HYPEREOSINOPHILIC SYNDROME AND WELLS SYNDROME (EOSINOPHILIC CELLULITIS): A CASE REPORT IN THE BRAZILIAN AMAZON

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Introdução

Hypereosinophilic syndromes (HES) constitute a group of rare diseases characterized by peripheral blood eosinophilia of 1.5 × 10 9/L or greater and evidence of end-organ manifestations attributable to eosinophilia and not otherwise explained in the clinical setting. In the USA, a prevalence of 5,000 cases per year is estimated (OLDÁN et al., 2009). HES are pleomorphic in clinical presentation and can be idiopathic or associated with a variety of underlying conditions, including allergic, rheumatologic, infectious, and neoplastic diseases. Epidemiologically, in HES, males are more affected than females (ratio of 9:1), predominantly between 20 and 50 years of age, although it can occur in children. The main objective of HES therapy is to control the number of eosinophils to prevent the progression of organ damage. Treatments include corticosteroid therapy, targeted therapy with a tyrosine kinase inhibitor (imatinib mesylate) for patients with FIP1L1-PDGFRA mutation and other chemotherapeutic agents (hydroxyurea, vincristine, etoposide, cyclosporine A and interferon-a as monotherapy or in combination with hydroxyurea, particularly in the myeloproliferative variant).

Casuística e Métodos

This is a descriptive study of the Case Study type, which will be made possible through research in the medical records of a specific patient and literature review, without causing pain or discomfort to the participant, without involving the collection of biological material, and without for profit (GIL, 2012). The information contained in this work will be obtained through analysis of medical records, registration of diagnostic methods to which the patient was submitted, and literature review.

Resultados

Initial laboratory tests, dated 05/10/2019, showed leukocytosis (leucometry of 15,900/mm³) with absolute and relative eosinophilia (30% eosinophils: 4,770/mm³), increased IgE (3,826 KU/L), high dose of vitamin B12 (1,145pg/ml). She performed a myelogram on 06/21/2019 which showed granulocytic series with relative and absolute hypercellularity with 22% eosinophils. Immunophenotyping with a moderately positive expression pattern for the hematopoietic marker CD45 and positive for the myeloid marker MPO and CD117. Positive for CD33 and positive for CD13. Positive for CD34 and HLA-DR immaturity markers. Absence of expression of B markers CD19, CD79a and negative for T markers CD3c and CD7. Negative for CD14, CD11b and CD64. Conclusion: Acute hypereosinophilic syndrome.

Resultados

On 07/17/2019, a biopsy of the popliteal region was performed with a finding of nonspecific chronic dermatitis with small vessel vasculitis and frequent eosinophils. FIPIL1/PDGFR ALFA associated fusion gene screening was performed and was negative. He underwent corticosteroid therapy from 07/04/2019 to 08/10/2020, with clinical and laboratory response. However, sequentially, in an attempt to gradually wean the steroid, there was recrudescence of hypereosinophilia in peripheral blood and bone marrow, in addition to worsening and intensification of skin lesions. A therapeutic proposal for systemic antineoplastic chemotherapy with hydroxyurea was made, with a favorable clinical and laboratory response, with a percentage drop in eosinophils from 32% (4,672/mm³) in September 2020 to 3% (306 eosinophils/mm³) in April 2021. In September 2021 (3576 eosinophils/mm³) and December 2021 (2831 eosinophils/mm³), the patient had new relapses, having undergone methylprednisolone pulse therapy for 3 days from 11/24 to 11/26/2021 and a new pulse from 12/17 to 12/22/2021, with improvement, using hydroxyurea until the conclusion of this report, in April 2022 (306 eosinophils/mm³).

Image 01: Right upper limb with Wells syndrome

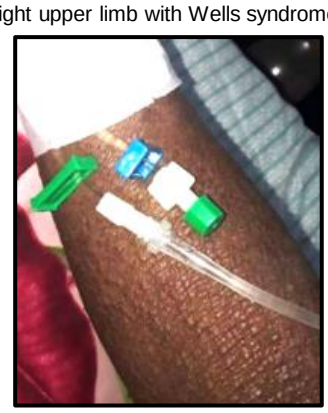
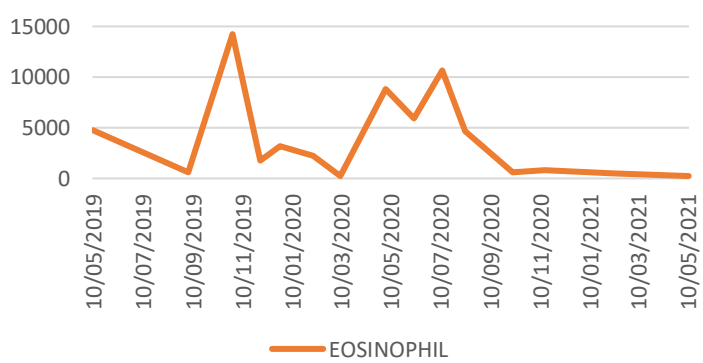


Image 02: Absolute number of eosinophils in peripheral blood



Conclusão

Hypereosinophilic syndrome is a rare disease characterized by persistent hypereosinophilia associated with evidence of some organic lesion with clinical manifestations and varied prognoses. All skin lesions should be investigated for diagnostic elucidation due to the risk of synchronous oncological pathologies. The Food and Drug Administration (FDA) approved on September 25, 2020 the monoclonal antibody directed to interleukin 5 mepolizumab for the treatment of patients with hypereosinophilic syndrome lasting more than 6 months in the USA. Efforts should be made to treat patients more homogeneously through national and international cooperation networks and sequential studies are needed to improve prognosis and outcomes.

Contato

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