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CASE REPORT

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Unilateral diffuse alveolar haemorrhage with microscopic polyangiitis: A case report

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Abstract

Diffuse alveolar haemorrhage (DAH) is a life-threatening condition caused by widespread damage to the small pulmonary vessels. Common chest imaging findings in patients with DAH show bilateral diffuse airspace opacities. DAH complicating antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, including microscopic polyangiitis, should be considered as a differential diagnosis in patients with progressive dyspnea, reduced haemoglobin levels, and alveolar opacities on chest imaging. We report the case of a 78-year-old woman who presented with unilateral DAH, severe dyspnea, and anaemia. DAH was confirmed using bronchoalveolar lavage. Laboratory test results, including ANCA, positive anti-myeloperoxidase antibody, and negative anti-proteinase 3, led to a diagnosis of microscopic polyangiitis. Rituximab and methylprednisolone were administered. The patient's symptoms, laboratory test results, and chest radiography findings improved after the initiation of treatment. This case highlights the importance of prompt recognition of clinical symptoms and signs, including dyspnea and anaemia, for the diagnosis of DAH.

KEYWORDS

bronchoalveolar lavage, microscopic polyangiitis, unilateral diffuse alveolar haemorrhage

INTRODUCTION

Diffuse alveolar haemorrhage (DAH) is a rare and potentially life-threatening pulmonary disease caused by a variety of diseases associated with hemoptysis, anaemia, bilateral diffuse lung infiltration, and acute respiratory failure.[1] The typical manifestations include hemoptysis accompanied by bilateral diffuse opacities in chest radiographs and decreased haemoglobin levels.[1] Considering the high mortality, prompt diagnosis and immediate treatment are required. However, its clinical presentation is non-specific, making early diagnosis difficult. Herein, we report the case of a woman who was diagnosed with DAH with microscopic polyangiitis (MPA), although she presented with unilateral diffuse opacities on chest radiography.

CASE REPORT

A 78-year-old woman presented to the emergency department of our institution with complaints of dyspnea for the past 2 days. The patient had a history of hypertension, diabetes mellitus, and hypothyroidism. The initial blood pressure, respiratory rate, and body temperature were 132/54 mmHg, 22 breaths/min, and 36.4°C, respectively. Her saturation was 82%, and she immediately received oxygen at 4 L/min. Chest radiography showed unilateral infiltration, and chest computed tomography (CT) revealed unilateral diffuse geographic consolidation in the entire right lung (Figure 1). Initial laboratory findings were as follows: low haemoglobin (7.1 g/dL), creatinine (1.69 mg/dL), and blood urea nitrogen (BUN, 38.7 mg/dL) levels. The urine dipstick test showed proteinuria (1+) with hematuria (3+).

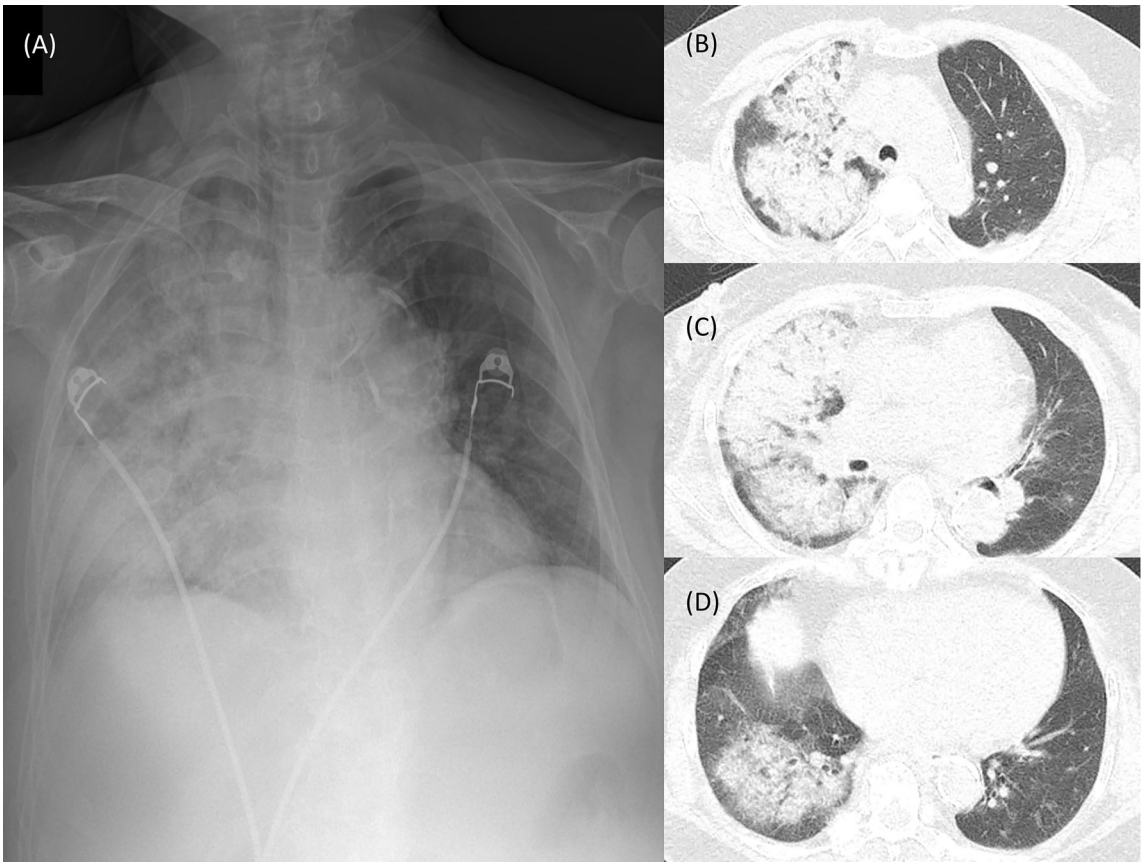

FIGURE 1 The consolidation of the geographic appearance on chest CT sparing the left hemithorax due to a complete unilateral right sided diffuse alveolar hemorraghe in microscopic polyangiitis. (A) chest X-ray. (B) chest CT (axial plane) in upper lung field. (C) chest CT (axial plane) in middle lung field. (C) chest CT (axial plane) in lower lung field

The radiography of paranasal sinuses excluded sinusitis. On the second day of admission, flexible bronchoscopy for bronchoalveolar lavage (BAL) was performed to evaluate pulmonary infiltration and the cause of anaemia. The BAL fluid analysis suggested DAH because serial aliquots showed progressively increased blood return (Figure 2), and bacterial, fungal, and viral infections were ruled out. Antineutrophil cytoplasmic antibody (ANCA) examination revealed positive anti-myeloperoxidase (anti-MPO) antibody (120 IU/mL) and negative anti-proteinase 3 (anti-PR3) antibody (0.3 IU/mL). Urinary dysmorphic erythrocytes were 10%.

Renal ultrasound and biopsy were recommended to confirm glomerulonephritis; however, considering the patient's general condition, treatment was initiated on the second day after admission with intravenous rituximab (500 mg) and methylprednisolone (125 mg). Four weekly doses of rituximab were administered. Methylprednisolone (125 mg) was injected every 12 h for 3 days and tapered (60, 50, 40, and 30 mg/day for week 1, 2, 3, and 4, respectively). Serial chest radiographs taken after the beginning of treatment showed a gradual decrease in infiltration of the right lung (Figure 3). The amount of oxygen supplied to the patient was also tapered on day 11 of admission. The patient was discharged on the 25th day of admission with oral prednisolone (25 mg) daily. The day before discharge, BUN and creatinine levels also showed improvement (BUN 18.5 mg/dL, creatinine 0.89 mg/dL).

DISCUSSION

The most common chest radiographic finding of DAH is diffuse bilateral alveolar opacification that sometimes has a central and lower lung distribution.[2] DAH is known to increase mortality in patients with MPA; especially in an acute clinical setting, 30% of patients do not survive an episode of DAH.[1] In this case, the patient presented with unilateral alveolar opacification on chest radiographs and anaemia without hemoptysis. Based on the clinical symptoms and signs, including dyspnea and anaemia, DAH was suspected.

MPA is an idiopathic autoimmune disease characterized by systemic vasculitis that predominantly affects the small size arteries, and is associated with the presence of ANCA.[3] Common clinical manifestations include rapidly progressive pauci-immune glomerulonephritis and alveolar haemorrhage.[3] Pulmonary involvement among MPA patients has

been reported to be 60%–70%, with pathologic capillaritis being the most common manifestation.[1] Approximately 29%–36% of MPA patients are known to show DAH.[1]

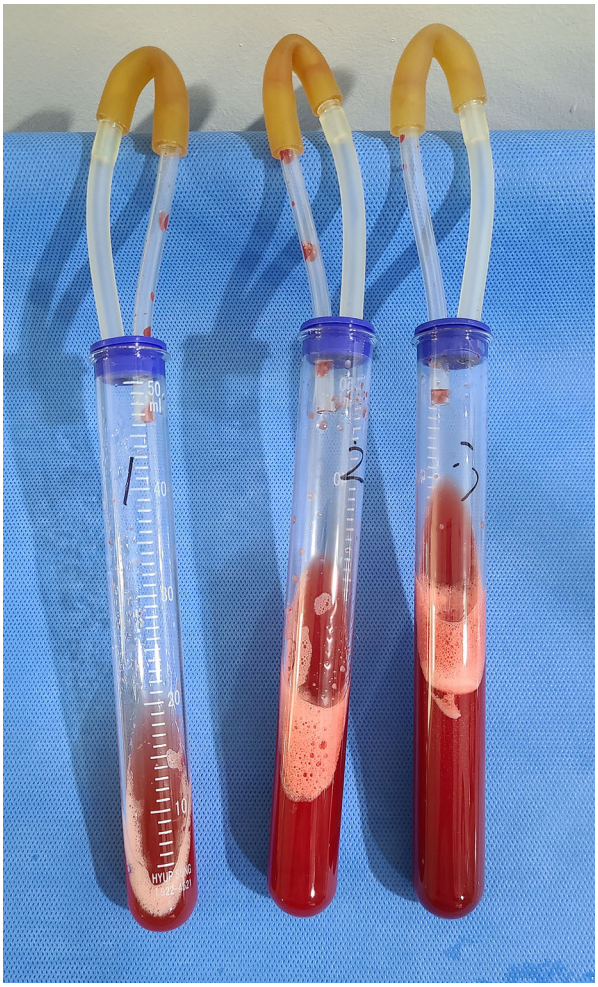

FIGURE 2 Bronchoalveolar lavage fluid suggesting diffuse alveolar hemorrage. Serial aliquots showing progressively increased blood return

Neutrophilic infiltration of lung interstitium leads to necrosis of alveolar structures, loss of capillary structural integrity, and spilling of red blood cells into the alveolar space and interstitium.[4] Therefore, although a focal or unilateral opacity distribution does not exclude haemorrhage, chest radiographs show patchy, bilateral airspace opacities, usually involving both lung fields.[2]

In a study of 112 patients with DAH of various causes, 13 patients (11.2%) exhibited unilateral infiltrates on chest radiographs. One of 39 patients with immune-related DAH had unilaterality on chest radiographs.[5] Another study of 39 patients with DAH with a proven immunological origin revealed that two patients (5.1%) showed unilateral infiltrates.[6] Although predominantly right-sided DAH was often caused by pulmonary congestion resulting in heart failure, the study of unilateral DAH is rare.[5] However, some cases on unilateral DAH due to granulomatosis with polyangiitis, negative pressure pulmonary edema, or a drug have been reported.[5] In our case, the patient was diagnosed with MPA; to the best of our knowledge, this is the first reported case of unilateral DAH as an initial presentation of MPA.

DAH is recognized by hemoptysis, anaemia, diffuse radiographic pulmonary infiltrates, and hypoxemic respiratory failure. Hemoptysis is initially absent in up to 33% of DAH cases.[1] In our case, the patient complained of dyspnea without any other symptoms. Initial decreased haemoglobin levels helped to promptly consider DAH. However, because of the life-threatening condition, we could not pathologically confirm glomerulonephritis or DAH, which is a limitation of our study. However, we were able to diagnose MPA based on the presence of anti-MPO antibody and sequential BAL aliquots.[1]

Tomos et al. reported the case of a man who was diagnosed with unilateral DAH in MPA and treated with methylprednisolone and cyclophosphamide.[7] In life-threatening cases, both rituximab and cyclophosphamide, in combination with glucocorticoids, have been used for inducing

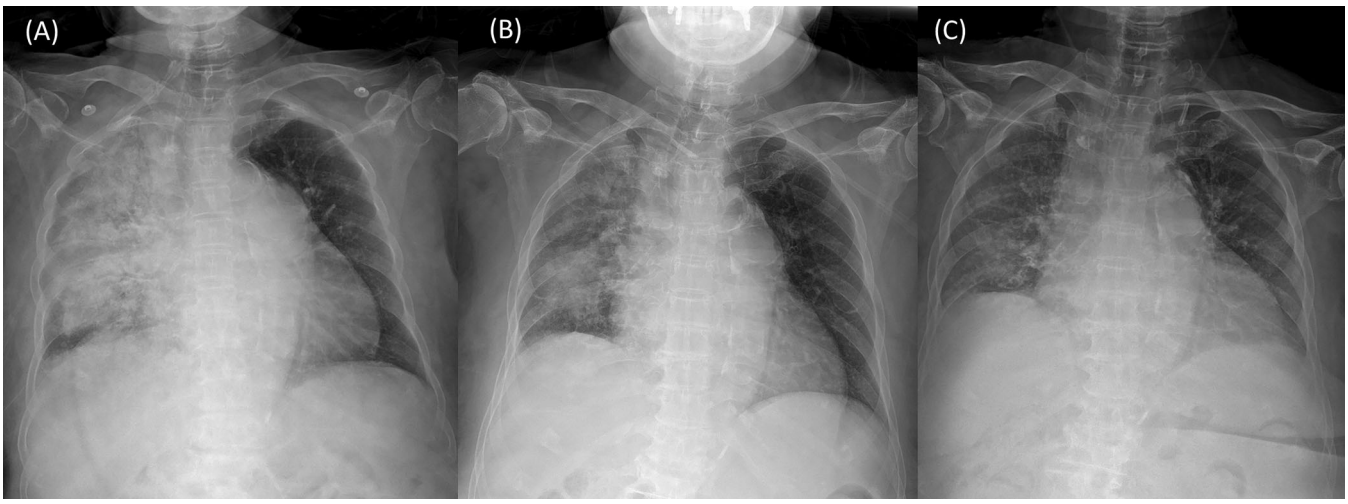

FIGURE 3 The serial chest radiographs taken after initiation of treatment. (A) Day zero of intravenous rituximab and corticosteroid administration. (B) Day 7 after intravenous rituximab and corticosteroid administration. (C) Day 21 after intravenous rituximab and corticosteroid administration

remission of GPA and MPA.[8] Because rituximab is considered less toxic than cyclophosphamide, the guidelines of the American College of Rheumatology recommend treatment with rituximab in combination with glucocorticoids.[8] In our patient's case, treatment with rituximab and methylprednisolone showed improvement in both the symptoms and lesions on radiographs.

Unilateral DAH can occur in MPA and may not always include hemoptysis. Therefore, patients with decreased haemoglobin levels without hemoptysis could still have DAH, and the possibility of MPA in patients with unilateral pulmonary infiltration should not be neglected.

AUTHOR CONTRIBUTIONS

Tae Gyoung Kim and Hyung Koo Kang was responsible for the conceptualization and drafting of the manuscript. Jiyeon Kang, Woo Jung Seo, Jieun Kang, So Hee Park, Hyeon-Kyoung Koo, Hye Kyeong Park, Sung-Soon Lee, Jung Gon Kim drafted the manuscript and approved the final manuscript.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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