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Pulmonary Presentation of Microscopic Polyangiitis Mimicking Pneumonia: A Case Report and Review of the Literature

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Abstract

Microscopic polyangiitis (MPA) is a rare subtype of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), characterized by pauci-immune necrotizing inflammation of small vessels, primarily affecting the kidneys, lungs, skin, and peripheral nervous system.

Keywords: Microscopic polyangiitis (MPA); Alongside granulomatosis polyangiitis (GPA); eosinophilic granulomatosis polyangiitis (EGPA)

Introduction

Microscopic polyangiitis (MPA) is a rare subtype of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV), characterized by pauci-immune necrotizing inflammation of small vessels, primarily affecting the kidneys, lungs, skin, and peripheral nervous system [1]. Alongside granulomatosis with polyangiitis (GPA) and eosinophilic granulomatosis with polyangiitis (EGPA), MPA has an estimated annual incidence of 20–30 cases per million population [2]. Myeloperoxidase (MPO)-ANCA positivity is a hallmark of MPA, distinguishing it from proteinase 3 (PR3)-ANCA-associated GPA in most cases [3]. Pulmonary involvement occurs in up to 70% of MPA cases, manifesting as diffuse alveolar haemorrhage, interstitial lung disease, or focal consolidations that mimic infectious pneumonia [4]. These overlapping clinical and radiological features frequently lead to misdiagnosis, particularly in elderly patients with comorbidities, delaying therapy and increasing the risk of irreversible organ damage [5]. Untreated AAV carries a mortality rate exceeding 80% within one year, highlighting the urgency of early diagnosis [6]. This case report describes a patient with MPA presenting as persistent pneumonia and reviews similar cases to highlight diagnostic challenges, clinical clues, and optimal management strategies.

Case Presentation

Patient information

Mr. A.B., a 73-year-old Caucasian male with a history of hypertension and stage 3 chronic kidney disease (baseline eGFR 40 mL/min/1.73m²), presented to Mater Private Hospital Mackay, Queensland, Australia, with a three-week history of progressive lethargy, reduced exercise tolerance, exertional dyspnoea, and a productive cough with white sputum. He denied haemoptysis, haematuria, fevers, night sweats, weight loss, or recent travel. His medications included amlodipine 5 mg daily and perindopril 4 mg daily. Mr A.B. was a non-smoker with no occupational exposures or known allergies.

Clinical Findings

On examination, the patient was afebrile (temperature 36.8°C), normotensive (blood pressure 132/78 mmHg), with a respiratory rate of 18 breaths/min and oxygen saturation of 96% on room air. Chest auscultation revealed decreased breath sounds and coarse

crackles in the left mid and lower zones. Cardiovascular examination identified a grade 3/6 pansystolic murmur radiating to the axilla, consistent with mild mitral regurgitation. There were no cutaneous lesions, joint swelling, or neurological deficits suggestive of systemic vasculitis.

Diagnostic Assessment

Initial laboratory results included:

- White cell count: 20.7 $\times 10^9$/L (neutrophils 16.2 $\times 10^9$/L)
- C-reactive protein (CRP): 76 mg/L (normal <5 mg/L) [7]
- Erythrocyte sedimentation rate (ESR): 92 mm/hr (normal <20 mm/hr)
- Urea: 16.6 mmol/L (normal 3.0–8.0 mmol/L)
- Creatinine: 167 µmol/L (eGFR 31 mL/min/1.73m², worsened from baseline 40 mL/min/1.73m²)
- Urinalysis: Proteinuria (5+), microscopic haematuria (242 $\times 10^6$/L erythrocytes), pyuria (209 $\times 10^6$/L leukocytes), and red cell casts [8]
- Haemoglobin: 112 g/L (normal 130–180 g/L)

Initial Imaging: Chest X-ray showed left-sided consolidation in the lingula, and high-resolution computed tomography (HRCT) confirmed left lingular consolidation with bronchial wall thickening, bibasal atelectasis, and ground-glass opacities [9]. No cavitary lesions, nodules, or pleural effusions were noted.

Initial Management and Progression

The patient was diagnosed with community-acquired pneumonia and treated with intravenous ceftriaxone (1 g daily) and azithromycin (500 mg daily). After five days, his dyspnoea worsened, and renal function declined further (creatinine 198 µmol/L, eGFR 26 mL/min/1.73m²) [10]. Repeat HRCT showed persistent left-sided consolidation with new patchy infiltrates in the right lower lobe.

Further Investigations

Additional workup included:

- Sputum culture: Negative for bacterial, fungal, and mycobacterial pathogens
- Blood cultures: No growth after 48 hours
- Autoimmune serology: Positive MPO-ANCA (titre 1:160), negative PR3-ANCA, negative antinuclear antibody (ANA), normal complement levels (C3, C4) [3]
- Renal ultrasound: Normal-sized kidneys, preserved cortical thickness, mild right perinephric fluid, no hydronephrosis
- Echocardiogram: Normal left ventricular systolic function (ejection fraction 71%), mild aortic sclerosis, no vegetations
- Bronchoscopy: was planned but could not be done because of urgency. It usually shows erythematous bronchial mucosa; bronchoalveolar lavage (BAL) usually shows hemosiderin-laden macrophages ruling out diffuse alveolar haemorrhage and no malignant cells ruling out malignancy [11].

Differential Diagnosis

The initial differential included:

1. Community-acquired pneumonia with sepsis-induced acute kidney injury
2. Atypical infections (e.g., tuberculosis, fungal pneumonia)
3. Pulmonary embolism with infarction
4. Pulmonary-limited vasculitis or systemic AAV
5. Malignancy (e.g., lung cancer, less likely given imaging)
6. Drug-induced interstitial lung disease (unlikely, no culprit medications)

Progressive renal impairment, active urinary sediment, and MPO-ANCA positivity favoured systemic vasculitis over infectious or malignant aetiologies.

Diagnosis

A working diagnosis of microscopic polyangiitis with pulmonary and renal involvement was established. A renal biopsy was scheduled to confirm pauci-immune necrotizing glomerulonephritis, the histopathological hallmark of MPA [12].

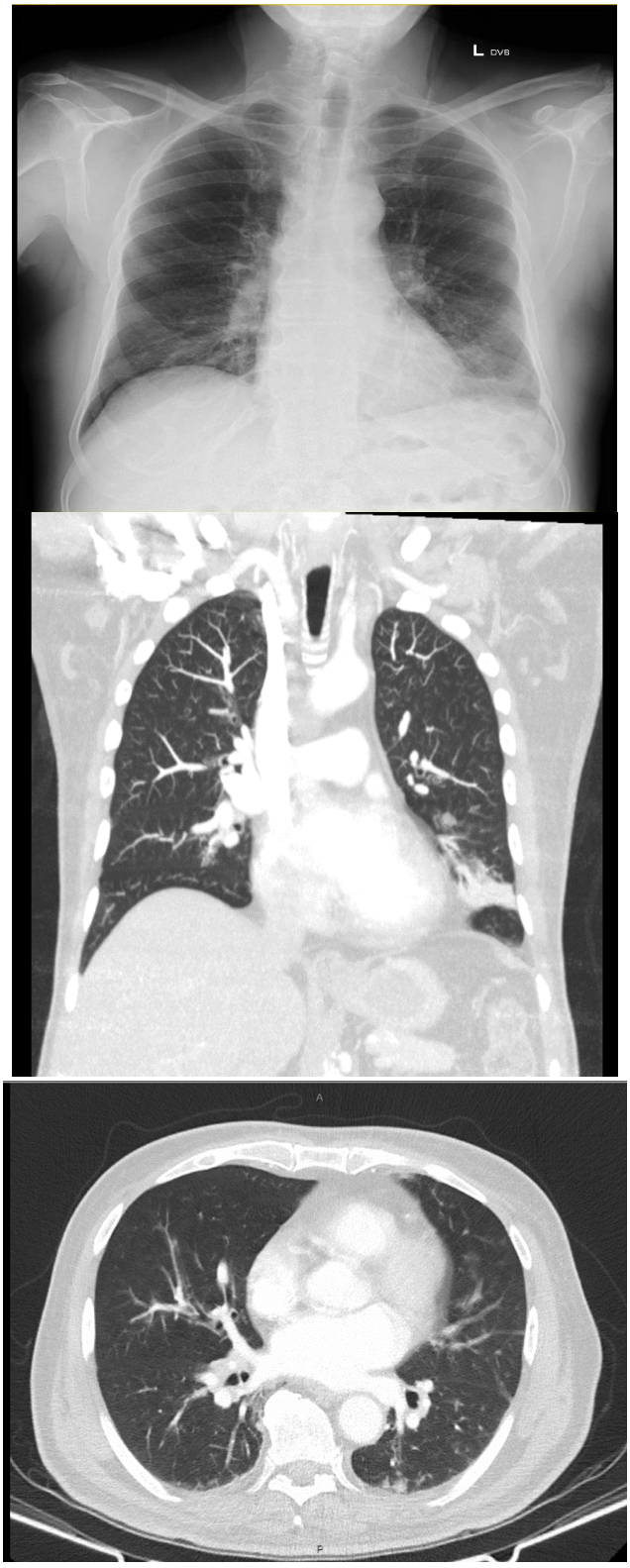
Management

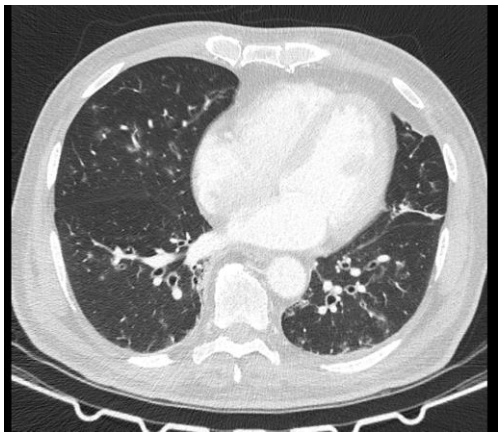
The patient was urgently referred to nephrology. Supportive care included discontinuation of perindopril to avoid further renal injury, optimization of fluid balance, and daily renal function monitoring. Pending biopsy results, the nephrology team planned induction therapy with high-dose corticosteroids (methylprednisolone 500 mg IV daily for 3 days, followed by prednisone 1 mg/kg/day) and rituximab (375 mg/m² weekly for 4 weeks) [13]. Plasmapheresis was considered for potential alveolar haemorrhage or severe glomerulonephritis. Prophylactic trimethoprim-sulfamethoxazole was planned to prevent Pneumocystis jirovecii pneumonia during immunosuppression [14]. The patient was counselled on the risks of therapy, including infections and corticosteroid-related side effects, and a multidisciplinary team (nephrology, respiratory, and rheumatology) was engaged to coordinate care.

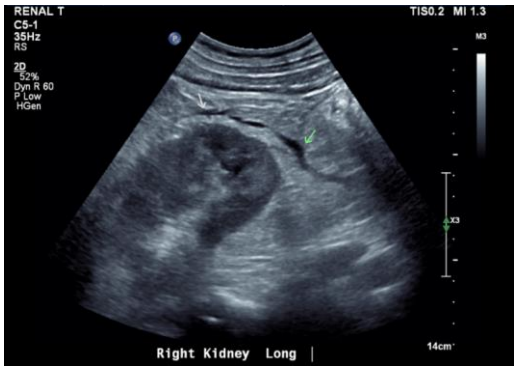
Discussion

Microscopic polyangiitis is a systemic small-vessel vasculitis with a predilection for renal and pulmonary involvement [7]. Pulmonary manifestations, seen in 10–30% of cases, range from subtle consolidations to life-threatening diffuse alveolar haemorrhage [15]. Radiologically, findings such as ground-glass opacities, consolidations, or interstitial patterns are non-specific, overlapping with infectious, neoplastic, or embolic processes [9].

This case illustrates the diagnostic challenge of pulmonary vasculitis mimicking community-acquired pneumonia, a common scenario in elderly patients with comorbidities [5].







Comparison with Other Case Reports

Several published cases highlight similar diagnostic challenges:

1. Chen described a 68-year-old woman with fever, cough, and bilateral infiltrates treated as pneumonia for 14 days. Persistent haematuria, declining renal function, and MPO-ANCA positivity led to an MPA diagnosis, confirmed by renal biopsy showing crescentic glomerulonephritis. The patient responded to cyclophosphamide and steroids [16].
2. Sato reported a 70-year-old man with unilateral consolidation and no initial renal symptoms, diagnosed as MPA via bronchoscopy and MPO-ANCA testing. Rituximab resolved pulmonary capillaritis [17].
3. Yamamoto et al. (2020) reviewed five cases of pulmonary vasculitis misdiagnosed as pneumonia, with delays of 7–28 days. Common features included non-resolving infiltrates, elevated CRP, and late renal involvement [18].
4. Watanabe-Imai described a 65-year-old man with isolated pulmonary consolidation as the sole MPA manifestation, diagnosed via transbronchial biopsy showing capillaritis. Corticosteroids alone achieved remission [19].
5. Kim reported a 72-year-old man with MPA presenting as organizing pneumonia. Progressive renal failure and MPO-ANCA positivity prompted immunosuppression, but the patient developed Aspergillus infection, highlighting treatment risks [20].

6. Lee described a 67-year-old woman with cough, dyspnoea, and bilateral consolidations initially treated as pneumonia. Persistent symptoms, haemoptysis, and MPO-ANCA positivity led to an MPA diagnosis after 10 days. Renal biopsy confirmed vasculitis, and rituximab with steroids improved outcomes [21].
7. Park reported a 75-year-old man with unilateral consolidation and mild haematuria, misdiagnosed as pneumonia for 12 days. MPO-ANCA positivity and renal biopsy confirmed MPA; plasmapheresis was used for alveolar haemorrhage, with good response [22].
8. Tanaka described a 69-year-old man with MPA presenting as interstitial lung disease mimicking atypical pneumonia. Delayed diagnosis (21 days) due to negative initial ANCA testing highlighted the need for repeat serology in suspected cases [23].

These cases consistently demonstrate:

- Poor response to antibiotics, with persistent or worsening infiltrates [18].
- Systemic inflammation (elevated CRP/ESR) disproportionate to infectious findings [7].
- Late emergence of renal abnormalities (haematuria, proteinuria, declining eGFR) [16].
- Positive MPO-ANCA serology, though occasionally delayed or initially negative [23].
- Diagnostic delays of 7–28 days, increasing the risk of organ damage [18].

Our patient's presentation aligns with these patterns, particularly the initial misdiagnosis of pneumonia, persistent symptoms despite antibiotics, and late detection of renal involvement. The absence of haemoptysis, as in Watanabe-Imai [19] and Sato [17], underscores that MPA can present with subtle pulmonary findings, complicating diagnosis.

Diagnostic Pitfalls and Clues

The overlap between pulmonary vasculitis and pneumonia poses significant diagnostic challenges. Key pitfalls include:

- Over-reliance on infectious aetiologies: Elderly patients with comorbidities are often presumed to have pneumonia, delaying ANCA testing [5].
- Non-specific imaging: Consolidations and ground-glass opacities are common to both MPA and infection [9].
- Delayed renal involvement: Renal abnormalities may emerge late, as seen in Sato [17] and our case.
- False-negative ANCA: Initial negative serology, as in Tanaka [23], may mislead clinicians, necessitating repeat testing.

Clinical clues to suspect MPA include:

- Non-resolving symptoms despite appropriate antibiotics [18].
- Active urinary sediment (red cell casts, haematuria) suggesting glomerulonephritis [8].
- Systemic inflammation (elevated CRP/ESR) without clear infectious source [7].
- MPO-ANCA positivity, though PR3-ANCA may occasionally be seen in MPA [3].

A systematic diagnostic approach should include:

- Serology: ANCA (MPO, PR3), ANA, and complement levels to exclude other vasculitides [3].
- Imaging: HRCT chest to characterize pulmonary patterns [9].
- Renal evaluation: Urinalysis for active sediment and renal function monitoring [8].
- Invasive procedures: Bronchoscopy with BAL to rule out infection or haemorrhage; renal biopsy to confirm pauci-immune glomerulonephritis [12].

Renal biopsy is the gold standard, typically showing focal or diffuse crescentic glomerulonephritis with minimal immune deposits [12]. In our case, the planned biopsy was critical to confirm MPA and guide therapy.

Management Strategies

Treatment of MPA aims to induce remission, prevent relapses, and minimize complications. Current guidelines recommend:

- Induction therapy: High-dose corticosteroids (methylprednisolone 500–1000 mg IV daily for 3 days, followed by prednisone 1 mg/kg/day) combined with rituximab (375 mg/m² weekly for 4 weeks) or cyclophosphamide (15 mg/kg IV every 2–3 weeks) [13]. Rituximab is preferred for its efficacy and lower toxicity, as demonstrated in the RAVE trial [24]. Our patient's planned regimen aligns with this approach.
- Plasmapheresis: Indicated for severe alveolar haemorrhage or rapidly progressive glomerulonephritis, as in Park [22]. It removes circulating ANCA and inflammatory mediators but carries risks of bleeding and infection [25].
- Maintenance therapy: Azathioprine (2 mg/kg/day) or methotrexate (20–25 mg/week) for 12–24 months to prevent relapses, as shown in the MAINRITSAN trial [26].
- Supportive care: Discontinuation of nephrotoxic drugs, blood pressure control, and prophylaxis against opportunistic infections (e.g., trimethoprim-sulfamethoxazole for Pneumocystis jirovecii) [14].

Monitoring and Complications: Close monitoring is essential due to the high risk of treatment-related complications:

- Infections: Immunosuppression increases susceptibility to infections, as seen in Kim et al., where the patient developed Aspergillus infection [20]. Prophylactic antibiotics and antifungal agents are critical [14].

- Corticosteroid side effects: Osteoporosis, diabetes, and hypertension require regular screening and management [13].
- Renal outcomes: Up to 30% of MPA patients progress to end-stage renal disease despite treatment [6]. Early therapy, as planned in our case, improves renal survival.
- Relapses: Relapse rates are 20–40% within 5 years, necessitating long-term follow-up [26].

Multidisciplinary Approach: Effective management requires collaboration between nephrologists, rheumatologists, respiratory physicians, and pathologists. In our case, the multidisciplinary team ensured timely biopsy planning, treatment initiation, and infection prophylaxis. Patient education on therapy risks and adherence is also critical.

Lessons Learned

1. Non-resolving pneumonia with systemic or renal abnormalities should prompt ANCA testing [18].
2. Repeat serology may be needed if initial ANCA is negative, as in Tanaka [23].
3. Renal biopsy is essential for definitive diagnosis and to exclude mimics (e.g., anti-GBM disease) [12].
4. Early immunosuppression prevents organ damage, but infection prophylaxis and monitoring are vital [14].
5. Multidisciplinary care optimizes outcomes in complex cases [13].

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