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(CASE REPORT)

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Microscopic polyangiitis: Diagnostic challenges and pulmonary manifestations: A case study

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Abstract

Microscopic polyangiitis (MPA) is a systemic necrotizing vasculitis that primarily affects small-caliber vessels. Previous studies have reported a pulmonary involvement frequency of 22% to 29%. Glomerulonephritis and alveolar hemorrhage are common features. It is usually associated with anti-neutrophil cytoplasmic antibodies (ANCA) directed against myeloperoxidase (MPO), which are considered pathogenic. The presence of positive p-ANCA is a guiding element in the diagnosis of MPA. We provide a detailed description of the clinical manifestations, diagnostic approach, and treatment of MPA in a 62-year-old man who presented with alveolar hemorrhage but no renal involvement. The treatment included the use of high-dose corticosteroids to suppress the autoimmune response. Life-threatening or organ-threatening diseases are treated with glucocorticoids and cyclophosphamide (in bolus). If renal function is preserved, methotrexate may be considered to induce remission, and maintenance therapy is recommended with azathioprine, while mycophenolate mofetil can be used as a second-line medication.

Keywords: Diffuse Alveolar Hemorrhage; Corticosteroids; Microscopic Polyangiitis; Ground-Glass Opacity; Corticosteroid-Sensitive

1. Introduction

Microscopic polyangiitis (MPA) is a vasculitis associated with ANCA. Its histological characteristic is necrotizing vasculitis of small vessels with few or no immune deposits (pauci-immune vasculitis). In Western countries, MPA has a lower prevalence than Wegener's disease, affects men more than women, and typically begins after the age of 50. The two organs most commonly affected, and often critical for prognosis, are the kidneys and the lungs. One of the most concerning clinical aspects of pulmonary alveolar hemorrhage (PAH) is its potentially life-threatening nature. PAH can also concurrently or sequentially affect other organs such as the nervous system, skin, musculoskeletal system, as well as the heart, eyes, and intestines. Therapeutic decisions should be based on the severity and pattern of organ involvement, adhering to the five-factor score (FFS). This case report presents an unusual clinical manifestation of PAH in a 62-year-old man exhibiting alveolar hemorrhage without renal involvement.

2. Case Presentation

We present the case of a 62-year-old patient, a former chronic smoker, with no notable medical history. The patient was hospitalized for an etiological workup of diffuse interstitial pneumonia, associated with chronic anemia in the context of general health deterioration. Upon examination, the patient reported stage III dyspnea according to Sadoul and low-grade hemoptysis for four months. The physical examination at admission revealed a conscious patient with pale skin and mucous membranes, a pale conjunctiva, tachycardia at 120 beats per minute, and a saturation of 97% on room air.

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Ear, pleuropulmonary and cardiac examinations were normal. A chest X-ray showed an interstitial syndrome with bilateral infiltrative opacities **(**Figure 1).

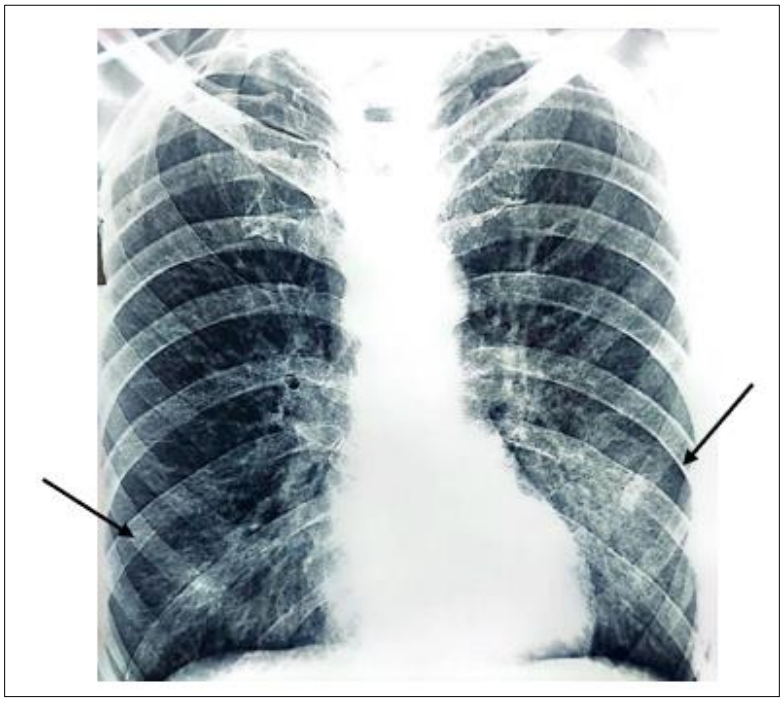

Figure 1 Posteroanterior chest X-ray demonstrating bilateral interstitial syndrome. The image shows diffuse bilateral infiltrative opacities predominantly in the middle and lower lung regions

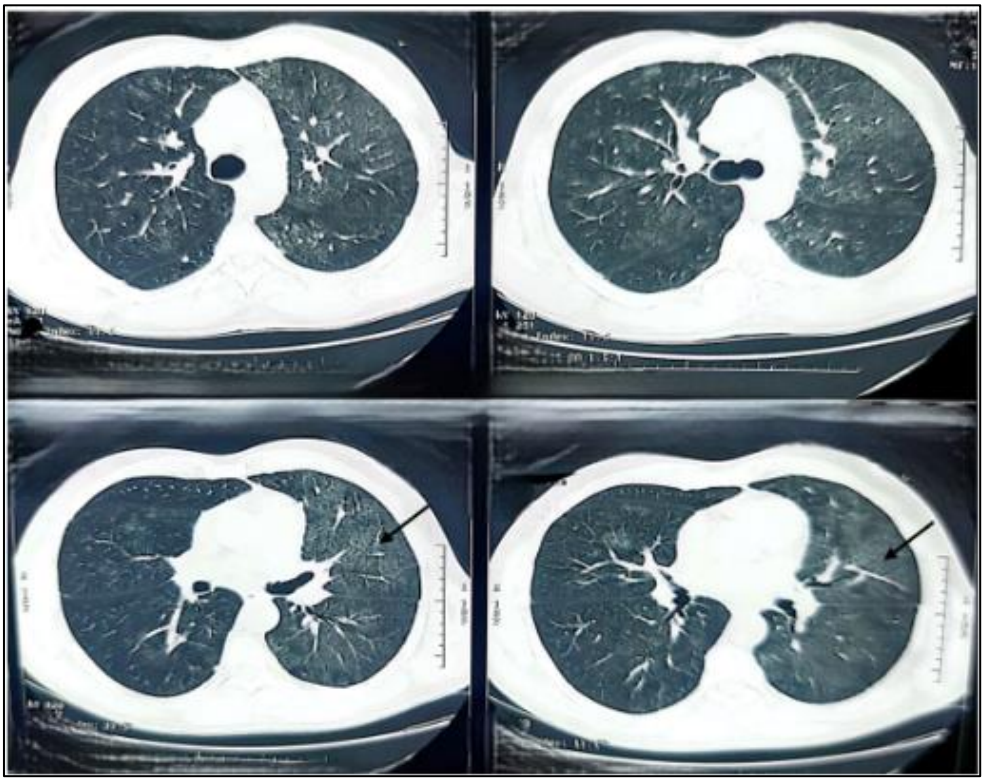

Figure 2 Chest computed tomography in the parenchymal window showing alveolar hemorrhage. Slices show diffuse bilateral ground-glass opacities with sparing of the subpleural region, suggestive of alveolar hemorrhage

A chest computed tomography (CT) scan showed diffuse bilateral ground-glass opacities more pronounced in the upper lobes with sparing of the subpleural region, suggesting alveolar hemorrhage (Figure 2). A biological assessment revealed hypochromic microcytic anemia, a negative C-reactive protein, a normal white blood cell count, negative 24-hour proteinuria, and normal blood urea levels. Sputum tests for mycobacteria were negative. An immunological evaluation revealed the presence of ANCA with cytoplasmic fluorescence (p-ANCA) (> 1/20 U), while c-ANCA was negative, strongly suggesting pulmonary alveolar hemorrhage (PAH).

Bronchoscopy with bronchoalveolar lavage (BAL) and Prussian blue staining revealed a macrophagic alveolitis with a predominance of histiocytes and hemosiderosis, with an estimated Golde score of 3. The results of the electromyography (EMG) and 24-hour proteinuria are within normal limits. The diagnosis of pulmonary alveolar hemorrhage (PAH) was based on the presence of suggestive clinical signs, including hemoptysis, anemia, and a characteristic appearance on the CT scan: multifocal ground-glass opacities with peripheral sparing and the presence of siderophages in the BAL. The diagnosis was also based on the classification criteria of the American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) 2022: presence of p-ANCA and radiographic abnormalities of the chest (≥ 5 points) (Figure 3).

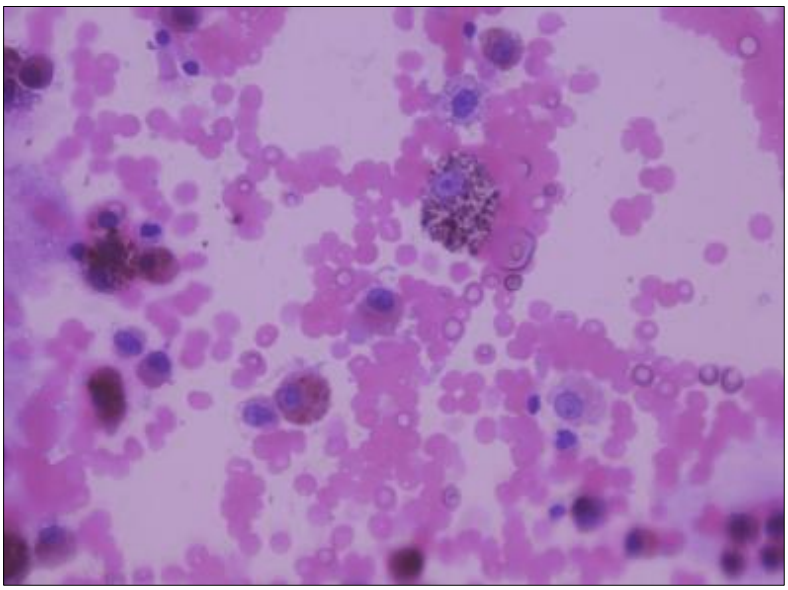

Figure 3 Bronchoalveolar lavage showing hemosiderosis. Bronchoalveolar lavage reveals a macrophagic predominant histiocytic alveolitis, with hemosiderosis. Prussian blue staining showed intense pigmentation of hemosiderin, with the nuclei of the histiocytes remaining visible. This pigmentation is mostly rated at 3 according to the Golde score and was evaluated on 100 histiocytic cells

The treatment is tailored to the severity of the disease, assessed by a prognostic score, the Five-Factor Score (FFS of 0). The patient received high-dose intravenous corticosteroids with methylprednisolone (1,000 mg/day) for three consecutive days. The treatment was then switched to oral prednisone (1 mg/kg/day), with a gradual dose reduction until a maintenance dose of 10 mg/day was reached. After the high-dose treatment, asthenia and cough resolved, with significant improvement in the ground-glass opacities observed on the chest CT scan, showing a reduction in both their extent and density. Following the definitive diagnosis of microscopic polyangiitis, the subsequent course was favorable, with good improvement in symptoms.

3. Discussion

Microscopic polyangiitis (MPA) is a rare systemic vasculitis that primarily affects small vessels, often manifesting as pulmonary and renal involvement. In the case presented, a 62-year-old patient developed alveolar hemorrhage, a symptom often associated with MPA, which warrants particular attention due to its clinical implications.

Alveolar hemorrhages in the context of MPA generally result from inflammation and necrosis of the pulmonary vessels. Studies show that nearly 70% of patients with MPA present with respiratory symptoms, including cough, dyspnea, and hemoptysis [1]. Alveolar hemorrhage, in particular, is a serious event that can lead to acute respiratory distress, necessitating prompt intervention [2].

Alveolar hemorrhage is a common and clinically significant complication in patients with microscopic polyangiitis (MPA). The typical thoracic radiological patterns of alveolar hemorrhage show focal or diffuse areas of opacification and/or ground-glass consolidation [3]. However, these findings are not specific, and other etiologies, such as pulmonary infections and interstitial pneumonia, present similar radiographic anomalies. Bronchoscopy is the best method for specifically diagnosing alveolar hemorrhage and for excluding other diseases [4]. Concurrently, renal involvement often manifests as glomerulonephritis, characterized by hematuria and proteinuria, which can progress to renal failure [5]. Renal biopsy is crucial for confirming the diagnosis, showing deposits of IgG and C3 associated with glomerular lesions.

In addition to pulmonary and renal involvement, microscopic polyangiitis (MPA) can also cause cutaneous, neurological, and gastrointestinal manifestations. Skin eruptions, such as purpura and nodules, are often observed [6]. In our case, the patient may also present with skin signs, indicating systemic involvement.

Neurological manifestations, although less common, can include peripheral neuropathies and, in severe cases, ischemic strokes due to cerebral vasculitis [7]. Gastrointestinal manifestations, while rare, can present as abdominal pain or gastrointestinal bleeding due to vascular involvement of the mesenteric vessels.

Microscopic polyangiitis is one of the three vasculitides associated with antineutrophil cytoplasmic antibodies (ANCA), and 75 to 80% of patients have pANCA with a typical myeloperoxidase specificity (anti-MPO). Patients without poor prognostic factors, according to the five-factor score, can theoretically be treated with corticosteroids alone, with immunosuppressants added only in cases of treatment failure or relapse.

Patients with poor prognostic factor(s) should be treated upfront with a combination of corticosteroids and immunosuppressants, primarily intravenous cyclophosphamide in bolus doses, along with plasmapheresis in cases of severe renal failure. Once remission is achieved, a less toxic maintenance treatment with azathioprine or methotrexate will be prescribed as a follow-up. The maintenance treatment should last at least 12 months [8]. The role of new biotherapies is still to be defined, and studies are ongoing. Remission is achieved in over 80% of cases by following these therapeutic recommendations, and the relapse rate is around 30% at five years, which is lower than that observed in Wegener's granulomatosis.

4. Conclusion

Microscopic polyangiitis is a serious systemic disease that can manifest with varied and sometimes subtle symptoms. Its diagnosis, which can be complex for clinicians, relies on often nonspecific clinical signs, typical histological abnormalities, and the frequent presence of autoantibodies, particularly anti-MPO p-ANCA. This article presents a clinical case of a 62-year-old man with an atypical presentation of microscopic polyangiitis, confirmed by positive ANCA, evocative imaging results, and abnormalities found during bronchoalveolar lavage. Rapid identification of this disease is crucial, especially in atypical cases. Through a rigorous differential diagnostic approach that considers infections, neoplasms, and other immunological disorders, we established the diagnosis of microscopic polyangiitis. Accelerating the diagnostic process is essential to enable early and effective treatment, thereby minimizing the risks of disease progression.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

‘The present research work does not contain any studies performed on animals/humans subjects by any of the authors’.

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