TREATMENT OF ROSAI DORFMAN´S HSTIOCYTOSIS: CASE REPORT

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BACKGROUND AND IMPORTANCE

Rosai Dorfman disease (DRD) is a rare non-Langerhans histiocytosis. This is →

NO ESTABLISHED TREATMENT

FEW THERAPEUTIC OPTIONS

TREATMENT WITH LIMITED EVIDENCE

DRD has recently been related to the identification of mutations in the mitogenic activation protein kinase (MAPK)-dependent signaling pathway, being an interesting target for its treatment.

AIM AND OBJECTIVES

This report will discuss the case of a patient with DRD who **responded adequately** to targeted therapy with **TRAMETINIB**.

MATERIALS AND METHODS

A bibliographic review of cases described with similar symptoms was carried out. The Pharmacy Service collaborated in the search for a possible effective treatment and justified the need to start treatment with a MEK inhibitor.

RESULTS

45-year-old patient being followed up for gastrointestinal episodes and lymphadenopathy who was diagnosed with histiocytosis compatible with DRD in 2021. It was started treatment with **corticosteroids**, which in the event of refractoriness was changed to **peginterferon alfa** without response. At the beginning of March 2023, he was admitted to the ward due to deep vein thrombosis and pulmonary thromboembolism. He received a new treatment regimen with **anakinra** for 13 days without success.

Corticosteroids 1 mg/kg → Peginterferon alfa 90 mcg → Anakinra 100 mg/0,67 ml → Trametinib 1mg/day

The case is consulted and it is decided to change to a MEK inhibitor. Its use is requested outside of indication despite not obtaining any alteration in the MAPK pathway. **Trametinib** was started at a dose of 1 mg/day. After 3 months, she currently has good tolerance with platelet counts of 37,000 and decreased lymphadenopathy.

As toxicity to trametinib, acneiform rash have been reported.

CONCLUSION AND RELEVANCE

- No well-defined protocol for the treatment of DRD and therefore they represent a diagnostic and therapeutic challenge.
- This case contributes to the limited data published on targeted therapy with MEK inhibitors in DRD when cases are refractory to traditional therapies.

BIBLIOGRAPHY

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Abstract number: 4CPS-049

ATC-code: L01- ANTINEOPLASTIC AGENTS