East African Scholars Journal of Medical Sciences
Abbreviated Key Title: East African Scholars J Med Sci
ISSN: 2617-4421 (Print) & ISSN: 2617-7188 (Online)
Published By East African Scholars Publisher, Kenya

Volume-8 | Issue-6 | Jun-2025 | DOI: https://doi.org/10.36349/easms.2025.v08i06.004

OPEN ACCESS

Case Report

The Hypereosinophilic Syndrome: About 2 Cases

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Article History
Received: 01.05.2025
Accepted: 05.06.2025
Published: 13.06.2025

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: Hypereosinophilia syndrome is a group of rare diseases characterized by persistent blood eosinophilia (eosinophil count > 1.5 G/L for more than 6 months) associated with organ damage due to infiltration or degranulation of eosinophils. The diagnostic criteria include blood eosinophilia, involvement of multiple organs, and exclusion of other causes. There are three main subtypes: myeloproliferative, lymphocytic, and idiopathic. The first-line treatment is based on corticosteroids, and the prognosis of the disease is variable. Regular monitoring is essential. In this work, we report two distinct observations of HES.

Keywords: The Hypereosinophilic, Eosinophilic Polymorphonuclear Cells, Corticosteroids.

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INTRODUCTION

Hyper eosinophilia syndrome (HES) is a heterogeneous entity, grouping together very diverse clinicobiological and pathophysiological situations that share the common feature of chronic blood eosinophilia (lasting more than six months) and sometimes associated with tissue lesions induced by eosinophils (Eos). In this work, we report two distinct observations of HES.

OBSERVATION

Observation 1

A 42-year-old woman was admitted to our training for the etiological assessment of hyper-eosinophilia. She had been followed in pulmonology for asthma for 15 years and treated pulmonary tuberculosis. She was referred to hematology due to the discovery of persistent hyper-eosinophilia following an episode of exacerbation of her respiratory symptoms characterized by productive cough with sputum and nocturnal dyspnea associated with asthenia and night sweats.

The clinical examination found eczema on the hands as well as wheezing on pulmonary auscultation. There was no tumor syndrome. The blood test revealed leukocytosis at 19.3 Giga/L with eosinophilia at 11.5 Giga/L, total IgE levels at twice the normal range, LDH at 385 U/L, and vitamin B12 at 642 pg/ml. The protein electrophoresis showed an oligoclonal profile of immunoglobulins. The ANCA test returned negative. The bone marrow examination noted the presence of hyperplasia of the eosinophilic lineage. The bone marrow biopsy showed a rich marrow with medullary eosinophilia. The parasitological examination of sputum and stool came back normal. The ECG and echocardiogram were normal.

The thoracic scan showed a diffuse reticular infiltration (Figure 1) and the pulmonary function test indicated severe obstructive ventilatory disorder, with bronchoalveolar lavage revealing eosinophilic alveolitis. The search for Bcr-Abl, jak2V617F, and FIP1L1-PDFRα, as well as the search for mutations in PDGFRβ, FGFR1, and C-Kit all returned negative results. The diagnosis of myeloid SHE was made and the patient was placed on imatinib 400 mg/day with good progression.

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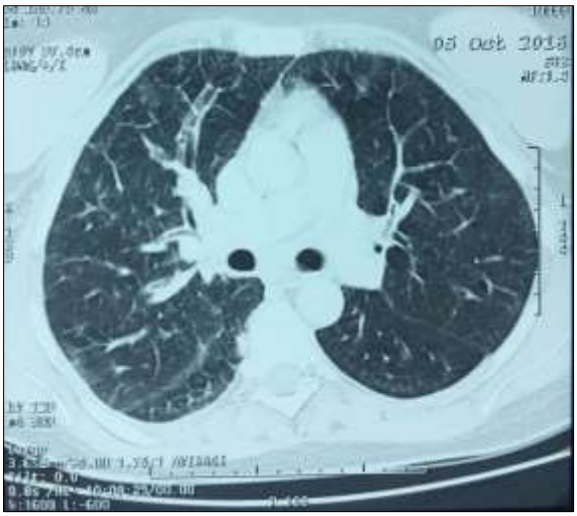

Figure 1: Thoracic CT showing diffuse reticular infiltration

Observation 2

A 41-year-old man consulted for edema of the lower limbs, associated with digestive symptoms consisting of chronic vomiting and abdominal pain. The clinical examination revealed lower limb edema reaching up to the root of the thigh with scrotal edema, associated with bilateral cervical, right axillary, and bilateral inguinal lymphadenopathy. There was no hepatomegaly or splenomegaly. The blood count showed leukocytosis at 37.94 giga/L with eosinophilia at 30.53 giga/L. The bone marrow analysis evidenced eosinophilia at 46%. The cytogenetic study showed no abnormalities in the bone marrow karyotype. The parasitological examination of the stools returned negative. The search for ANCA returned negative.

A CT TAP was performed showing gastric and intestinal thickening with retroperitoneal poly-ADP (Fig 2), complemented by gastroscopy and colonoscopy with staged biopsies that all returned normal results without any particularities. The ECG and echocardiography were normal. The search for the Bcr-Abl transcript or FIP1L1-PDFRα was normal. Flow cytometry and the search for T clonality were negative. The diagnosis of a lymphoid hyper-eosinophilic syndrome was retained, and the patient was put on corticosteroid therapy at a dose of 1mg/kg/day of prednisone with good clinical and biological improvement. However, he developed corticosteroid dependence.

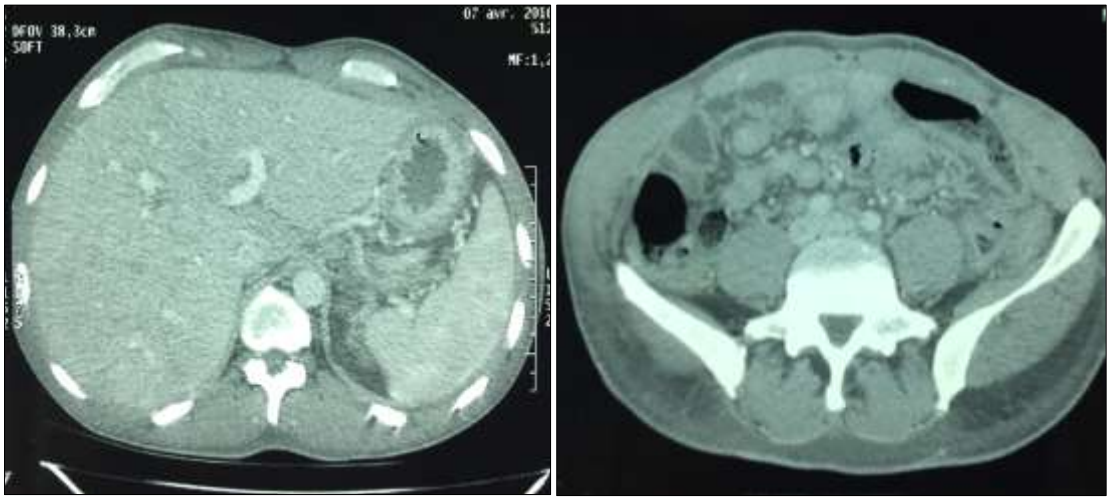

Figure 2: Abdominal CT showing gastric thickening (2A) and jejunal thickening with retro-peritoneal poly-ADP (2B)

DISCUSSION

The term eosinophilic hypersyndrome (HES) has been used for over 35 years to describe a wide variety of clinicobiological manifestations that share the common feature of unexplained blood eosinophilia (HE) (> 1.5 G/L), chronic (lasting more than six months), and associated with tissue lesions induced by eosinophil granulocytes, thus defining the criteria set by Chusid *et al*., Recent advancements have allowed for the identification of subgroups of patients within this heterogeneous entity, in whom a molecular explanation for the HE has been evidenced [1-3]:

1. Myeloproliferative or Myeloid Hyper Eosinophilic Syndromes

✓ Myeloproliferative syndromes "identified" and Hypereosinophilia:

chronic myeloid leukemia (CML) with translocation 9-22 responsible for a BCR-ABL fusion gene, other typical myeloproliferative syndromes often associated with the V617F mutation of JAK2 (polycythemia vera, essential thrombocythemia, idiopathic myelofibrosis), the 8p11 myeloproliferative syndrome (stem cell leukemia lymphoma), involving type 1 fibroblast growth factor (FGF) receptor, chronic myelomonocytic leukemia with translocation 5-12, involving the platelet-derived growth factor (PDGF) receptor, and finally, systemic mastocytosis with D816V mutation of C-Kit.

However, these chromosomal abnormalities were only exceptionally found in the SHE, and the myeloproliferative nature was often asserted based on a set of clinicobiological arguments: hepatosplenomegaly, myelemia, cytopenia (anemia, thrombocytopenia), elevation of vitamin B12, and reticulin fibrosis in the bone marrow.

✓ Chronic eosinophilic leukemia associated with FIP1L1-PDGFRA:

This is a translocation t(1;4) (q44;q12), a fusion gene FIP1L1-PDGFRA (F/P), initially resulting from an interstitial deletion of 800 kb on chromosome 4. This clonal cytogenetic anomaly has allowed, according to WHO criteria, the classification of this particular form of myeloproliferative disorder (MPD) as chronic eosinophilic leukemia F/P+ (CEL F/P+), the term most commonly used in the international literature.

2. Lymphoid Hyper-Eosinophilic Syndromes

The "lymphoid" variant of SHE results from a dysregulation of lymphocyte homeostasis with increased secretion of eosinophil growth factors (primarily IL-5) by Th2 lymphocytes. Various studies have highlighted, through flow cytometry, "aberrant" T phenotypes. These patients more often have skin involvement (including angioedema), and cardiac involvement is rare.

3. Idiopathic SHE:

Defined by an unexplained SHE. Long-term follow-up of these patients often allows them to be reclassified as myeloid SHE (often with FIP1L1) or lymphoid (T cell clonality).

The treatment of SHE has long relied on corticosteroids, although their eosinophil-blocking mechanisms are poorly understood. Hydroxyurea and interferon (INF) are used in cases of corticosteroid failure or for corticosteroid-sparing purposes. Initially proposed in combination for SHEM, these molecules remain indicated as second-line treatment (alone or in combination) in most variants of SHE, but their long-term efficacy and tolerance are limited, leading to treatment discontinuation in 75% of patients. Numerous other immunosuppressants have been proposed anecdotally in SHE: etoposide, vincristine, cyclophosphamide. Alemtuzumab, a monoclonal anti-CD-52 antibody, could be an interesting alternative. Finally, allogeneic stem cell transplantation has been reported in refractory SHE [1-5].

This therapeutic arsenal has been considerably modified thanks to a better understanding of the molecular mechanisms underlying eosinophilic disorders, allowing the development of targeted therapies, mainly tyrosine kinase inhibitors (Imatinib) and anti-IL-5 monoclonal antibodies (Reslizumab and Mepolizumab) [2, 3].

CONCLUSION

Despite these significant advances, nearly half of the SHE remain unexplained at the molecular level (idiopathic SHE). The identification of new tyrosine kinases (or new fusion proteins involving PDGFRA), the identification of specific biomarkers for lymphoid or myeloid variants should allow for better characterization of SHE in the future and an appropriate therapeutic choice.

Conflict of Interest: The authors declare that they have no conflict of interest.

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Cite This Article: Yassine Marjane, M.A. Aznag, H. Dergaoui, El M Awati, M. Chakour (2025). The Hypereosinophilic Syndrome: About 2 Cases. *East African Scholars J Med Sci, 8*(6), 210-212.