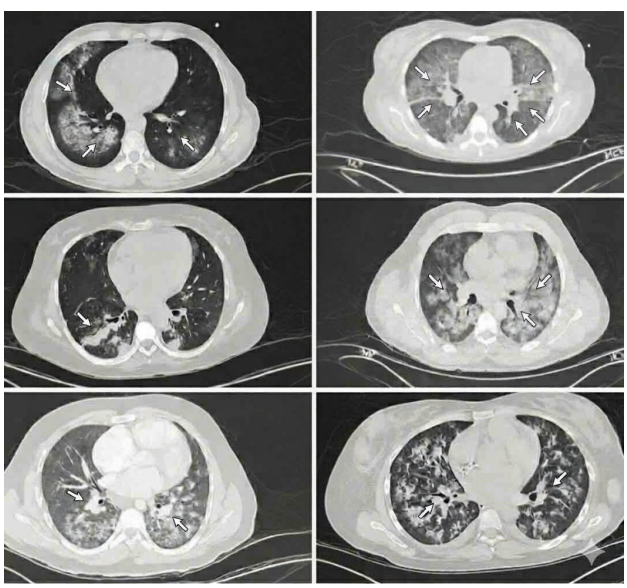

Figure 2: CECT scan Chest demonstrating Diffuse alveolar hemorrhage

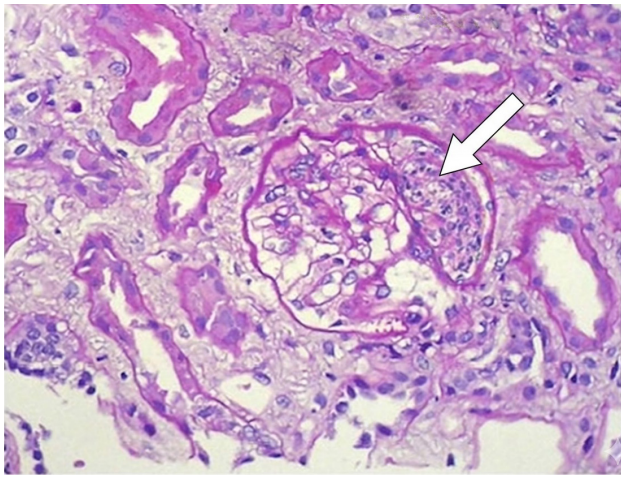

Figure 3: Kidney biopsy showing rapidly progressive (crescentic) glomerulonephritis.

The patient was immediately started on pulse intravenous methylprednisolone (1 g/day for three days) followed by high-dose oral prednisolone (1 mg/kg/day) and rituximab (375 mg/m² weekly for four doses). Despite this induction regimen, hemoptysis persisted and renal function did not improve. After multidisciplinary discussion with rheumatology and nephrology, therapeutic plasmapheresis (three plasma-volume exchanges) was instituted on alternate days for three sessions. Within 48 hours of initiating plasmapheresis, hemoptysis markedly decreased and ceased completely by the third session. Serum creatinine improved to 1.7 mg/dL within one month and stabilized at 1.4 mg/dL at third month of discharge. The patient was transitioned to oral prednisolone with tapering and continued rituximab maintenance. He remained asymptomatic at one-month follow-up with normalized renal function and no recurrence of hemoptysis.

The patient provided written informed consent for the publication of this case report and any accompanying images.

Discussion

This case highlights the diagnostic challenges of granulomatosis with polyangiitis (GPA) in tuberculosis-endemic regions. The patient initially presented with hemoptysis and a right lung mass that closely mimicked post-primary tuberculosis—a common finding reported in several studies.[8,9] Recurrent sinusitis, rapidly progressive glomerulonephritis, normal procalcitonin levels, and ultimately positive C-ANCA were the critical clues that steered the diagnosis toward vasculitis. Differentiating the two conditions on imaging can be difficult[9]; however, the appearance of diffuse ground-glass opacities consistent with pulmonary hemorrhage on CT (as opposed to cavitary or tree-in-bud patterns typical of TB) helped refine the differential once clinical worsening occurred.[10,11]

Standard induction therapy for organ-threatening GPA consists of high-dose glucocorticoids plus either cyclophosphamide or rituximab.[12] Rituximab has demonstrated non-inferiority to cyclophosphamide in the RAVE trial and is often preferred due to its more favorable safety profile.[13] In this patient, however, there was no significant clinical or laboratory improvement after starting steroids and rituximab, which prompted escalation to plasmapheresis. Although the PEXIVAS trial questioned the routine use of plasma exchange in ANCA-associated vasculitis[14], subgroup analyses and real-world experience support its role as rescue therapy in life-threatening diffuse alveolar hemorrhage (DAH) refractory to immunosuppression[15]. Our patient showed rapid recovery after just three sessions, supporting emerging evidence that plasmapheresis can quickly remove pathogenic ANCA antibodies and inflammatory mediators, thus bridging the time until rituximab achieves its full effect[16-18].

This case emphasizes the need to maintain a high index of suspicion for GPA even when initial findings suggest an infectious cause. Early ANCA testing in patients with unexplained pulmonary-renal syndrome can speed up diagnosis and help prevent irreversible organ damage.

Conclusions

In regions where tuberculosis is highly prevalent, Granulomatosis with Polyangiitis (GPA) can easily be mistaken for pulmonary TB because their clinical presentations overlap so closely. Clinicians should maintain a high index of suspicion for GPA when a patient presents with a combination of hemoptysis, sinusitis and kidney dysfunction.

When diffuse alveolar hemorrhage (DAH) occurs, ordering a prompt C-ANCA test, aggressively treating with immunosuppressants and moving to plasmapheresis if refractory to standard therapy can be life saving. This case report contributes valuable insights to the limited medical literature surrounding GPA patients who are initially misdiagnosed with TB, highlighting how plasma exchange can successfully rescue a patient when rituximab therapy fails to yield a complete response.

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: Prasanna Bahadur Amatya

Acquisition, analysis, or interpretation of data: Milan Bajracharya

Drafting of the manuscript: Prasanna Bahadur Amatya

Critical review of the manuscript for important intellectual content: Milan Bajracharya,Kashyap Dahal

Supervision: Samrat Shrestha

Disclaimer: AI-assisted tools (e.g.Grammarly) were used solely for grammar correction and language polishing. The authors reviewed and edited all content and take full responsibility for the final manuscript.

References

1. Yaseen K, Mandell BF: ANCA associated vasculitis (AAV): a review for internists. Postgrad Med. 2023, 135:3-13. 10.1080/00325481.2022.2102368
2. Yoo J, Kim HJ, Ahn SS, Jung SM, Song JJ, Park YB, Lee SW: The utility of the ACR/EULAR 2017 provisional classification criteria for granulomatosis with polyangiitis in Korean patients with antineutrophil cytoplasmic antibody-associated vasculitis. Clin Exp Rheumatol. 2018, 36:85-87.
3. McDonald JB, Edwards RW.: "Wegener's Granulomatosis"—A Triad. JAMA. 1960, 173:1205-1209. 10.1001/jama.1960.03020290031006

4. Arora R: Massive alveolar haemorrhage: a rare life threatening complication of Wegener's granulomatosis—report of a rare case.Quant. Imaging Med Surg. 2014, 4:424-427. 10.3978/j.issn.2223-4292.2014.07.03
5. Bam N, Shrestha B, Pokhrel M: Mishra AK.Granulomatosis with Polyangiitis Masquerading as Recurrent Tuberculosis and Non-Resolving Pneumonia: A rare case report.Journal of. Advances in Internal Medicine. 2025:86-90. 10.3126/jaim.v14i2.88405
6. Mahmood FS, Schwartz E: Kurup S, et al.A diagnostic dilemma: differentiating between granulomatosis with polyangiitis and tuberculosis.Clinical Medicine. 2013:411-413. 10.7861/clinmedicine.13-4-411
7. Sánchez Álamo B, Moi L, Bajema I etal EUVAS: Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis. Nephrol Dial Transplant. 2023, 30:1655-1665. 10.1093/ndt/gfac320
8. Gayathri D, Ralapanawa U, Shyamali I etal: Navigating a Diagnostic Dilemma: A Case Report of Overlapping Presentation of Granulomatosis With Polyangiitis and Tuberculosis. Cureus. 2025, 19:84378. 10.7759/cureus.84378
9. Martins JD, Soares C, Barreiros C etal : Granulomatosis With Polyangiitis Misdiagnosed as Tuberculosis: A Case Report.. Cureus. 2024, 16:e71834.. 10.7759/cureus.71834
10. Zhu Y, Zhu J, Luo R etal: Granulomatosis with polyangiitis. IDCases. 2024, 16:01970. 10.1016/j.idcr.2024.e01970
11. Miller WT Jr, Panosian JS: Causes and imaging patterns of tree-in-bud opacities. Chest. 2013, 144:1883-92. 10.1378/chest.13-1270
12. Kronbichler A, Bajema IM, Bruchfeld A etal: Diagnosis and management of ANCA-associated vasculitis. Lancet. 2024, 17:683-698. 10.1016/s0140-6736(23)01736-1
13. Carranza-Enríquez F, Meade-Aguilar JA, Hinojosa-Azaola A: Rituximab treatment in ANCA-associated vasculitis patients: outcomes of a real-life experience from an observational cohort. Clin Rheumatol. 2022, 41:2809-2816. 10.1007/s10067-022-06192-1
14. Bensalem A, Mulleman D, Paintaud G etal: Non-Linear Rituximab Pharmacokinetics and Complex Relationship between Rituximab Concentrations and Anti-Neutrophil Cytoplasmic Antibodies (ANCA) in ANCA-Associated Vasculitis: The RAVE Trial Revisited. Clin Pharmacokinet. 2020, 59:519-530. 10.1007/s40262-019-00826-5
15. Fussner LA, Flores-Suárez LF, Cartin-Ceba R etal ; PEXIVAS Investigators: Alveolar Hemorrhage in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: Results of an International Randomized Controlled Trial (PEXIVAS). Am J Respir Crit Care Med. 2024, 1:1141-1151. 10.1164/rccm.202308-1426oc
16. Toyoda T, Yates M, Watts RA: Is There Still a Role of Plasma Exchange in the Current Management of ANCA-Associated Vasculitides?. Curr Rheumatol Rep. 2022, 24:111-117. 10.1007/s11926-022-01064-8
17. Hellmich B, Sanchez-Alamo B, Schirmer JH etal: EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update. Ann Rheum Dis. 2024, 2:30-47. 10.1136/ard-2022-223764
18. Dhooria A, Naidu GSRSNK, Misra DP etal: Indian Rheumatology Association guidelines for the management of ANCA associated vasculitis. Autoimmun Rev. 2024, 23:103647. 10.1016/j.autrev.2024.103647