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Idiopathic hypereosinophilic syndrome: a case report

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Abstract

Idiopathic hypereosinophilic syndrome(IHES) is defined as eosinophilia > $1.5X10^9$/L persisting at least for 6 months, for which no underlying cause can be found and is associated with signs of organ involvement and dysfunction. A 55year old Male, presented with abdominal discomfort,hepatosplenomegaly. No history of parasitic infection, drug reaction or allergy was noted. Stool examination for parasite and other relevant investigations were unremarkable. Hemogram done on day 1 and day 21, In between Diethyl carbamezine was given. Hemogram showed total count 86X109/L and $67X10^9$/L, and absolute eosinophil count $36.1X10^9$/L and $25.5X10^9$/Lrespectively. Hemogram before 1 year showed total count of $56X10^9$/L and absolute eosinophil count $21.8X10^9$/L. Bone marrow aspiration revealed hypercellular marrow with increase no of eosinophilic precursor, without disproportionate increase in myeloblasts and normal erythropoiesis and megekaryocytopoiesis. Based on clinico-hematological findings, diagnosis of idiopathic hypereosinophilic syndrome was made.

Key Words: Idiopathic hypereosinophilic syndrome, eosinophilia, chronic eosinophilic leukemia.

INTRODUCTION

Idiopathic hypereosinophilic syndrome(IHES) is defined as eosinophilia > $1.5X10^9$/L persisting at least for 6 months, for which no underlying cause can be found and is associated with signs of organ involvement and dysfunction [1]. It is a rare entity, the true incidence of which is not known due to its overlapping features with chronic eosinophilic leukemia, not otherwise specified.[1]

CASE SUMMARY

55 year Male, presented to clinician with complain of abdominal discomfort and diarrhea. On examination, mild hepatosplenomegaly was noted. No history of parasitic infection, drug reaction or allergy was noted.

Investigations : USG abdomen was unremarkable except for mild hepatosplenomegaly. Stool examination for parasite negative. Hemogram was done on day1 and day21, In between Diethyl carbamezine was given. Hemogram showed total count $86X10^9$/L and $67X10^9$/L, neutrophils 37% and 40%, lymphocytes 09% and 08%, eosinophils 42% and 38%, monocytes 01% and 02%, myelocytes 06% and 07%, metamyelocytes 04% and 04%, myeloblasts 01% and 01%, platelets $284X10^9$/L and $262X10^9$/L, and absolute eosinophil count $36.1X10^9$ /L and $25.5X10^9$ /L respectively. Hemogram also done before 1 year which showed total count of $56X10^9$/L, eosinphils 39% and absolute eosinophil count $21.8X10^9$/L.

Morphologically, eosinophils were showing mainly hyposegmentation with occasional hypersegmented eosinophils. Few eosinophilic myelocytes were also noted(Figure I). Bone marrow aspiration revealed hypercellular marrow with increase no of eosinophilic precursor(Figure II), without disproportionate increase in myeloblasts. Erythropoiesis and megekaryocytopoiesis were normal. Patient is being followed closely for last 25 months and is healthy except for hepatosplenomegaly and persistent peripheral blood eosinophilia.

DISCUSSION

The term hypereosinophilic syndrome was first coined in 1968 to regroup patients with a number of closely related disorders all characterized by chronic increase in peripheral blood eosinophil levels and organ damage related to eosinophilic infiltration [2]. Current definition of Idiopathic HES was proposed by chusid in 1975 stating that eosinophilia > $1.5X10^9$/L persisting at

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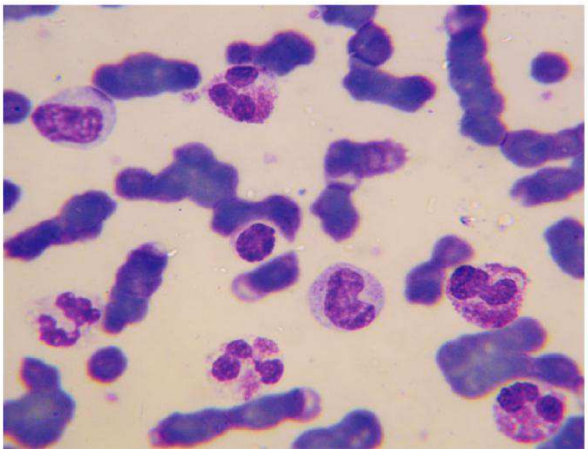

Figure 1 Peripheral Smear: Hyposegmented eosinophils and eosinophilic myelocytes. (Leishman Stain X1000).

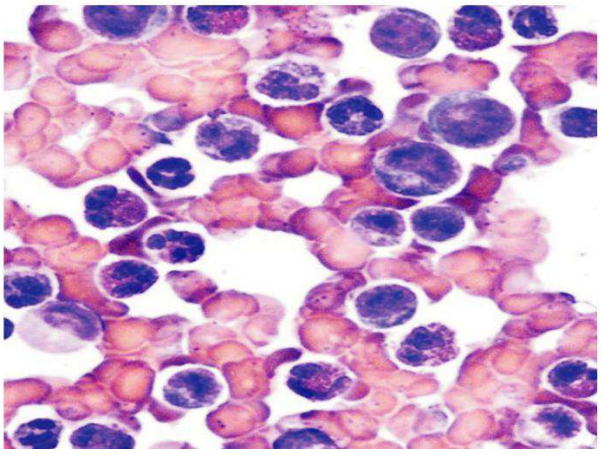

Figure 2 Bone marrow :Hypercellular marrow with increased eosinophil precursors(Leishman Stain X1000).

least for 6 months, for which no underlying cause can be found and is associated with signs of organ involvement and dysfunction.[2-4] In 2001, the WHO adopted a step-wise approach for Categorizing eosinophilia into primary, secondary (reactive), and idiopathic.[3] IHES is a rare entity, the true incidence of which is not known due to its overlapping features with Chronic eosinophilic leukemia, not otherwise specified.[1] It usually affect adults between the ages of 20 and 50 years with Male:Female ratio of 9:1 [2,4]. It is a multisystem disorder with invariable involvement of Bone Marrow and Peripheral Blood[1]. Clinical manifestations are variable and related to release of highly cationic molecules such as ECP(eosinophilic cationic protein), MBP(major basic protein), oxydating molecules such as EPO(eosinophilic peroxidase) and enzymes such as elastase and collagenase[2]. Hepatosplenomegaly is not uncommon and seen in 30-50% cases [1].Other systems like central nervous system, heart, skin and gastrointestinal tract may also involve. Peripheral Smear shows mature eosinophilia with occasional eosinophilic myelocytes. Eosinophils show increased size, sparse granulation with cleareareasofcyto plasm, and nuclear hypersegmentation or hyposegmentation.[1,4,5]

Neutrophiliaandmono cytosis of ten accompanies eosinophilia. Bone marrow reveals marrow with increase no. of eosinophilic precursor, without disproportionate increase in myeloblasts and normal erythropoiesis and megekaryocytopoiesis.[1]

Diagnosis of IHES is based on following criteria,[1,5] there is an eosinophil count of >1.5X10^9/L persisting for at least 6 months, reactive eosinophilia excluded by appropriate thorough investigation, AML, MPN, MDS and systemic mastocytosis are excluded, a cytokine producing T cell population excluded and, there is a tissue damage as a result of eosinophilia. Prognosis is good with 5 year survival is 80%, however, marked splenomegaly, increased blasts and dysplatic features in other myeloid lineages worsens prognosis.[1]

CONCLUSION

IHES is a rare hematological disorder with clinical heterogeneity. It is characterized by peripheral blood eosinophilia of unknown origin exciding 1.5X10^9/L persisting at least for 6 months. Present case was diagnosed as a IHES based on clinical features and hematological findings.

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