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Overlap Syndrome: Systemic Lupus Erythematosus and ANCA Vasculitis in a Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: Microscopic polyangiitis is a systemic necrotising vasculitis that predominantly affects small-calibre vessels. It may be associated with systemic lupus erythematosus (SLE) as part of an overlap syndrome, but there are very few reports of this in the literature.
Case Report: We report the case of a 19-year-old patient admitted with moderate haemoptysis, epistaxis and petechial purpura revealing intra-alveolar haemorrhage with a strongly positive P-ANCA. In addition, blood tests for antinuclear antibodies and rheumatoid factor were positive, with renal involvement suggesting associated lupus as part of an overlap syndrome. Progression was good with corticoide and immunosuppressant treatment.
Conclusion: The clinical presentation of overlap syndrome is rare and severe, with a dual immunological profile of antinuclear antibodies and neutrophil cytoplasm.

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Keywords: Overlap; treatment; Lupus; corticoids; haemorrhage.

ABREVIATIONS

SLE : Systemic Lupus Erythematosus
ANCA : Neutrophil Cytoplasmic Antibodies
ENT : Ear Nose Throat
AV : ANCA vasculitides
ANA : Antinuclear Antibodies

1. INTRODUCTION

Microscopic polyangiitis is a systemic necrotizing vasculitis that predominantly affects small-caliber vessels. The most typical clinical manifestations are extracapillary glomerulonephritis and alveolar hemorrhage. This is a vasculitis associated with neutrophil cytoplasmic antibodies (ANCA). The p-ANCA type, with anti-myeloperoxidase specificity, is found in 75-80% of patients [1]. ANCA is negative, but may be associated with systemic lupus erythematosus (SLE) as part of an overlap syndrome. Very few reports of this condition have been published in the literature.

2. CASE REPORT

We report the case of a 19-year-old female patient with no previous pathological history who was admitted for moderate hemoptysis, associated with epistaxis and petechial purpura, with generalized mucocutaneous pallor, all of which had been evolving for 3 months in a context of declining general condition. The patient was admitted to hospital on an emergency basis, with conditioning and injections of haemostatic treatment.

Chest imaging revealed an interstitial syndrome with bilateral ground-glass, predominantly at the bases (Fig. 1).

Blood count was consistent with poorly tolerated iron-deficiency hypochromic microcytic anemia, with a hemoglobin level of 4.8g/dL, for which the patient received 3 packed red blood cells. Sedimentation rate was accelerated and CRP was elevated to 54mg/l. An emergency bronchioloalveolar lavage confirmed intra-alveolar haemorrhage on macroscopic examination (Fig. 2), with a Golde score of over 20. In the search for ENT involvement, nasofibroscopy showed inflammation with nasal ulceration of the inferior turbinate and bulging of the upper wall of the cavum, and a biopsy was unremarkable.

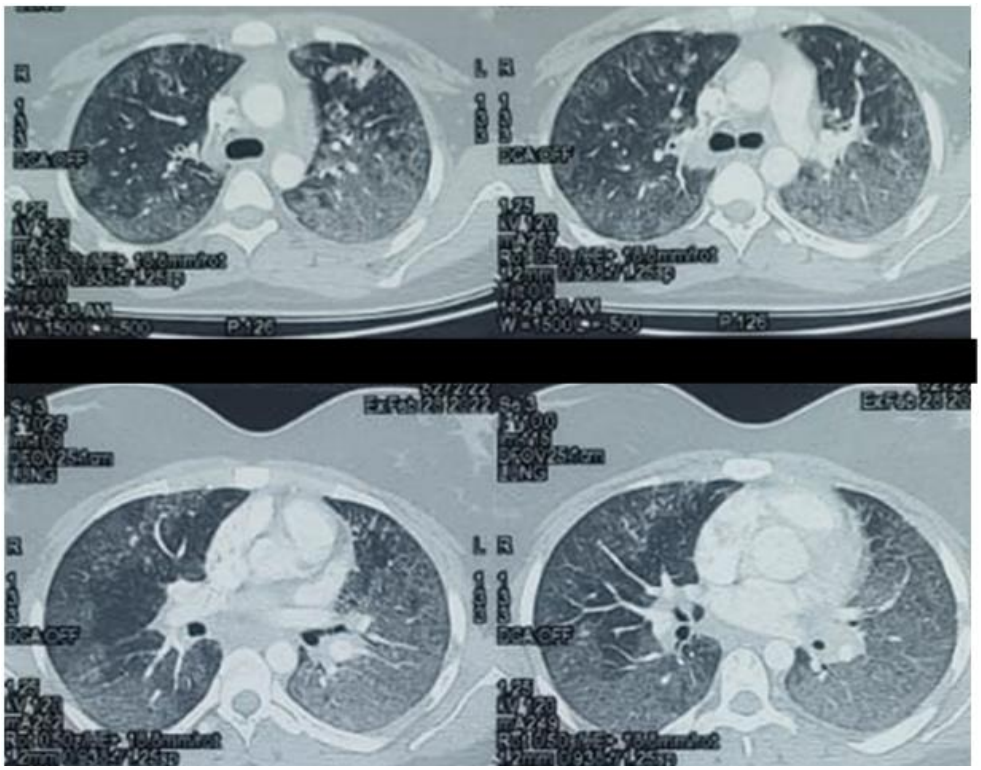

Fig. 1. Chest CT scan showing diffuse ground-glass appearance

Fig. 2. Macroscopic appearance of frankly haemorrhagic bronchoalveolar lavage

The Blondeau scan revealed a deviated nasal septum with no evidence of acute or chronic sinusitis (Fig. 3). The renal work-up was in favor of a pure nephrotic syndrome, with 24h proteinuria at 3.5g/24h and albuminemia at 25g/L. Renal ultrasound was unremarkable, so a renal biopsy was indicated. The blood immunoassay showed strongly positive P-ANCA. The diagnosis of microscopic polyangiitis was thus accepted. The biopsy showed stage 3 extramembranous glomerulonephritis with vascular deposition of IgG, IgM and C3. In addition, blood tests for antinuclear antibodies and rheumatoid factor were positive at 320 and 16.06 IU/ml respectively. Thus, the patient also met the diagnostic criteria for SLE. The patient was started on bolus corticosteroids, followed by oral corticosteroids at a dose of 1mg/kg/d with progressive tapering, then bolus cyclophosphamide at 3-week intervals, followed by azathioprine.

Hemoptysis subsided with radiological clearance (Fig. 4).

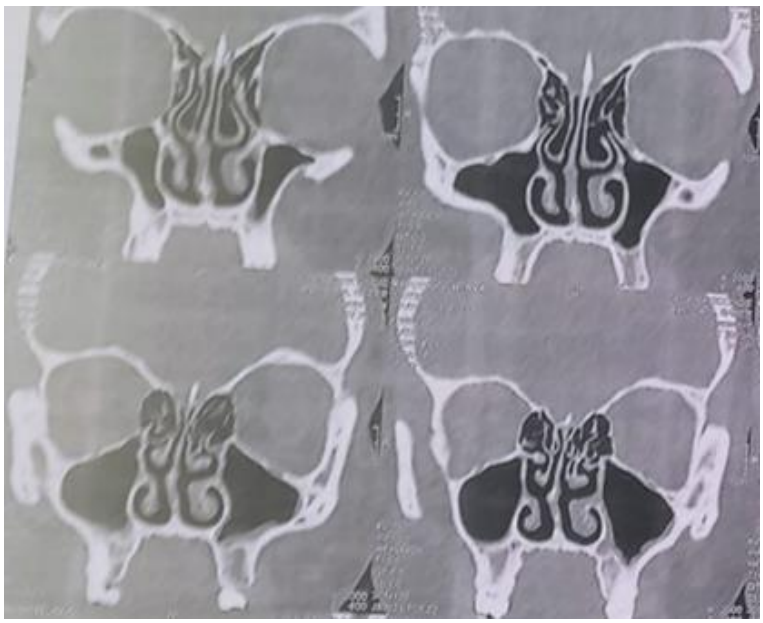

Fig. 3. Blondeau scan in favour of a deviated nasal septum

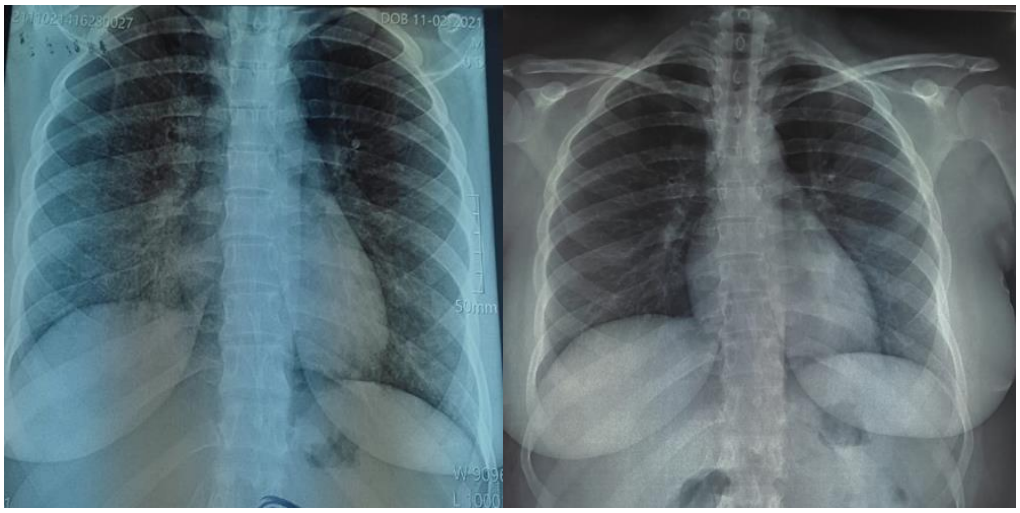

Fig. 4. Radiological clean-up after 3 months of treatment

3. DISCUSSION

Overlap syndromes are defined by the presence of criteria for several systemic diseases (at least two) at the same time in the same patient [1]. Unlike systemic lupus erythematosus (SLE), ANCA vasculitides (AV) have rarely been associated with other autoimmune diseases [2]. The SLE /AV overlap syndrome is a rare condition defined by the existence of a systemic disease meeting both SLE and AV classification criteria. The diagnosis of SLE may precede, follow or be concomitant with that of AV, as was the case in our patient. Females were frequently involved.

SLE and AV are two autoimmune diseases that differ in their autoantibody profile (anti-nuclear antibodies ANA in SLE and anti-neutrophil cytoplasm antibodies in AV), resulting in different systemic involvement. Renal involvement is common in both diseases, It is about glomerulonephritis type with immune complex deposits in SLE, and pauci-immune extracapillary glomerulonephritis type without immune complex deposits in AV [3,4].

Although rare, the SLE / AV overlap syndrome could be explained by immunopathological mechanisms common to both SLE and AV, in particular neutrophil activation and the involvement of ketosis, a cellular immune process that affects neutrophil polynuclear cells, releasing fibers composed of DNA and proteins called NETs, whose function is to trap pathogenic microorganisms. Significant netosis and impaired clearance of NETs are involved in the pathophysiology of tissue damage observed in various autoimmune diseases and small-vessel vasculitides [5]. The occurrence of SLE and AV has been reported under anti-TNF therapy [6].

In patients with lupus nephropathy, overlapping histological lesions with necrotizing extracapillary glomerulonephritis, as defined for vasculitis, make the diagnosis of an overlap syndrome. This may be associated with granulomatous inflammation of the entire respiratory tract in the SLE/Wegener association, eosinophil-rich granulomatous inflammation of the respiratory tract associated with asthma and hypereosinophilia in the SLE/granulomatosis eosinophilia with polyangiitis association, pneumo-renal syndrome without granulomatous inflammation in the SLE/microscopic polyangiitis association, and with or without ANCA positivity by IFI and/or ELISA. In the case of extra-capillary necrotizing glomerulonephritis of a AV diagnosed 1st, overlapping histological lesions with the presence of unexpected deposits of immune complexes (C3, IgG, C1Q) or validation of at least four diagnostic criteria for SLE, including ANCA, and/or anti-DNA-native positivity will make the diagnosis of an overlap syndrome [7,8].

An overlap syndrome is excluded in patients with lupus nephropathy associated with the presence of ANCA only, without clinical or histological arguments for ANCA vasculitis, or in patients with necrotizing extracapillary glomerulonephritis associated with the presence of antinuclear antibodies (ANA) or native anti-DNA but without clinical or histological arguments for SLE or positivity of antibodies directed against the

glomerular basement membrane [3]. It should be noted that genuine vasculitis can occur in the course of SLE, but rarely meets the criteria for classification as AV [9].

Currently, there are no treatment guidelines, and management decisions are often left to clinicians, depending on the predominant features of ANCA disease or lupus nephritis at the time of presentation [10]. Systemic corticosteroid therapy in the form of a bolus of methyl-prednisolone, 1 g/day for 3 days, followed by prednisone at a dose of 1mg/kg/day, combined with a synthetic antimalarial (hydroxychloroquine) has been described in the literature [11]. Accelerated biopsy and initiation of treatment are crucial to preserve renal function [12] Plasma exchange in patients with severe ANCA-associated vasculitis did not reduce the incidence of death or end-stage renal disease. However, it has been shown to be beneficial in patients with diffuse alveolar hemorrhage, compared with patients without diffuse alveolar hemorrhage [13].

4. CONCLUSION

The association of SLE and AV with renal involvement remains rare. The clinical presentation of SLE/AV overlap syndrome is severe, with a dual immunological profile of ANA and neutrophil anti-cytoplasm. In our patient, the evolution was favorable under treatment, but strict monitoring remains important to detect a potentially serious relapse.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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