Multiple Cavitary Lung Lesions in ANCA-Negative Small Vessel Vasculitis: A Case Report

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ABSTRACT

Multiple cavitary lung lesions present a diagnostic challenge, with a broad differential diagnosis, including infectious, malignant, and autoimmune etiologies. ANCA-associated vasculitis (AAV), particularly in its seronegative form, is a rare yet important cause that is frequently overlooked, as pulmonary involvement is uncommon and its manifestations can closely mimic infection or malignancy. We report a case of a 30-year-old male who presented with a three-week history of dyspnea and hemoptysis, and a background of membranoproliferative glomerulonephritis confirmed by renal biopsy three months prior, consistent with pauci-immune glomerulonephritis. Chest radiography and contrast-enhanced CT revealed multiple bilateral cavitary lung lesions with air-fluid levels, micronodules, and centrilobular ground-glass opacities. Extensive infectious workup, including sputum and bronchoalveolar lavage cultures, acid-fast bacilli smear, GeneXpert, and fungal studies, was negative. Cytological examination excluded malignancy. Both p-ANCA and c-ANCA were negative. Based on the 2022 ACR/EULAR classification criteria, a diagnosis of ANCA-negative microscopic polyangiitis (MPA) was established, with a total score of +6 derived from imaging findings consistent with interstitial lung disease and histopathological evidence of pauci-immune glomerulonephritis in a prior renal biopsy. Immunosuppressive therapy with methylprednisolone and mycophenolate sodium was initiated, resulting in complete radiological resolution of all cavitary lesions within two months. This case highlights that cavitary lung lesions may represent a rare pulmonary manifestation of ANCA-negative MPA. A negative ANCA result should not exclude vasculitis when clinical suspicion is high, and tissue biopsy remains essential for definitive diagnosis in atypical presentations.

Keywords*: Anti-neutrophil cytoplasmic antibody-associated vasculitis, microscopic polyangiitis, antibodies, antineutrophil cytoplasmic, lung diseases, cavitary lung lesions.*

INTRODUCTION

Multiple cavitary lung lesions pose a diagnostic challenge in clinical practice, with a differential diagnosis spanning infectious, neoplastic, and autoimmune etiologies.[1] Among these, ANCA-associated vasculitis (AAV), a group of systemic autoimmune diseases characterized by necrotizing inflammation of small blood vessels, represents an important yet frequently overlooked consideration.[2] AAV encompasses three subgroups: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA).[3-4]

Among all systemic vasculitides, AAV is the most common cause of pulmonary manifestations, with presentations ranging from interstitial lung disease to diffuse alveolar hemorrhage.[5] The development of cavitary lung lesions within this context, however, is a rare occurrence that often mimics infectious or malignant disease, making early recognition difficult. This diagnostic challenge is further complicated when ANCA serology returns negative.[6] Although ANCA is detectable in the majority of AAV cases, seronegative disease has been documented, and in such cases, diagnosis confirmation is based on clinical assessment and histopathological examination.[7] A negative ANCA result in the setting of an atypical presentation, such as multiple pulmonary cavitations, can lead to substantial diagnostic delays and deferred immunosuppressive therapy, with considerable consequences for patient outcomes.

We report a case of a 30-year-old male who presented with multiple bilateral pulmonary cavitary lesions against a background of pre-existing membranoproliferative glomerulonephritis. Infectious and malignant workup was negative, and ANCA serology was also negative for both p-ANCA and c-ANCA. He was ultimately diagnosed with ANCA-negative MPA at a tertiary referral center in Indonesia. This case draws attention to the importance of including AAV, even in its seronegative form, in the differential diagnosis of progressive multiple cavitary lung lesions that fail to respond to antimicrobial therapy, and reinforces the role of tissue biopsy in reaching a timely and accurate diagnosis.

CASE ILLUSTRATION

A 30-year-old male presented to the emergency department at RSUPN Cipto Mangunkusumo with a three-week history of dyspnea and hemoptysis. One month before admission, he started to develop a cough, fever, arthralgia, hair loss, and rash initially involving the axillary regions, which subsequently spread to the upper and lower extremities. The patient had a prior smoking history of one pack per day but had ceased smoking two months before presentation. The patient had an underlying diagnosis of nephrotic syndrome secondary to membranoproliferative glomerulonephritis, diagnosed by biopsy three months before admission. Renal biopsy revealed membranoproliferative glomerulonephritis with focal endocapillary proliferation and fibrocellular crescent formation in 1 of 16 glomeruli (6%), with minimal immune deposits of IgA and IgM and absent IgG and C3, consistent with pauci-immune glomerulonephritis. At presentation, he was receiving oral methylprednisolone of 16-8-8 mg daily and mycophenolate sodium (Myfortic) 720 mg twice daily for his kidney condition.

On physical examination, the patient exhibited signs of mild distress, with a heart rate of 111 beats/minute, respiratory rate of 24 breaths/minute, oxygen saturation of 94% on room air, and was afebrile. Lung auscultation revealed bilateral coarse crackles. Laboratory evaluation demonstrated hemoglobin 8.6 g/dL, leukocyte count 16,900/µL, and platelet count 160,000/µL. Differential counts showed basophils 0.6%, eosinophils 0.1%, neutrophils 85.1%, lymphocytes 7.0%, and monocytes 7.2%, indicating leukocytosis with marked neutrophilia and relative lymphopenia. Renal function tests revealed urea 46.9 mg/dL, creatinine 0.76 mg/dL, and eGFR 122.4 mL/min/1.73m², indicating preserved renal function despite elevated urea levels. The patient was also found to have hyperglycemia, with a random blood glucose level of 381 mg/dL and an HbA1c of 7.8%**.** Chest radiography performed in the emergency department revealed multiple cavitary lesions in both lungs (**Figure 1**).

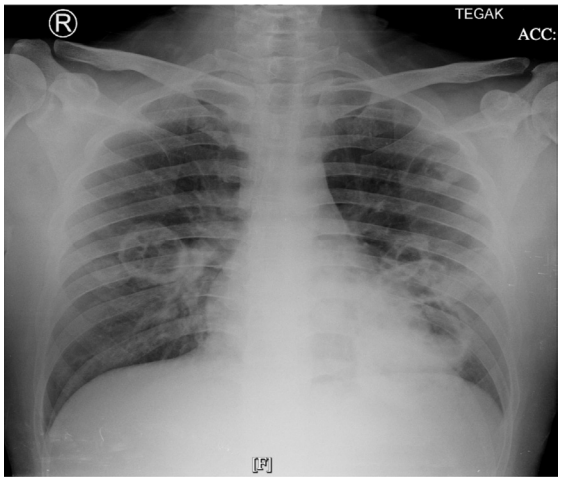


Figure 1. Initial chest radiography revealed multiple cavitary lesions in both lung fields.

The initial working diagnosis was multiple lung abscesses caused by atypical bacterial infection, with differential diagnoses including fungal infection and pulmonary tuberculosis. The patient was admitted for inpatient management and empirically treated with intravenous ampicillin-sulbactam 1.5 g four times daily and fluconazole 200 mg once daily. Tranexamic acid 500 mg was administered as needed for hemoptysis, along with other supportive symptomatic therapies. His existing methylprednisolone regimen was continued, while mycophenolate sodium was temporarily withheld because of suspected active infection.

During hospitalization, further diagnostic workup was performed. Contrast-enhanced thoracic computed tomography (CT) demonstrated multiple bilateral cavitary lung lesions with air-fluid levels of varying sizes, suggestive of multiple abscess formations, accompanied by multiple micronodules and centrilobular nodular ground-glass opacities in both lungs (**Figure 2)**. Acid-fast bacilli (AFB) smear and GeneXpert testing of sputum were negative for Tuberculosis. Sputum aerobic culture isolated *Acinetobacter junii.* KOH staining and fungal culture of the sputum were negative. Anti-HIV testing was nonreactive. Bronchoscopy revealed normal intraluminal findings in the lower respiratory tract. Bronchoalveolar lavage (BAL) specimens obtained during bronchoscopy were negative for bacterial and fungal pathogens on staining and culture examinations. BAL galactomannan testing was also negative. Cytological examination of the BAL specimen demonstrated no evidence of malignant tumor cells. A pulmonary biopsy was not performed at that time, as the clinical presentation was initially attributed to an infectious etiology and tissue sampling was not yet deemed necessary.

Follow-up chest radiography after seven days of antibiotic and antifungal therapy

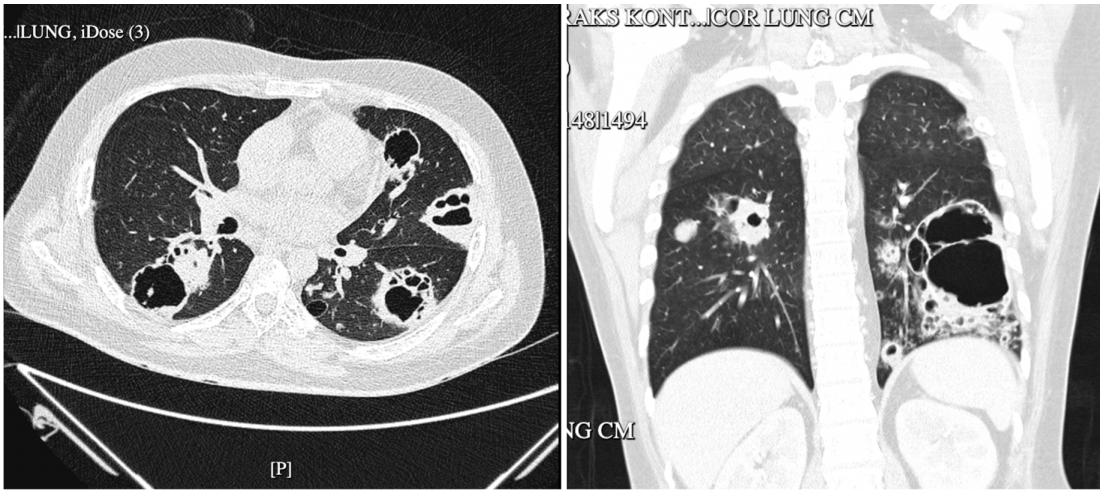


Figure 2. Contrast-enhanced chest CT at initial presentation demonstrating multiple round cavitary lesions, with irregular margins, thick walls, and non-uniform sizes, distributed across segments of both lungs. The dominant lesion is located in the left lung, comprising a large cavity surrounded by coalescent smaller cavities measuring 6 × 7.2 × 12 cm with air-fluid levels. Multiple subcentimeter cavities with similar characteristics and centrilobular micronodules are distributed throughout both lungs, most prominently surrounding the dominant lesion. Diffuse ill-defined centrilobular nodular ground-glass opacities are noted predominantly in the upper lobes of both lungs, suggestive of interstitial lung disease.

showed multiple bilateral cavitary lesions with air-fluid levels that remained relatively stable in size and number (**Figure 3**). After 12 days of hospitalization, the patient was discharged with improvement in dyspnea, although intermittent hemoptysis persisted. Discharge medications included oral amoxicillin-clavulanate, fluconazole, and tranexamic acid.

As the multiple cavitary lesions did not improve significantly with antimicrobial therapy and recurrent hemoptysis persisted, the patient was referred to the Allergy and Clinical Immunology division to evaluate for possible vasculitis, particularly given his systemic manifestations and prior renal involvement. Autoimmune biomarker profile evaluation demonstrated weakly positive lupus anticoagulant, negative anticardiolipin IgM, and normal complement C4 levels. Serologic testing for anti-glomerular basement membrane (anti-GBM), cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA), and perinuclear antineutrophil cytoplasmic antibody (p-ANCA) were all negative. The patient was subsequently clinically diagnosed with ANCA-negative small-vessel vasculitis. Mycophenolate sodium 720 mg twice daily was reinitiated, and the methylprednisolone regimen was maintained due to suspected ongoing active pulmonary vasculitic involvement.

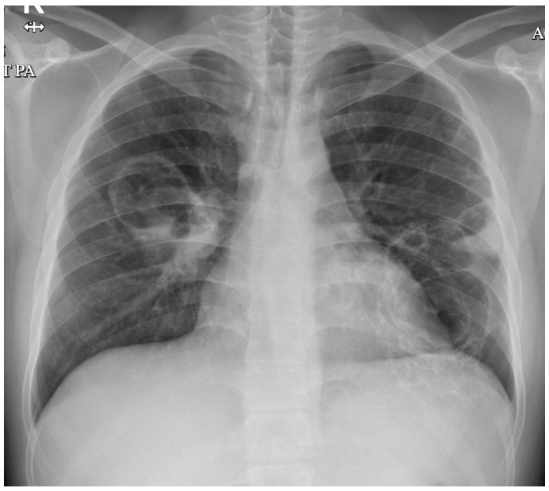


Figure 3. Follow-up chest radiography after one week of therapy revealed multiple cavitary lesions in both lung fields that remained relatively unchanged compared with the previous examination.

A repeat thoracic CT scan performed two months after initiation of immunosuppressive therapy demonstrated complete resolution of the cavitary lesions, with residual fibrotic changes in the previously affected areas (**Figure 4**). Clinically, the patient reported complete resolution of dyspnea and hemoptysis. Mycophenolate sodium at the dose being administered at the time was planned to be continued for at least two years until complete remission was achieved, with routine follow-up evaluations using thoracic CT scans every six months.

DISCUSSION

ANCA-associated vasculitis (AAV) is a group of systemic autoimmune diseases characterized by necrotizing inflammation of small blood vessels, manifesting in a wide spectrum of clinical features depending on

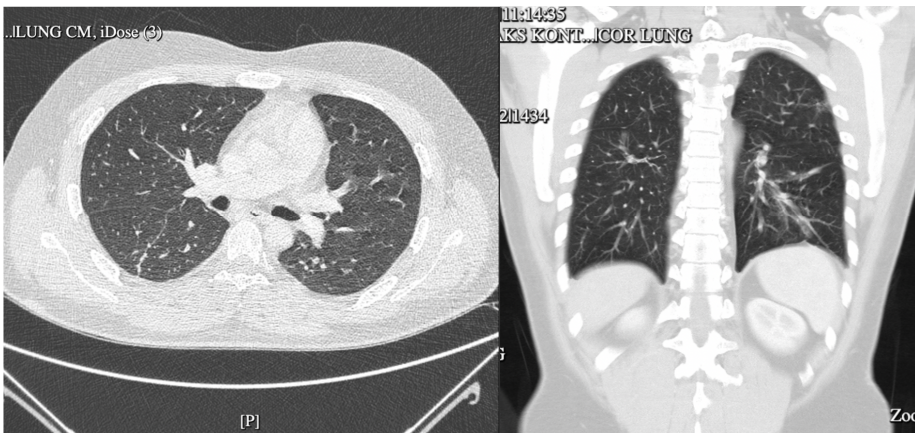


Figure 4. Follow-up thoracic CT scan obtained two months after initiation of immunosuppressive therapy showing resolution of the lung cavitary lesions, with residual fibrosis in the previously involved regions.

the organ system involved. Among all forms of systemic vasculitis, ANCA-associated vasculitis is notably the most frequent cause of pulmonary manifestations.[8] It encompasses three major disease subtypes: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA).[4] ANCA-associated vasculitis is an inherently uncommon condition, with a global pooled incidence of approximately 17.2 per million person-years.[9] Within this already rare population, ANCA-seronegative cases constitute only around 9% of all diagnoses. When further considering that cavitary pulmonary lesions represent merely 1.7% of pulmonary ANCA-associated vasculitis manifestations, as reported in studies of Chinese adult populations, the clinical scenario encountered in the present case emerges as a remarkably uncommon presentation.[10]

The cavitary formation in ANCA-associated vasculitis involves multiple overlapping mechanisms. In seropositive disease, circulating ANCA activates primed neutrophils, triggering the release of reactive oxygen species and proteolytic enzymes that damage small pulmonary vessel walls. However, vascular inflammation can also occur without ANCA, as cytokine signaling, dysregulated T-cell activity, B cells, anti-endothelial cell antibodies, and complement activation have each been implicated in driving endothelial injury through ANCA-independent pathways.[1,12] The resulting sustained vascular damage leads to tissue necrosis and liquefaction; as necrotic debris is expelled through the bronchial tree, an air-filled cavity forms within the lung parenchyma. This explains how cavitation may arise in both ANCA-positive and ANCA-negative small vessel vasculitis.[13-14]

The present case involves a male patient who developed multiple pulmonary cavitary lesions alongside respiratory complaints spanning at least three weeks. The temporal course of cavitary lung disease carries important diagnostic implications, as lesions evolving over less than twelve weeks tend to reflect acute infectious processes such as bacterial or fungal lung abscess, necrotizing pneumonia, or septic embolism. In contrast, a more chronic course is more suggestive of mycobacterial infection, underlying malignancy, or systemic autoimmune disease. Beyond chronology, the morphological pattern of the lesions further refines the differential diagnosis: A solitary cavity more commonly points toward primary lung cancer or lung abscess, while multiple bilateral cavities raise concern for systemic autoimmune conditions or metastatic spread. Additionally, in countries with a high prevalence of tuberculosis, such as Indonesia, pulmonary cavities mandate thorough investigation for mycobacterial infection. Overall, the differential diagnosis of multiple cavitary lung lesions must systematically address infectious, malignant, and autoimmune etiologies.[15-16]

In this patient, the initial presentation of hemoptysis, dyspnea, and laboratory findings of leukocytosis with neutrophilia raised suspicion for an infectious etiology, particularly atypical bacterial pneumonia, fungal infection, or pulmonary tuberculosis, given the subacute symptom course. However, repeated microbiological evaluation, including for tuberculosis, yielded no positive results. The patient also failed to demonstrate clinical improvement despite appropriate antibiotic and antifungal therapy. Malignancy was subsequently excluded following cytological analysis that revealed no evidence of malignant cells. As none of the workup results supported an infectious or malignant etiology, an autoimmune process was increasingly suspected, a suspicion further supported by a prior history of renal involvement.

In patients presenting with respiratory symptoms and pulmonary lesions that fail to improve despite antibiotic therapy, ANCA-associated vasculitis should be considered in the differential diagnosis. A comprehensive autoimmune workup, especially ANCA screening, alongside transthoracic biopsy are essential steps toward diagnosing vasculitis. In this patient, both p-ANCA and c-ANCA were performed and returned negative. Nonetheless, a negative ANCA result may not exclude ANCA-associated vasculitis diagnosis when clinical suspicion remains high, as ANCA negativity has been reported in 5–10% of GPA and MPA

cases and in up to 70% of EGPA cases. ANCA diagnostic accuracy also varies by disease subtype, in which PR3-ANCA (typically associated with c-ANCA) demonstrates higher sensitivity for GPA than MPA (74% vs. 11%), whereas MPO-ANCA (commonly associated with p-ANCA) is more sensitive for MPA than GPA (73% vs. 7%). These figures highlight that ANCA serology should complement, rather than replace, comprehensive clinical and histopathological evaluation.[17-18]

Pulmonary biopsy was not performed in this case during the admission, as the initial presentation was attributed to infection. Such diagnostic uncertainty is not uncommon; up to 73% of patients with systemic vasculitis are initially given an alternative diagnosis, most frequently infection. The diagnostic challenge was further heightened by the unusual presentation; there is even a report of pulmonary cavities ultimately diagnosed as concurrent ANCA-associated vasculitis and non-tuberculous mycobacterial infection, an overlap that underscores how vasculitis may be overlooked in this setting. More importantly, diagnostic delays in vasculitis have been associated with negative health consequences, and early tissue biopsy should be prioritized in cases of multiple cavitary lung lesions that do not respond to antimicrobial therapy.[19-21]

In this patient with small vessel vasculitis, a diagnosis of microscopic polyangiitis (MPA) was established. MPA is a subtype of ANCA-associated vasculitis characterized by necrotizing inflammation predominantly affecting small vessels with few or no immune deposits, most commonly involving the kidneys and lungs, with approximately 9% of cases being ANCA-seronegative. Applying the 2022 ACR/EULAR classification criteria (**Table 1**), this patient demonstrated diffuse nodular ground-glass opacities in both lungs on contrast-enhanced chest CT consistent with interstitial lung disease (+3), alongside a renal biopsy showing membranoproliferative glomerulonephritis with zero-to-low immune deposits, consistent with pauci-immune glomerulonephritis (+3). The absence of nasal involvement (0), negative c-ANCA and p-ANCA (0), and a normal eosinophil count (0) yielded a total score of +6, meeting the threshold for an MPA classification.[22]

Kidney involvement is one of the most frequent manifestations of MPA, occurring in up to 90% of patients in the form of glomerulonephritis. In this patient, a prior renal biopsy demonstrating pauci-immune glomerulonephritis had been performed before presentation, which ultimately directed clinical suspicion toward an underlying vasculitic process. Beyond renal disease, pulmonary involvement is found in up to 40% of MPA patients, with ground-glass opacities, reticular shadowing, and bronchiectasis among the most commonly reported CT findings. Notably, the predominant pulmonary manifestation in this patient, multiple bilateral cavitary lesions, is atypical for MPA and is far more characteristically associated with GPA. The combination of ANCA seronegativity, multiple pulmonary cavities, and concurrent renal involvement renders this case a rare and unusual presentation within the MPA spectrum.[23-25]

The management of ANCA-associated vasculitis, encompassing both MPA and GPA, follows a two-phase therapy approach based on EULAR recommendations: induction and

Table 1. Classification criteria for microscopic polyangiitis.[22]

Clinical criteria	
Nasal involvement: Bloody discharge, ulcers, crusting, congestion, blockage, or septal defect/perforation	-3
Laboratory, imaging, and biopsy criteria	
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies ANCA positive	+6
Fibrosis or interstitial lung disease on chest imaging	+3
Pauci-immune glomerulonephritis on biopsy	+3
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

maintenance. For nonsevere ANCA-associated vasculitis, defined as disease that is not organ- or life-threatening, glucocorticoids in combination with rituximab, methotrexate, or mycophenolate mofetil are recommended, while severe disease warrants glucocorticoids combined with rituximab or cyclophosphamide.[4] According to the 2024 KDIGO guidelines, ANCA-negative patients are to be managed using the same treatment principles as ANCA-positive cases, although dedicated studies for this subgroup remain lacking. Despite presenting with multiple pulmonary cavities and glomerulonephritis, this patient was clinically classified as nonsevere AAV, given hemodynamic stability, preserved eGFR, and the intermittent and non-massive nature of the hemoptysis. Given a prior history of immunosuppressive therapy, treatment was initiated with methylprednisolone and mycophenolate sodium in accordance with the nonsevere treatment recommendation.[26]

Approximately 74% of ANCA-associated vasculitis patients achieve remission following induction therapy.[27] Treatment response is generally assessed over the first several months, with remission formally evaluated at six months of induction. By consensus definition, response is defined by a reduction in disease activity of at least 50% without the emergence of new manifestations, while remission means absence of signs or symptoms.[4] In pulmonary ANCA-associated vasculitis, this is typically reflected by symptomatic improvement and radiological regression of pulmonary lesions on serial imaging. In this case, complete resolution of all cavitary lesions was observed within two months of initiating immunosuppressive therapy, a response that further supported the diagnosis of active pulmonary vasculitis. Unfortunately, pulmonary involvement in ANCA-associated vasculitis is associated with significantly higher rates of mortality and relapse compared to disease without pulmonary manifestations. Given that pulmonary involvement in ANCA-associated vasculitis carries a significantly higher risk of mortality and relapse, adequate treatment and routine follow-up with serial radiological examination remain an integral part of ongoing disease monitoring in this patient. Accordingly, this patient had 24–48 months of immunosuppressive therapy planned with serial chest CT monitoring to ensure sustained remission and early detection of relapse.[2,27-28]

Take-Home Message

Multiple cavitary lung lesions carry a broad differential diagnosis, with bacterial infection and tuberculosis remaining the foremost consideration in high-prevalence regions. In cases of progressive pulmonary cavitation unresponsive to antibiotic therapy, particularly when accompanied by systemic manifestations, ANCA-associated vasculitis should be considered even in the absence of positive ANCA serology. Tissue biopsy remains the most critical diagnostic tool when the clinical setting allows. Given that pulmonary involvement in ANCA-associated vasculitis is associated with poorer outcomes, early diagnosis, adequate treatment, and close monitoring are essential to improving prognosis.

CONCLUSION

This case illustrates that multiple cavitary lung lesions, while most commonly attributed to infectious or malignant etiologies, may represent a rare manifestation of ANCA-negative small vessel vasculitis. In patients presenting with multiple pulmonary cavitation that fails to resolve despite adequate antimicrobial therapy, ANCA-associated vasculitis should remain a differential consideration regardless of seronegative ANCA status. Early recognition, prompt initiation of immunosuppressive therapy, and long-term monitoring are essential as pulmonary involvement in ANCA-associated vasculitis is associated with significantly higher rates of mortality and relapse. This case further adds to the limited body of literature on ANCA-negative MPA presenting with cavitary lung lesions and highlights the critical need to identify and manage this uncommon presentation.

CONFLICT OF INTERESTS

The authors declare no conflict of interest.

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