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Research Article

A rare case report of massive alveolar hemorrhage, a rare and potentially fatal consequence of wegener's granulomatosis

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ABSTRACT
A rare, often fatal, and still curable consequence of Wegener's granulomatosis (WG), massive diffuse alveolar haemorrhage (DAH) is linked to death in over 66% of cases. DAH is a syndrome that can appear as a symptom of many different conditions. Since the best chance of survival for patients with WG, a multisystem necrotizing granulomatous vasculitis, is an early diagnosis and vigorous care with cytotoxic medications, it should always be taken into consideration as a potential etiological component in these individuals, whether or not other systems are involved. We report a rare case of 58-year-old man who presented with massive haemoptysis and epistaxis in emergency department and subsequently diagnosed as WG.

KEYWORDS: Diffuse alveolar haemorrhage (DAH); Wegener's granulomatosis (WG); computed tomography (CT); antineutrophil cytoplasmic antibodies (ANCA).

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INTRODUCTION

Wegener's granulomatosis (WG) consists of necrotizing granulomatous disorder in the nose and lung, necrotizing granulomatous vasculitis in generalized arterioles, and intractable vasculitis that features renal necrotic crescent forming nephritis. It is the most common of the antineutrophil cytoplasmic antibody – associated vasculitis which also includes Churg–Strauss syndrome and microscopic polyangiitis.[1]

Diffuse alveolar haemorrhage is a serious manifestation of granulomatosis with polyangitis (Wegener's) with high morbidity and mortality. The clinical triad of hemoptysis, anemia, and increasing hypoxemia characterizes it. In a suitable clinical context, bronchoalveolar lavage, lung biopsy, or the identification of C-antineutrophil cytoplasmic antibodies are used to confirm the diagnosis of granulomatosis with polyangitis.[2]

The diagnosis is difficult since the occurrence is abrupt and both symptoms and histology of the lesion are nonuniform and nonspecific. Since the early diagnosis is essential for the outcome, it should be considered in every patient of WG with severe pulmonary symptoms. Cyclophosphamide and corticosteroids are the recommended form of therapy for this disorder.

CASE REPORT

A 58-year-old male was admitted to our department with complaints of shortness of breath, nonproductive cough, epistaxis , hemoptysis, and low-grade fever since 10 days and pedal edema for last 4 days. Dyspnea was slowly progressive with three episodes of hemoptysis. patient .There was past history of CVA 2 years back and was on regular medications for the same. No history of allergic diathesis or bronchial

asthma. He was a farmer by occupation, tobacco chewer ,non-smoker, and occasional alcoholic.

On general examination, he was pale. His resting pulse rate was 108/min, blood pressure 100/60 mmHg, and respiratory rate 32/min. Chest examination was unremarkable on inspection, palpation, and percussion. On auscultation, bilateral diffuse course crepitations were present. Routine investigations showed were as follows: Hb – 6.4 g%, TLC – 6000/mm^3, DLC – neutrophils – 85%, lymphocytes – 10%, monocytes – 3%, platelet count – 3,27,000, erythrocyte sedimentation rate – 120, serum bilirubin – 0.8 mg/dl, SGPT – 40 IU and SGOT – 71 IU, blood urea – 144 mg/dl, and serum creatinine – 6.5 mg/dl. Arterial blood gas analysis done at the time of admission revealed hypoxia with PaO_2 44 mmHg and $PaCO_2$ 32 mmHg. Chest x-ray showed bilateral alveolar infiltrates [Figure 1]. High-resolution computed tomogram of chest suggestive of extensive consolidation with ground glass opacities with bilateral large areas of pulmonary alveolitis. [Figure 2].

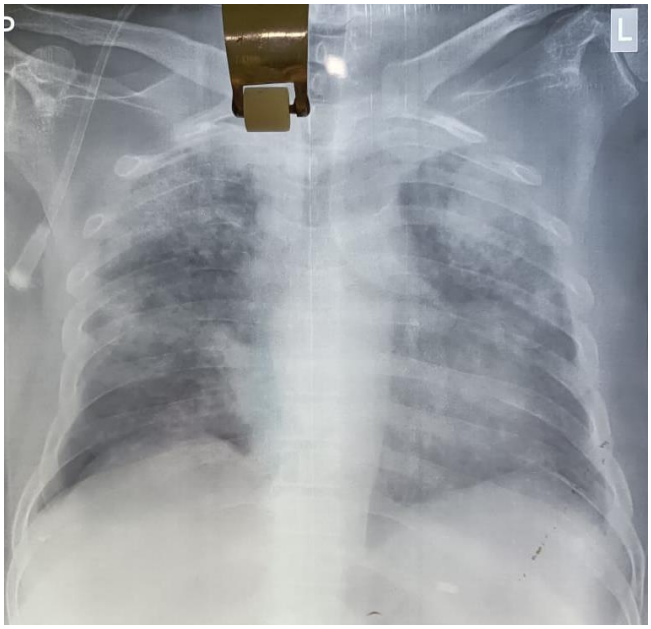

Figure 1: Bilateral alveolar infiltrates

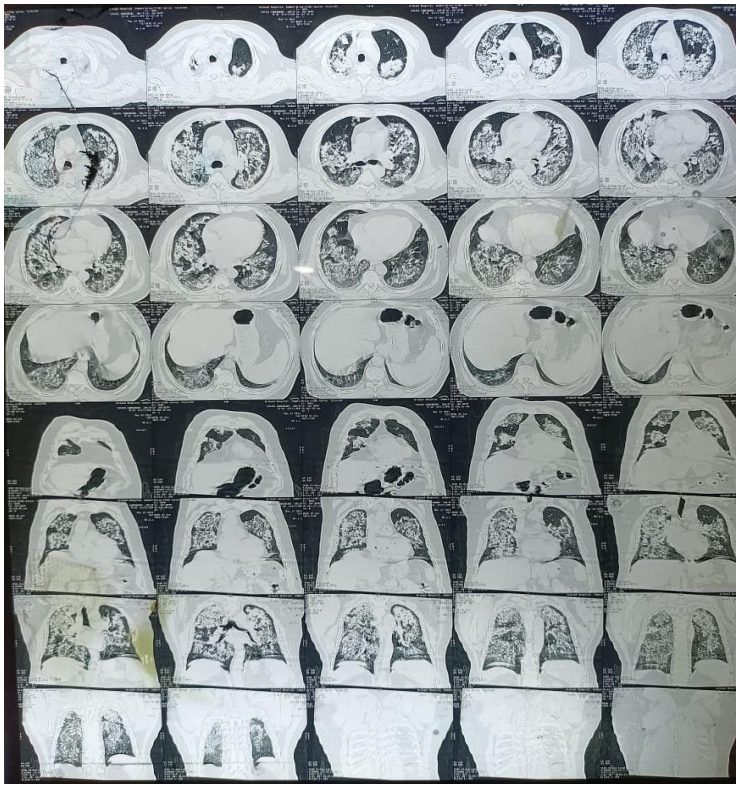

Figure 2 : Extensive consolidation with ground glass opacities with bilateral large areas of pulmonary alveolitis.

The triad of anemia, hemoptysis, and widespread air-space consolidation led us to think a possibility of systemic illness, prompting further investigation to determine the underlying cause. Urinalysis showed the presence of microscopic hematuria. Three consecutive sputum samples tested negative for acid-fast bacilli. Bleeding time and clotting time were within normal limits. To rule out any systemic vasculitis, the antinuclear antibody (ANA) profile and antineutrophil cytoplasmic antibody (ANCA) were sent. The ANA profile was negative but P-ANCA came out to be strongly positive. So the diagnosis of diffuse alveolar hemorrhage was made and WG was thought to be the cause. The patient was given cyclophosphamide pulse (1 g) along with pulse methylprednisolone (750 mg) daily. Four units of blood were transfused to correct anemia. Noninvasive ventilation was used to maintain oxygenation. After about 48 h of strict monitoring, he showed significant improvement clinically.. Radiograph taken after 6 days of treatment showed significant clearing [Figure 3]. The patient is clinically asymptomatic after five cycles of cyclophosphamide pulse therapy.

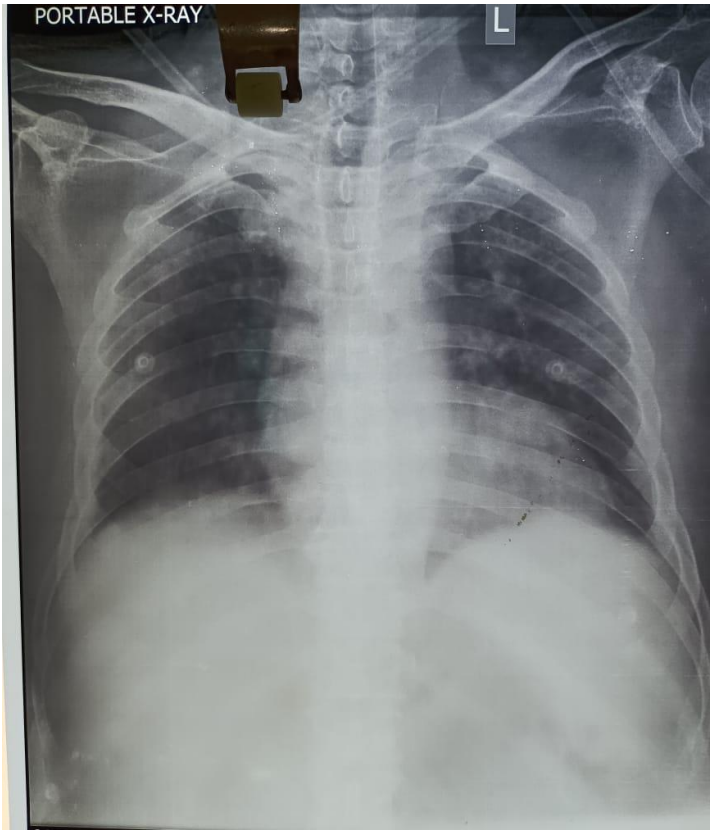

Figure 3: Follow-up chest radiograph showing significant radiological improvement.

DISCUSSION

Diffuse alveolar haemorrhage (DAH) refers to widespread bleeding within the alveoli due to significant damage to the alveolocapillary membrane of the lungs. DAH can indicate systemic disorders, as well as result from lung-specific injury. The majority of DAH cases are linked to capillaritis associated with systemic autoimmune disorders such as antineutrophil cytoplasmic antibody-associated vasculitis, antiglomerular basement membrane disease, and systemic lupus erythematosus.3. In rare instances, the pulmonary metastasis of angiosarcoma has also been noted as a cause of DAH.[4]

WG is a complex disease that presents with necrotizing granulomatous inflammation in the upper and lower respiratory tracts, as well as glomerulonephritis. It is a rare disease, with an estimated prevalence of 3 per 100,000, and the average age of onset is 40 years.5.The lung is frequently impacted in WG, with signs of involvement detected in more than 90% of patients throughout the disease progression. In 9% of cases, the lung is the sole organ affected.6. It can present as asymptomatic infiltrates or manifest with symptoms such as cough, hemoptysis, difficulty breathing, and chest discomfort. Even though the lung is frequently impacted in Wegener's Granulomatosis, the occurrence of diffuse alveolar hemorrhage due to capillaritis is rare, with an estimated frequency ranging between 7% to 45%.7.

Immunosuppressive agents represent the cornerstone of treatment for diffuse alveolar haemorrhage, particularly when linked to systemic or pulmonary vasculitis. In individuals experiencing life-threatening organ damage, the initial treatment involves employing high doses of cyclophosphamide and prednisolone to induce remission.[8] In addition to corticosteroids, other immunosuppressive medications like cyclophosphamide, azathioprine, mycophenolate mofetil, methotrexate, and etanercept can be utilized in cases of diffuse alveolar haemorrhage. This is especially true in severe cases, when standard corticosteroid treatment has proven ineffective, or if there is a specific underlying cause.[9] Furthermore, azathioprine is recommended for long-term maintenance

therapy. A recent multicenter study proposed that substituting azathioprine for 3-month cyclophosphamide to sustain remission is just as efficient as opting for an extended cyclophosphamide treatment.[10] Relapses are frequent even with ongoing immunosuppressive therapy, as half of the patients experience a relapse within 5 years. In addition to these developments, ongoing trials are investigating the potential of newer medications such as rituximab, deoxyspergualin, antithymocyte globulin, and interferons in the treatment of WG.[11]

CONCLUSION

Alveolar hemorrhage was linked to a mortality rate of over 50%, which partially reflects the seriousness of this Wegener's granulomatosis complication. Cytotoxic agents were used to treat all survivors; those who passed away received either no specific therapy, corticosteroid-only treatment, or passed away before cytotoxic therapy could be expected to be effective. Wegener's granulomatosis should be in the differential diagnosis for patients who present with alveolar hemorrhage with or without renal failure. An aggressive diagnostic approach and the earliest possible administration of cytotoxic drugs in combination with corticosteroids offers the best chance of survival in this fulminant condition.

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