RECURRING AND REFRACTORY DIFFUSE ALVEOLAR HEMORRHAGE AS THE FIRST MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS – CASE REPORT

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BACKGROUND

Diffuse alveolar hemorrhage (DHA) comprises an uncommon and potentially fatal manifestation in systemic lupus erythematosus (SLE). It occurs in less than 2% of patients and is associated with high mortality (up to 90%), and its treatment is based on case reports and expert opinion.

CASE REPORT

Female, 20 years old, history of mild cognitive disorder and previously healthy, subacute progressive condition of constitutional symptoms, alopecia, significant weight loss, and migratory arthritis. Acute worsening with fever, dry cough, dyspnea, pleuritic chest pain, and hemoptysis. On admission, she presented pallor, dyspnea, and desaturation, associated with severe anemia and lymphopenia, negative hemolysis tests, without other dysfunctions. Progressed with a drop in hemoglobin, performed tomography of the chest with a pattern suggestive of DHA (Figure 1), pleural and pericardial effusion, later confirmed in an echocardiogram. Clinical

Figure 1. Diffuse alveolar hemorrhage.

support started, with transfer to the ICU. Investigation showed positive ANA, anti-DNA, and SM in high titers, and consumption of complements, being diagnosed with severely active SLE. Hydroxychloroquine and pulse therapy with methylprednisolone 1 g/day for 3 days was started, followed by cyclophosphamide 0.6 g/m², with excellent clinical response, oxygen weaning, and recovery of hematimetric indices, with discharge. Despite the good initial evolution, it evolved sequentially with three episodes of recurrence of the episodes of DHA within 2 months. The first recurrence after 2 weeks, period of cyclophosphamide nadir, with increased corticosteroid therapy and association of rituximab 1 g D1, with new recovery and discharge. Before D15 of rituximab, a new episode of DHA, this time in association with nephritis and being opted for induction with pulse therapy with methylprednisolone and cyclophosphamide 1 g/m², and new clinical improvement. Bronchoscopy was performed, infectious and neoplastic differential diagnoses were excluded, with a pattern of chronic bronchitis. Soon after, she presented the third recurrence, with signs of infectious complication, hospitalization, antibiotic therapy, and immunoglobulin 2 g/kg in 5 days. After the last complication, he evolved with sustained clinical improvement, maintained on a monthly regimen of cyclophosphamide, having completed eight cycles and subsequent maintenance with azathioprine and weaning from corticosteroid therapy, with no further recurrences.

CONCLUSION

Diffuse alveolar hemorrhage is a rare manifestation in SLE, severe and associated with a poor prognosis, dependent on early diagnosis and treatment to minimize damage and risk of death. We present a case of great complexity and refractoriness, with different therapeutic lines, but with a favorable outcome.

KEYWORDS

Diffuse alveolar hemorrhage, Systemic lupus erythematosus, First manifestation.