USE OF INTRAVENOUS IMMUNOGLOBULINS IN DIFFUSE ALVEOLAR HEMORRHAGE SECONDARY TO SYSTEMIC LUPUS ERITHEMATOSUS: CASE REPORT AND REVIEW OF THE LITERATURE

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INTRODUCTION

Diffuse alveolar hemorrhage (DAH) in patients with Systemic Lupus Erythematosus (SLE) is a rare but potentially catastrophic pulmonary manifestation. Classical treatment consists of intravenous pulses of methylprednisolone followed by oral corticosteroids and intravenous cyclophosphamide. In refractory cases, plasmapheresis or intravenous immunoglobulins (IVIG) can be used. A case of a patient with DAH secondary to SLE who required IVIG treatment is presented.

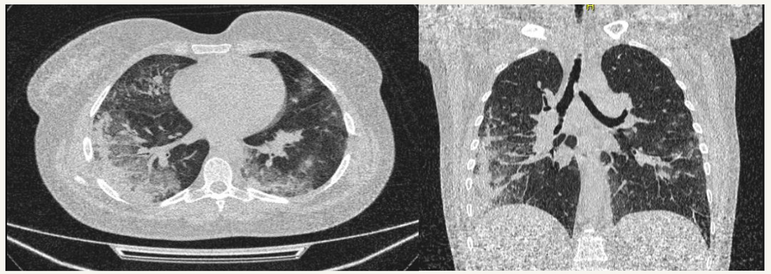
CASE DESCRIPTION

A 38-year-old female patient was admitted to the hospital because of fever, dyspnea and hemoptoic expectoration of recent onset. She also referred photosensitivity and arthralgias of small joints. Physical examination revealed fever, tachycardia, tachypnea, malar erythema, alopecia and hypoventilation.

Lab tests

- Hemoglobin: 9,1 gr/dl
- Hematocrit: 28%
- Erythrocyte sedimentation rate: 138 mm/h
- Serum creatinine: 1.44 mg/dl
- Proteinuria: 1.52 gr/24 hs
- Urine sediment: abundant red blood cells, leukocytes and proteins ++
- Positive ANA (titer 1/640, speckled pattern and titer 1/1280, homogenous pattern)
- Positive anti SSA/Ro
- Positive anti DNA (titer 1/160)
- C3 45 mg/dl, C4 8 mg/dl
- Negative ANCA, MPO and PR3

Chest Computed Tomography on presentation



Ground glass density and consolidation in both lungs suggestive of DAH and mild bilateral pleural effusion.

Bronchoalveolar lavage

Presence of abundant red blood cells and hemosiderin-laden alveolar macrophages.

Cultures: negative for bacteria, acid–fast bacilli and fungal infections.

CLINICAL COURSE

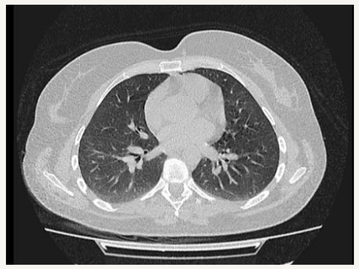
Three pulses of intravenous methylprednisolone on consecutive days were indicated, followed by oral prednisone (1 mg / kg / day).

→ On day 4 of hospitalization, she developed hemoptysis and respiratory failure associated with an acute drop in hematocrit (hematocrit 20%).

→ She required mechanical ventilation and 3 more pulses of intravenous methylprednisolone were indicated.

→ Plasmapheresis was initiated as rescue therapy, receiving 5 sessions in total, with no response to the treatment.

Due to the fact that the patient did not present a response to the treatment and also developed pneumonia associated with mechanical ventilation, IVIG therapy was initiated (2 g/kg/course divided into 5 days). This led to a sustained resolution of pulmonary hemorrhage and respiratory failure. Weaning from mechanical ventilation was achieved on day 5 of the first IVIG dose. Finally, cyclophosphamide was started.



Chest Computed Tomography after treatment with IGIV

DISCUSSION

DAH is a serious manifestation in SLE, with a mortality ranging from 28% to 64%. Its prevalence is variable: between 1.2 and 5.3% according to some series.

Although there are no well-established guidelines for treatment due to the lack of large clinical trials, DAH is generally treated with high-dose glucocorticoids and intravenous cyclophosphamide. In this case, it was required the use of plasmapheresis and IVIG therapy as rescue measures. IVIG are mainly polyvalent IgG and have immunomodulatory effect. Even though the evidence supporting their use is very low, they must be considered in patients with severe and refractory manifestations of SLE (DAH, thrombocytopenia, peripheral neuropathy, intestinal pseudo-obstruction and toxic epidermal necrolysis) or in those cases associated with infection or pregnancy.

CONCLUSIONS

DAH can be the initial manifestation of SLE. It is very important to make an early diagnosis and treatment should be started as soon as possible. IVIG therapy should be considered in severe and refractory cases.

Bibliography

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