



OFF-LABEL USE OF INTRAVENOUS CYCLOPHOSPHAMIDE IN SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING AS ACUTE LUPUS PNEUMONITIS: A CASE REPORT

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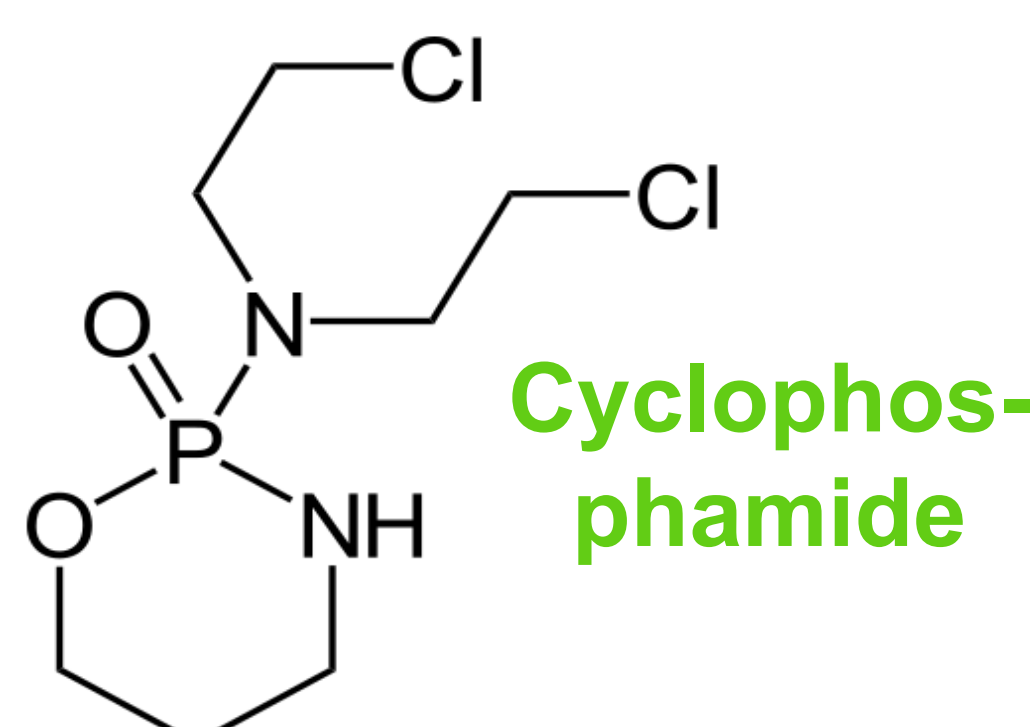
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BACKGROUND AND IMPORTANCE

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with wide-ranging pleuropulmonary manifestations. **Acute lupus pneumonitis (ALP)** is one of its uncommon complication. **Systemic steroids associated with immunosuppressive therapy (cyclophosphamide, rituximab, hydroxychloroquine and intravenous immunoglobulin)** are until the mainstream treatment of ALP.



AIM AND OBJETIVES

To describe the case of a **patient with ALP treated with intravenous cyclophosphamide** as well as to evaluate the **effectiveness and safety** of this treatment.

MATERIALS AND METHODS

1

We report a 67-year-old woman who went to the hospital due to fever, fatigue, myalgia, arthralgia, dyspnoea and dry cough with sputum for the past three weeks. Multiple and bilateral lung opacities were present on chest X-ray so she was diagnosed with community-acquired pneumonia.

2

The woman presented slight improvement despite empirical antibiotic and antifungal coverage. Subsequently, laboratory findings showed **leukopenia and positive anti-double-stranded-DNA antibodies** so the final diagnosis was **ALP secondary to SLE**.

3

Systemic steroid treatment was initiated with **high-dose of methylprednisolone and hydroxychloroquine**. **Due to the severity of the pulmonary involvement, it was requested to start treatment with intravenous cyclophosphamide**.

RESULTS

The patient received a total of three doses (600mg/m²) of intravenous cyclophosphamide. MESNA, ondansetron and oral hydration were prescribed as supportive treatment. Despite the decrease in inflammatory analytical parameters, the woman presented **modest reduction of lung injury and symptoms**. She reported high-grade myalgia and vomiting after first infusion, which was successfully treated with paracetamol and metoclopramide. **Sequential therapy with oral cyclophosphamide was considered, but because it is not funded for ALP and its adverse effect profile, treatment with methotrexate was started.** Currently, the patient continues treatment with methotrexate, hydroxychloroquine and oral steroids. Computed tomography, performed three months after ending intravenous cyclophosphamide, showed **stability of the disease**.



CONCLUSION AND RELEVANCE

Treatment with intravenous cyclophosphamide has not shown promising results in our patient although its safety profile is good. Because the therapeutic alternatives in patients with ALP are limited, **it would have been interesting to verify that sequential therapy with oral cyclophosphamide** improves the signs and symptoms of the disease, and long-term adverse effects could also be analyzed.