ISSN: 0975-3583,0976-2833 VOL15, ISSUE 11, 2024

Granulomatosis with Polyangiitis Presenting with Diffuse Alveolar Hemorrhage: A Rare Case Report

Dr Darshan S Likhiya[1], Dr Chandni S Likhiya[2], Dr Meet M Agola[3], Dr Chirag G Adroja[4]

[1]MBBS, DMRE, P D U Medical College, Rajkot, Gujarat, India

[2]MBBS, DDV, P D U Medical College, Rajkot, Gujarat, India

[3]MBBS, B J Medical College, Ahmedabad, Gujarat, India

[4]MBBS MD medicine, C U Shah Medical College, Surendranagar, Gujarat, India

Corresponding Author: Dr Darshan S Likhiya

ABSTRACT

Granulomatosis with Polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare systemic necrotizing vasculitis predominantly affecting small to medium-sized blood vessels, with a reported annual incidence of approximately 10-20 cases per million. The disease is often characterized by necrotizing granulomatous inflammation that can involve multiple organs, most commonly the lungs and kidneys. This case report presents an unusual instance of GPA with diffuse alveolar hemorrhage in an 18-year-old male, highlighting the importance of early diagnosis, comprehensive clinical evaluation, and multidisciplinary management in achieving optimal patient outcomes.

INTRODUCTION

Granulomatosis with Polyangiitis (GPA) is a potentially life-threatening autoimmune disorder primarily involving the respiratory tract and kidneys, with pulmonary and renal involvement being hallmark features (1). It is part of the ANCA-associated vasculitis (AAV) spectrum, with the presence of c-ANCA and PR3-ANCA often aiding in diagnosis. Despite being well-documented, GPA remains rare, and presentations with diffuse alveolar hemorrhage (DAH) are exceedingly uncommon, particularly in younger individuals (2). DAH, marked by rapid onset of hemoptysis, dyspnea, and respiratory compromise, requires prompt recognition and treatment due to its associated high mortality. Here, we report a rare case of GPA with DAH and multisystem involvement in an adolescent male, underscoring the critical role of early detection and intervention.

CASE PRESENTATION

An 18-year-old male presented to the emergency department with complaints of persistent cough, progressive dyspnea, low-grade fever, generalized weakness, and headaches for the past four weeks. His medical history revealed a recent viral infection, one month prior, for which

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 11, 2024

he received a blood transfusion. He also reported bilateral facial swelling, worsening exertional dyspnea, pleuritic chest pain, abdominal discomfort, and maculopapular rashes localized to his shins. Family history was significant for systemic lupus erythematosus in the maternal grandmother.

Physical Examination

On physical examination, the patient appeared moderately ill, with stable vital signs aside from mild tachycardia and elevated respiratory rate. Bilateral pitting edema of the face was noted, alongside maculopapular rashes on both shins and decreased breath sounds in the left lung field.

Laboratory Findings

- **Erythrocyte Sedimentation Rate (ESR)**: 90 mm/hr, indicating significant inflammation.
- **Urinalysis**: +3 albumin, with 10-12 pus cells and numerous red blood cells (RBCs), suggestive of renal involvement.
- **Immunologic Studies**: c-ANCA and PR3-ANCA were strongly positive, consistent with a diagnosis of GPA.

Imaging

- **Chest X-ray**: Demonstrated bilateral, multifocal nodular opacities.
- **High-Resolution CT (HRCT) Thorax**: Showed multiple nodular, ground-glass, and consolidatory opacities predominantly in the left lung, along with minimal left-sided pleural effusion, suggestive of diffuse alveolar hemorrhage.
- **CT Brain**: No acute intracranial abnormalities, though findings included left maxillary sinus disease.

Diagnosis

Based on the clinical presentation, serological findings, and imaging, a diagnosis of GPA with diffuse alveolar hemorrhage was established.

DISCUSSION

Granulomatosis with Polyangiitis is an inflammatory disorder characterized by necrotizing granulomatous inflammation and small-vessel vasculitis, primarily affecting the respiratory tract and kidneys. Environmental triggers, the presence of autoantibodies (c-ANCA and PR3-ANCA), and an overactive immune response with excessive cytokine release play significant roles in its pathogenesis. Although most common in middle-aged adults, GPA can present in young patients and its presentation with diffuse alveolar hemorrhage is rare, especially in the adolescent age group (3).

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 11, 2024

Pulmonary Involvement: Pulmonary manifestations, including nodules, cavitary lesions, and diffuse alveolar hemorrhage, are frequent in GPA. Diffuse alveolar hemorrhage is an acute, often life-threatening complication due to rapid bleeding into the alveolar spaces, necessitating urgent care (4,5).

Renal Involvement: The patient's renal findings, including proteinuria and hematuria, indicate active renal disease, a common feature in GPA. Renal involvement often leads to rapidly progressive glomerulonephritis, which underscores the importance of vigilant renal monitoring (6).

Dermatologic and Sinonasal Involvement: The maculopapular rash observed on the patient's shins and sinus involvement are reflective of GPA's systemic nature. Although less frequent, dermatologic findings such as palpable purpura and rashes are well-documented in GPA (7).

Management and Treatment

Given the patient's active symptoms and potential infection risk, induction therapy was initiated with rituximab, an anti-CD20 monoclonal antibody. Rituximab is increasingly favored in GPA treatment due to its efficacy and reduced risk of adverse effects compared to cyclophosphamide, which was avoided in this case due to concerns regarding infection susceptibility. Adjunctive high-dose corticosteroids were also administered to control inflammation and provide rapid immunosuppression (8).

Alternative Therapies: Other immunosuppressants, including methotrexate and azathioprine, as well as plasma exchange, may be considered in refractory cases or those with severe organ involvement (8).

Multidisciplinary Approach: Management involved collaboration between pulmonologists, nephrologists, and immunologists to ensure comprehensive care and regular monitoring. Routine follow-ups and periodic imaging are crucial to monitor disease progression and response to treatment.

CONCLUSION

This case illustrates a rare presentation of GPA with diffuse alveolar hemorrhage in a young male patient, highlighting the diagnostic and therapeutic challenges associated with this form of ANCA-associated vasculitis. Early recognition of GPA, with prompt initiation of immunosuppressive therapy, is crucial to prevent irreversible organ damage and improve prognosis. Given the multisystem involvement in GPA, a multidisciplinary approach and vigilant follow-up are essential for optimal management and to minimize the risk of relapse.

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 11, 2024

REFERENCES

1. Comarmond C, Cacoub P. Granulomatosis with polyangiitis (Wegener): clinical aspects and treatment. Autoimmun Rev. 2014 Nov;13(11):1121-5. doi: 10.1016/j.autrev.2014.08.017. Epub 2014 Aug 20. PMID: 25149391.
2. Qasim A, Patel JB. ANCA-Associated Vasculitis. [Updated 2024 Aug 31]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK554372/
3. Potentas-Policewicz M, Fijolek J. Granulomatosis with polyangiitis: clinical characteristics and updates in diagnosis. Front Med (Lausanne). 2024 Aug 27;11:1369233. doi: 10.3389/fmed.2024.1369233. PMID: 39257888; PMCID: PMC11385631.
4. Lohrmann C, Uhl M, Kotter E, Burger D, Ghanem N, Langer M. Pulmonary manifestations of wegener granulomatosis: CT findings in 57 patients and a review of the literature. Eur J Radiol. 2005 Mar;53(3):471-7. doi: 10.1016/j.ejrad.2004.04.016. PMID: 15741022.
5. Sacoto G, Boukhlal S, Specks U, Flores-Suárez LF, Cornec D. Lung involvement in ANCA-associated vasculitis. Presse Med. 2020 Oct;49(3):104039. doi: 10.1016/j.lpm.2020.104039. Epub 2020 Jul 7. PMID: 32650042.
6. Masiak A, Zdrojewski Z, Pęksa R, Smoleńska Ż, Czuszyńska Z, Siemińska A, Kowalska B, Stankiewicz C, Rutkowski B, Bułło-Piontecka B. The usefulness of histopathological examinations of non-renal biopsies in the diagnosis of granulomatosis with polyangiitis. Reumatologia. 2017;55(5):230-236. doi: 10.5114/reum.2017.71638. Epub 2017 Oct 28. PMID: 29332961; PMCID: PMC5746633.
7. Manfredi A, Cassone G, Izzo R, Dallagiacoma G, Felicetti M, Cariddi A, Berti A, Giollo A, Nannini C, Bettio S, Monti S, Conticini E, Govoni M, Quartuccio L, Argolini LM, Kaleci S, Emmi G, Dejaco C, Padoan R, Dagna L, Rossini M, Cantini F, Montecucco C, Frediani B, De Vita S, Caporali R, Muratore F, Sebastiani M, Salvarani C; Study Group of the Italian Society for Rheumatology (SIR). Interstitial lung disease in microscopic polyangiitis and granulomatosis with polyangiitis: demographic, clinical, serological and radiological features of an Italian cohort from the Italian Society for Rheumatology. Clin Exp Rheumatol. 2023 Apr;41(4):821-828. doi: 10.55563/clinexprheumatol/xu4hmh. Epub 2022 Oct 6. PMID: 36200955.

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 11, 2024

8. Berti A, Caporali R, Montecucco C, Paolazzi G, Monti S. Aging in Primary Systemic Vasculitis: Implications for Diagnosis, Clinical Manifestations, and Management. Drugs Aging. 2019 Jan;36(1):53-63. doi: 10.1007/s40266-018-0617-4. PMID: 30484240.