



A Case of Idiopathic Hypereosinophilic Syndrome

KEYWORDS

Hypereosinophilic syndrome, granulocyte-macrophage colony-stimulating factor, cytokines (IL-5).

Dr. Abhishek Sharma

A632 vidhyut nagar ,ajmer road, Jaipur (Raj)

Dr. Anil Mangal

SMS Hospital , Jaipur (Raj)

ABSTRACT *Hypereosinophilic syndrome is defined as a prolonged state (more than six months) of eosinophilia (greater than 1500 cells/ μ L), without an apparent etiology and with end-organ damage. Presentation is nonspecific and virtually any organ can be affected. We present here a case of persistent hypereosinophilia with lung and bone marrow involvement and without any identifiable cause so diagnosis of IDIOPATHIC HYPEREOSINOPHILIC SYNDROME was made.*

INTRODUCTION

The term hypereosinophilic syndrome (HES) was first introduced in 1968 by Hardy and Andersen [1]. criteria for idiopathic diagnosis of HES were defined by Chusid et al. 1975 [2]. HES is a rare syndrome characterized by persistent and marked eosinophilia along with organ system damage. HES has non specific clinical presentation but can be lethal without treatment, especially in patients with a myelodysplastic variant of HES. However the pathophysiology of HES is poorly understood, dysregulation of cytokines IL-5, IL-3, granulocyte-macrophage colony-stimulating factor [GM-CSF] responsible for the maturation of eosinophils is a primary feature [3,4] Of these, IL-5 have the greatest role in the regulation of eosinophil maturation[3]. There is no FDA-approved treatment for HES as yet; current strategies are designed to lower blood eosinophils and attempt to limit end-organ damage.

CASE REPORT

A 30yr old male admitted in our hospital presented with complaints of dry cough and fever for 2mnths and history of shortness of breath for 1month days. There was significant past history of dry cough for which he has been taking treatment from a allergy clinic.

On examination patient was pale and febrile. On systemic examination there was b/l rhonchi and end inspiratory creptations present in basal areas.

Lab investigations showed Hb-14.2 TLC-72310, PLT-1.73lkh, DLC-82% of eosinophils was seen. PBF – leucocytosis with marked eosinophilia. Total eosinophil count-52000cumm, s.lgE-2129. LFT, RFT and serum electrolytes were normal.

Sputum for AFB, gram stain, KOH and culture was normal but sputum for eosinophil count was more than 4%. Spirometry showed mild restriction. Stool for ova and cysts were negative. His autoimmune profile was negative.

HRCT thorax showed small nodules with ground glass haziness seen in periphery of lungs.

Next a bone marrow examination was done, which was suggestive of hypereosinophilic syndrome with normal erythropoiesis and mild depression of thrombopoiesis.

Since patient had persistent hypereosinophilia with lung and bone marrow involvement and without any identifiable cause a diagnosis of IDIOPATHIC HYPEREOSINOPHILIC SYNDROME was made.

DISCUSSION

Diagnostic criteria of hypereosinophilic syndrome are (1). Eosinophil count greater than $1.5 \times 10^9/L$. (2) Symptoms present for more than 6 months (3). Evidence of multiple-organ dysfunction, most frequently involving the heart, the central or peripheral nervous system and the lungs (4). Exclusion of known causes of eosinophilia (e.g., parasitosis, immunodeficiency and malignant disease)[2]

Three recently defined subtypes of HES have been described: 1) myeloproliferative characterized by chromosomal abnormality, tyrosine kinase involvement, elevated levels of dysplastic mast cells in the bone marrow. 2) lymphocytic variant characterized by an abnormal T cell (aberrant phenotype) with increased production of interleukin 5 (IL-5), interleukin 3 (IL-3), and GM-CSF, and idiopathic variants [4] The basic pathology of HES is the sequestration of eosinophils in organ tissues or systems. Eosinophil-derived neurotoxin, eosinophil cationic protein and major basic protein are enzymes released by eosinophils that cause endothelial damage and promote fibrosis, thrombosis and infarction.[5]

Clinical features: Any organ system may be affected in HES, but the heart, central nervous system (CNS), respiratory system and skin are commonly involved. Thromboembolic disease is not infrequent. Major symptoms of hypereosinophilic syndrome include the following:

The cardiac involvement is very frequent, and leading cause of mortality. This occurs in three stages: (1) acute necrosis in initial stage of disease that usually asymptomatic; (2) thrombotic phase; and (3) endomyocardial fibrosis. Common manifestation in these stage include chest pain, dyspnea, or orthopnea.

Hematologic manifestations include fatigue, anemia, splenomegaly (occur in 40% of patient) and hypercoagulability.

Neurologic symptoms are: Embolic or thrombotic strokes or TIA, neuropsychiatric symptom and peripheral neuropathy.

Respiratory manifestations are as follows: A chronic, persistent dry cough is the most common respiratory symptom. Patient may have recurrent angioedema. dyspnea pulmonary fibrosis are other manifestation. Bronchospasm and asthmatic symptoms are infrequent.

Other manifestation are arthralgias myalgias pruritus abdominal pain, diarrhoea, fever and night sweats [6]

D/D of hypereosinophilia- malignant disease, parasitosis and other infections.

CONCLUSION

HES can present nonspecifically, with multiple cutaneous, immunologic, ocular, rheumatologic and gastrointestinal manifestations.

Although, as in our case, other comorbidities such as asthma can mask the accurate diagnosis of HES with pulmonary involvement, early diagnosis often leads to the most appropriate management.

ACKNOWLEDGEMENT

I would like to express my deep sense of gratitude and reverence to Dr. Mahendra K. Agarwal and Dr.C.L Nawal professor dept of medicine for their constant guidance, encouragement and support.

The patient is the centre of medical universe around which all our work revolves and towards whom our efforts tend.

REFERENCES

1. Hardy WR, Anderson RE. (1968) The hypereosinophilic syndrome. *Annals Intern Med*; 68:1220–1229.
2. Chusid MJ, Dale DC, West BC, Wolf SM. (1975) The hypereosinophilic syndrome: analysis of fourteen cases with review of the literature. *Medicine (Baltimore)*; 54:1–27
3. Weller PF, Bubley GJ. (1994) The idiopathic hypereosinophilic syndrome. *Blood*; 83(10):2759–2779.
4. Roufosse F, Cogan E, Goldman M. (2003) The hypereosinophilic syndrome revisited. *Annu Rev Med*; 54:169–184.
5. Steven M. , Holland, John I. Gallin, *Disorders of Granulocytes and Monocytes*, (2015) *Harrison's Principles of Internal Medicine*, 19 Edition, Vol 1, 423.
6. Venkata A. S., Paul S., (2016) Hypereosinophilic Syndrome Clinical Presentation, [http:// http://emedicine.medscape.com/article/202030-clinical](http://emedicine.medscape.com/article/202030-clinical)