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Hypereosinophilic Syndrome, Erythroderma as Clinical Debut. Case Report and Literature Review

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ABSTRACT

Eosinophilia is a common finding in clinical practice, but when elevated values of eosinophils are found, it constitutes a diagnostic challenge. Hypereosinophilic syndrome is a group of rare disorders defined by persistent blood hypereosinophilia >1.5 × 109/l and associated to organ damage that result in a wide variety of clinical manifestations: fatigue with nonspecific skin lesions, to endomyocardial fibrosis, neurological compromise and life-threatening evolution.

The prognosis of the disease is variable and depends on the variant and the availability of specific treatment. 1- 2

We present the clinical case of a patient with a history of B symptoms and the presence of persistent erythroderma. The patient underwent several complementary studies, including skin biopsy without presenting a definitive diagnosis. Laboratory with persistence of hypereosinophilia that guided us towards the diagnosis of Idiopathic Hypereosinophilic Syndrome.

KEYWORDS: Eosinophil, Dermatitis, Erythroderma

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INTRODUCTION

Eosinophilia is a common finding in clinical practice, but when elevated values of eosinophils are found, it constitutes a diagnostic challenge. Hypereosinophilic syndrome is a group of rare disorders defined by persistent blood hypereosinophilia >1.5 × 109/l and associated to organ damage that result in a wide variety of clinical manifestations: fatigue with nonspecific skin lesions, to endomyocardial fibrosis, neurological compromise and life-threatening evolution.

The prognosis of the disease is variable and depends on the variant and the availability of specific treatment. [1- 2]

We present the clinical case of a patient with a history of B symptoms and the presence of persistent erythroderma. The patient underwent several complementary studies, including skin biopsy without presenting a definitive diagnosis. Laboratory with persistence of hypereosinophilia that guided us towards the diagnosis of Idiopathic Hypereosinophilic Syndrome.

CLINICAL CASE

A 34-year-old male with no significant chronic-degenerative history. He started symptoms 7 months earlier, with fever of 39 °C predominantly at night, diaphoresis, asthenia, adynamia, and weight loss of 8 kg in one month. Later with itching and erythema in the extremities, spreading to the chest and abdomen. (Figure 1-2)

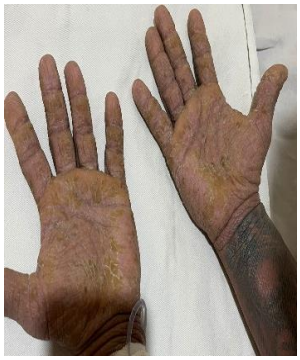

Fig. 1. Skaly skin and lichenification áreas.

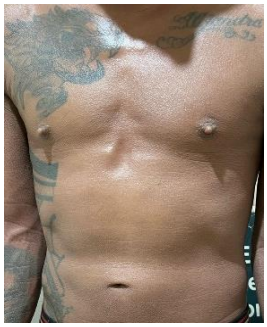

Fig. 2. Erythroderma of abdomen.

Streptolysins were reported in more than 1600, for which he was treated with amoxicillin / clavulanate, benzathine penicillin and betamethasone. Without clinical improvement, a skin biopsy was taken, finding *pityriasis rubra pilaris*; being treated with deflazacort, hydroxyzine, cetirizine, methotrexate and prednisone 0.5/mg/kg/hr, which he voluntarily discontinued due to edema of the extremities. Despite treatment he developed cervical, inguinal and axillary lymph node growths. Suspecting mycosis fungoides, a new skin biopsy was taken, which documented psoriasiform and spongiotic dermatitis with lymphohistiocytic infiltrate and eosinophilia.

Laboratories with 20,000 leukocytes, 10,000 eosinophils, 8,000 neutrophils, 1000 lymphocytes, 359,000 platelets. Due to the presence of eosinophilia, evaluation by hematology was requested, where a new Biometry is performed with persistence of eosinophilia at levels of 9,500 to 120,000. Biopsy of bone marrow showed: hypercellular bone marrow with accentuated eosinophilia, presence of immature forms in 10 %. (Figure 3-4).

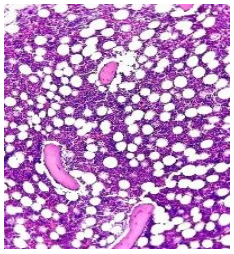

Fig. 3. Hypercelular bone marrow.

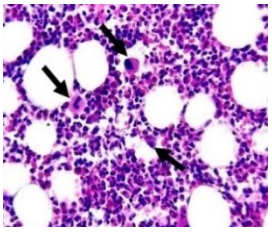

Fig.4. Eosinophilic promielocytes and mature forms.

Hypereosinophilic syndrome was diagnosed, TORCH and hepatitis panel were requested, both being negative. He started treatment with dexamethasone 16mg every 24 hours and imatinib, showing a decrease in eosinophilia and significant clinical improvement.

DISCUSSION

The 2016 World Health Organization recognizes a category of myeloid/lymphoid neoplasms with prominent eosinophilia (M/Leo) and genetic rearrangements of PDGFRA/B, FGFR1, or JAK2.

In a patient with myeloid characteristics, tests for myeloid/lymphoid neoplasms with prominent eosinophilia should be performed; if this pathology is ruled out, a diagnosis of chronic eosinophilic leukemia will be considered. If secondary causes are excluded, the diagnosis of idiopathic HES is possible.[1]

The initial concept of hypereosinophilic syndrome (HES) was introduced by Hardy and Anderson in 1968, later in 1975 Chusid et al established the first diagnostic criteria: persistent absolute blood eosinophil count greater than 1500/mm^3 for more than 6 months, with evidence of tissue and organ damage, without any identifiable cause of eosinophilia. In 2011, the International Working Group on Eosinophilic Disorders (ICOG-EO), maintained the criteria regarding the level of eosinophilia in the blood, but modified the duration of hypereosinophilia to 1 month, adding tissue eosinophilia and forms of asymptomatic, associated and eosinophilia. superimposed.

HypereosinophiliaIt is a rare, multisystemic and heterogeneous syndrome with significant mortality. It is characterized by an absolute concentration of eosinophils, greater than 1500/mm^3 on two consecutive occasions, persistent at least for 1 month. It is considered moderate with concentrations between 1,500 and 5,000/mm3, and severe, when it is higher than 5,000/mm3.

Regarding the pathophysiology,the cytokines that stimulate eosinophil production in the bone marrow are IL-3, IL-5, and granulocyte-macrophage colony-stimulating factor (GM-CSF); produced by CD4 and CD8 T cells in peripheral blood and inflamed tissues. Of these 3 cytokines, IL-5 causes terminal differentiation of eosinophils and is the target of medical treatment. Another pathogenic mechanism is an intrinsic defect of eosinophil-committed neoplastic progenitor cells, caused by mutations involving PDGFR or FGFR1.[3]

The condition occurs in men with an estimated 9:1 ratio, especially in FIP1L1-PDGFR α, who are almost exclusively male. However, in the lymphoid and idiopathic variant, the sex ratio seems closer to 1:1.The most frequently affected organs are skin, lungs, intestine, heart, kidneys, eyes and the peripheral nervous system. It is more common in men with a mean age of onset of 50 years. The most serious complication is heart disease. [4]

Hypereosinophilic syndrome is classified into:

- Family or hereditary.
- Secondary due to causes of reactive eosinophilia,such as helminth infection or drug hypersensitivity.
- Secondary (lymphocytic variant) with cytokine production from clonal and/or phenotypically aberrant T cells.
- Primary (myeloid), Molecular and cytogenetic studies are performed.
- Overlay.
- Idiopathic or of undetermined significance.[5]

The clinical presentations of HES are variable and can affect any organ. In the present case, the patient presented severe peripheral eosinophilia with erythroderma, proving dermal involvement by histopathological examination, without presenting compromise at any other systemic level. [6-7]

The most frequent initial manifestations are dermatological, which are characterized by being nonspecific and variable, therefore, clinical suspicion or knowledge of the entity is deficient.

Expressions in the skin are generally estimated at 69%, in a series of 44 patients with positive PDGFRA-MHES they were observed in 57% and up to 80% in 21 patients with lymphocytic variant CD3-CD4+ HES (LHES).

Cutaneous exteriorization requires a differential diagnosis with urticaria, mycosis fungoides, adverse drug reactions, contact dermatitis, and atopic dermatitis. Attention should be paid to pruritic erythematous papules, urticaria, angioedema, dermographism, oral and genital ulcers; erythema annulare centrifugum, acral blisters, and erythroderma. Histopathologic examination of the skin lesion is usually nonspecific, with viable eosinophilic infiltration. [8-9]

On the other hand, pulmonary and gastrointestinal manifestations are the second manifestation observed, followed by cardiac and neurological involvement. The most common symptoms of HES are fatigue, itching, and shortness of breath. [10-11]

Treatment should not be delayed. Patients with life-threatening complications such as the current case, which manifests as severe erythroderma, marked eosinophilia, should be treated with high doses of corticosteroids, pending a definitive diagnosis. The response to Imatinib, a tyrosine kinase inhibitor in FIP1L1-PDGFRα positive patients, reaches almost 100%. The low dose of 100 mg/day offers hematologic and molecular remission in most patients. [12]

CONCLUSION

Hypereosinophilic syndrome is a rare disease, sometimes fatal, and one of the organs that can be affected is the skin. Its diagnosis most of the time is given by ruling out other pathologies, and it is possible that only one organ is affected, as in the case of our patient. When the debut is with dermal involvement, the diagnosis is often delayed due to its pleomorphic manifestations and its insidious evolution. Therefore, it is crucial that in all cases of erythroderma, hypereosinophilic syndrome be considered as a differential diagnosis, to initiate timely treatment and improve the patient's clinical prognosis.

CONFLICT OF INTEREST.

The authors declare no conflict of int

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