

Case Report

Granulomatosis with polyangiitis masquerading as community acquired pneumonia: a case report

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ABSTRACT

A 32-year-old female with a known history of hypothyroidism presented with a 10-day history of fever, cough, fatigue, and breathlessness, along with initial symptoms of myalgia and headache. There was no prior history of chronic respiratory illness or tuberculosis exposure. Clinical examination revealed fever, tachypnoea, hypoxia, and pallor, with localized coarse crepitations over the right lung fields. Laboratory evaluation showed anemia, neutrophilic leukocytosis, elevated CRP and ESR, and a negative procalcitonin. COVID-19 rapid antigen test was positive. Chest X-ray revealed right-sided consolidation with bilateral patchy opacities. Despite supportive care and broad-spectrum antibiotics, the patient deteriorated, requiring non-invasive ventilation. Chest CT revealed bilateral subpleural and peribronchial consolidations with cavitation and mediastinal lymphadenopathy. Microbiological workup including blood/sputum cultures and CBNAAT was negative. Bronchoscopy showed mucopurulent secretions, but BAL panel was negative. Autoimmune workup was unremarkable. CT-guided lung biopsy revealed granulomatous inflammation with vasculitis, confirming granulomatosis with polyangiitis (GPA). The patient showed marked clinical and radiological improvement with corticosteroids and Rituximab.

Keywords: Community acquired pneumonia, Granulomatosis with polyangiitis, Non-resolving pneumonia, Vasculitis

INTRODUCTION

Respiratory illnesses with fever, cough, and breathlessness pose a diagnostic challenge due to overlapping presentations of infectious, inflammatory, and autoimmune conditions. A systematic approach, including clinical evaluation, imaging, and laboratory investigations, is crucial for accurate diagnosis and timely management.

This is the case of a 32-year-old female who presented with features suggestive of community acquired pneumonia. However, further workup and lung biopsy revealed an autoimmune etiology, leading to a significant clinical improvement with immunosuppressive therapy. This case highlights the importance of considering vasculitis in patients with atypical pulmonary presentations.

CASE REPORT

32-year-old female with hypothyroidism presented with a 10-day history of fever, cough, fatigue, and breathlessness. She also gives history of myalgia, headache in the initial days of illness. No history of prior chronic respiratory illness, weight loss, night sweats or contact with tuberculosis. Examination revealed fever (temperature: 101 °F), tachypnoea (respiratory rate: 24/minute), hypoxia (spo₂: 92% in room air) and pallor. She didn't have any generalised lymphadenopathy or clubbing. Upper respiratory tract was normal. On auscultation, reduced air entry with coarse crepitations were present on the right side- axillary, infra-axillary and infrascapular areas. Initial workup showed mild anemia (Hb: 10.8), neutrophilic leukocytosis (Total count: 18500; N63.2%, L26.7%, E5.9%), and elevated inflammatory markers (CRP:47.5,

ESR:90). Platelet count was normal. Serum procalcitonin was negative. COVID-19 rapid antigen test was positive. Chest X-ray showed consolidation of right middle and lower lobes with bilateral patchy non-homogenous opacities. Liver and renal function tests were normal. Patient was admitted and was started on oxygen support, broad spectrum parenteral antibiotics and other supportive measures. CT imaging revealed no features suggestive of COVID-19. It revealed subpleural and peribronchial consolidatory patches in bilateral lung parenchyma, largest in right middle and lower lobes. Focal tiny cavitation was seen within the consolidatory patch of right lower lobe, mediastinal lymphadenopathy was also seen.

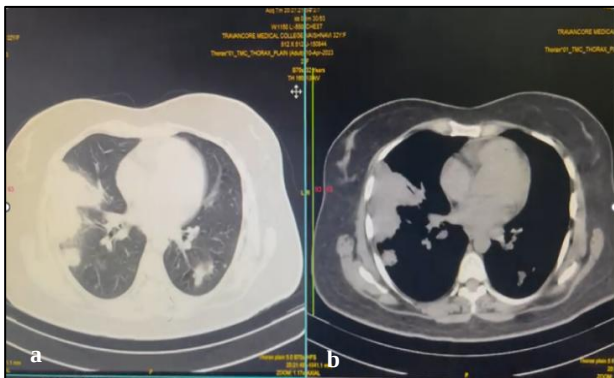


Figure 1 (a and b): CT image of right middle and lower lobe consolidation.

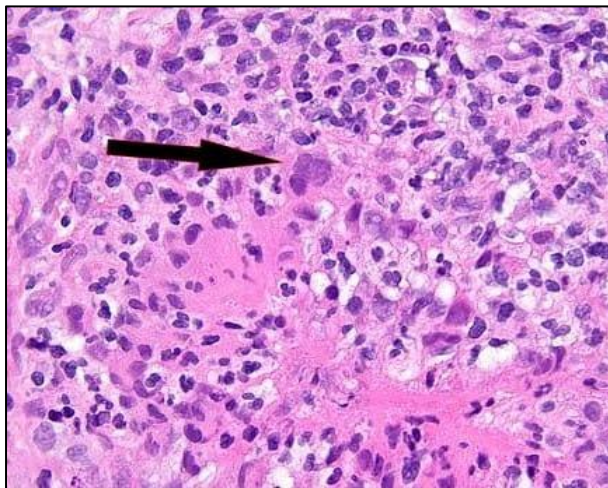


Figure 2: Lung biopsy showing necrotizing granuloma.

Blood and sputum cultures, sputum CBNAAT were negative. On subsequent days patient had clinical deterioration requiring NIV support. Repeat imaging showed radiological worsening also. She was started on parenteral steroids and antibiotics were hiked. Bronchoscopy was performed, which showed features suggestive of infection (mucopurulent secretions/ no endobronchial growth), but the BAL-Biofire pneumonia panel was negative. ANA profile, S.ACE, c-ANCA and p-ANCA were negative. CT-guided lung biopsy was

performed, which showed zones of granulomatous inflammation with vasculitis consistent with granulomatosis with polyangiitis. The patient improved significantly on steroids and Rituximab with radiographic resolution on follow up.

DISCUSSION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare, necrotizing vasculitis primarily affecting small to medium-sized vessels. It commonly involves the upper respiratory tract (sinuses, nose), lungs and kidneys, leading to inflammation and granuloma formation with a characteristic association to antineutrophil cytoplasmic antibodies (ANCA), particularly anti-proteinase 3 (PR3) antibodies. Despite its well-defined clinical features, GPA can present in diverse and atypical manners, often leading to diagnostic challenges.¹

This is a unique case of GPA mimicked as community-acquired pneumonia. The patient had consolidation with fever, cough, breathlessness, and neutrophilic leucocytosis. Our patient didn't have any renal or upper respiratory tract involvement, either. Initial investigations, including a positive COVID-19 rapid antigen test, led to a presumptive diagnosis of COVID-19 pneumonia or a secondary bacterial pneumonia. However, the patient's clinical deterioration and radiological findings prompted further evaluation. CT imaging findings were atypical for COVID-19 pneumonia and raised suspicion for an alternative diagnosis.

Our patient presented with non-resolving pneumonia, having failed to respond to initial treatment. The underlying causes of non-resolving pneumonia can generally be divided into two broad categories: infectious and non-infectious. Approximately 40% of cases are due to infectious etiologies, which may include primary infections, persistent infections, or nosocomial (hospital-acquired) infections.² Approximately 20% of presumed non-resolving pneumonias are due to non-infectious causes.³ In such cases, further evaluation with advanced radiologic imaging, non-invasive sampling, and bronchoscopy is recommended to assess the lungs more thoroughly and to collect specimens for microbiological testing and other diagnostic analyses.⁴

Similar cases have been reported where GPA mimicked infectious pneumonia, leading to initial misdiagnosis and delayed treatment.⁵ Reports of atypical presentations of GPA mimicking cardiovascular, or gastrointestinal conditions are also there.⁶ There are case reports where TB and GPA coexist.⁷ Treatment recommendations in GPA includes induction of remission with rituximab or cyclophosphamide (CYC) and steroids and maintenance with azathioprine or steroids or mycophenolate mofetil.⁸ In this case, the lack of a clear infectious etiology, combined with the biopsy findings, helped guide the diagnosis toward a vasculitis process rather than an

infectious one. The patient's response to steroids and Rituximab further supports the diagnosis of GPA. Diagnosing GPA often requires a high level of clinical suspicion, as its initial symptoms are typically non-specific. Most patients receive a diagnosis between 3 to 12 months after symptom onset.⁹ This case emphasizes the importance of a multidisciplinary approach, including imaging, histopathological analysis, and the use of targeted therapies, in reaching a final diagnosis in complex cases with overlapping clinical features.

CONCLUSION

This case underscores the importance of considering autoimmune diseases like granulomatosis with polyangiitis in the differential diagnosis of lung lesions, especially when infectious causes are excluded. GPA should be included in the differential diagnosis for any case of necrotizing pneumonia that does not respond to standard treatment.¹⁰ It also highlights the potential for rapid clinical improvement with immunosuppressive therapy in appropriately diagnosed cases, particularly with Rituximab in severe GPA. Further studies, including the exploration of the role of early lung biopsy in cases of suspected vasculitis, are needed to refine the diagnostic approach to such complex presentations.

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