Case Reports

Hypereosinophilic syndrome

M Dissanayake[1] and **M K R Gunawardana**[2]

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Summary

We report a case of Hypereosinophilic Syndrome (HES) presenting with dermatitis of 6 years duration. She also had aquagenic pruritus of six years duration. Successfully treated with corticosteroids. Therapeutic alternatives are discussed.

The idiopathic hypereosinophilic syndrome (HES) is an infrequent entity defined by an eosinophilic count $> 1.5 \times (10)^9/1$ and the specific infiltration of different organs not attributable to another disease. Patients with HES may very rarely manifest as skin lesions as the first and only manifestation. The prognosis for these patients has been to a large extent improved by greater awareness of this condition.

Introduction

Relatively little is known about the physiological role of the eosinophil or of the basic mechanism responsible for the development of eosinophilia. Association of hyper-eosinophilia with blood dyscriasis, skin disease, autoimmune disease, bronchial asthma, parasitic infestation including helminthiasis and filariasis, drug reactions, Hodjkins lymphoma, etc. is well known[1]. A complete check up including clinical, laboratory and radiological data allows the identification of an etiology in majority of cases. In some cases the etiology remains obscure. Among these unexplained situtations, the rare idiopathic hypereosinophilic syndrome requires special attention, because of it's fatal course due to cardiac involvement which can remain clinically silent for a long period of time.

The term hypereosinophilic syndrome was first coined by Hardy and Anderson to include variety of disorders associated with eosinophilia[2]. The criteria introduced by Chusid et al have however defined the syndrome more specifically[3]. These criteria include:

1. Persistent eosinophilia of over $1.5 \times (10)^9/1$ (1500/Cu.mm) for longer than 6 months,
2. Lack of evidence of the known causes of eosinophilia, and
3. Multiple organ involvement.

A case of HES presenting with associated skin disease is described.

Case report

A 35-year-old female patient presented in March, 1996 with an itchy skin rash of six years duration. She also had aquagenic pruritus. Patient had asthma in childhood. No history of a cough or wheeze in the recent past.

On examination she had dry erythematous papular lesions prominent on both thighs (Figure 1), lower legs around the wrist and extensor aspects of elbows (Figure 2). Spleen was 2cm enlarged. Right axillary lymph nodes were palpable. On investigating patient was found to have a total white cell count of 18,800 with 85% eosinophilia (Absolute count of 15,980). ESR was 3mm in the first hour. A diagnosis of atopic eczema with tropical pulmonary eosinophilia was entertained and was treated with diethyl – carbamazipine, chlopheneramine, and betamethasone ointment locally. There was no response.

Further investigations were carried out to determine the cause of eosinophilia. Hb was normal. Blood picture shows mature eosinophilia. No AOC in stool. Filarial antibody titre was nil. Chest X-Ray, ECG and Echocardiogram were normal. Bone marrow examination showed hyperplasia of eosinophil precursors consistent with hypereosinophilic syndrome (HES). Axillary lymph node biopsy showed eosinophilic infiltrate without any abnormal cells. Clinical examination and investigation failed to show more usual manifestations of (HES), especially cardiac, pulmonary or neurological involvement.

[1] *Teaching Hospital, Kurunegala.*

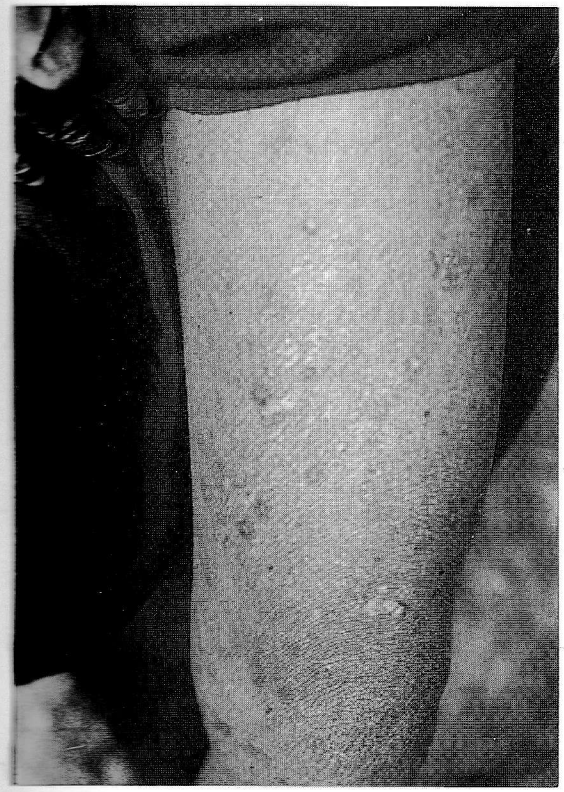

Figure 1. Dry erythematous papular lesion on the thigh

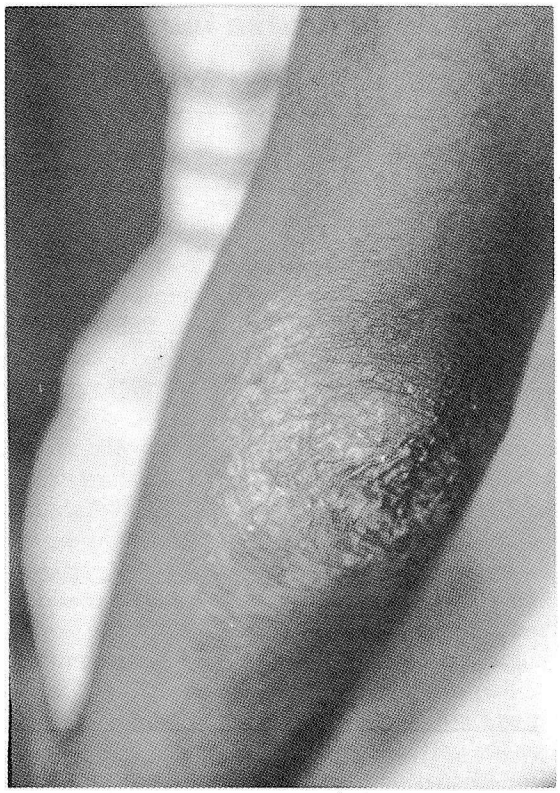

Figure 2. Lesion on extensor aspect of the elbow

Discussion

The diagnosis of hypereosinophilic syndrome was made according to criteria of Chusid et al, namely;

1. persistence of significant eosinophilia (over 1500) for more than 6 months. (Diagram-1).
2. lack for evidence of any known cause of eosinophilia. Although she has had childhood bronchial asthma she did not have pulmonary symptoms after childhood. The bone marrow and peripheral blood picture excluded an eosinophilic leukemia. Lymph node biopsy excluded a Hodgkin's lymphoma.
3. there was evidence of multiple systems involvement. There was evidence of reticuloendothelial system involvement (splenomegaly, lymphadenopathy and bone marrow infiltrate) and involvement of the skin. The cardiovascular, pulmonary and nervous system were not overtly affected.

Patient was started on topical clobetasole propionate and antihistamine. The effect on blood eosinophili and skin was insignificant. Prednisolone 15 mg daily resulted in marked fall in her circulating eosinophil count and considerable reduction in her pruritus. After terminating prednisolone eosinophil count rose to the original level. Restarted steroids and blood eosinophil started rising. A course of psoralance 10 mg thrice a week and UV treatment started. There was no significant reduction in symptoms and eosinophil count started to fall. Symptoms and eosinophil count controlled when steroids was re-instituted.

Skin lesions are commonly present in patients with an incidence of between 27% and 53%[4]. Skin lesion as the first and only (at least for several years) manifestation of the HES are very rare. The lesions are frequently non specific, either red pruriginous papular eruption over bony prominences or angioedema/urticaria[4]. Aquagenic pruritus has been described in the HES previously. The histology of the affected skin in the HES shows a mixed inflammation in the perivascular region.

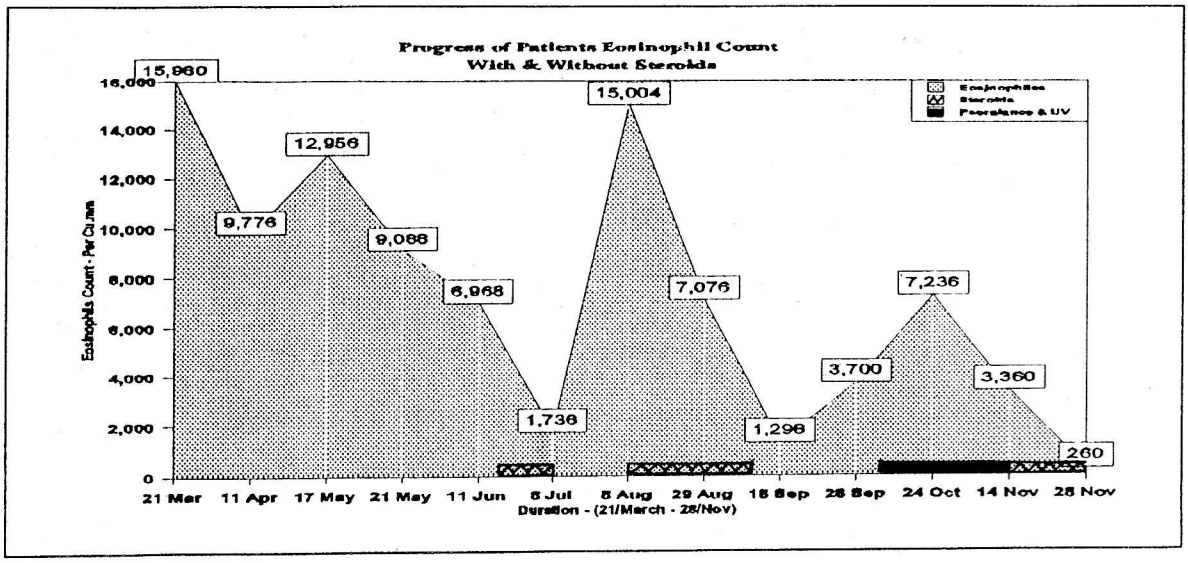

Diagram 1

Idiopathic hypereosinophilic syndrome is an infrequent entity characterized by sustained over production of eosinophilic infiltration of bone marrow, heart and other organs. The syndrome is associated with cardiac, hematological, pulmonary neurological and cutaneous involvement. It has wide range of severity, some patients suffer from severe fatal course while others present only skin involvement or asymptomatic. Nevertheless, the prognosis has been correlated with heart involvement which usually result in a restrictive cardiomyopathy.

Although a notable prolongation in survival may be achieved in these patients with the administration of glucocorticoids and cytolytic drugs; mainly hydroxyurea, there remain a group of patients who do not respond to these therapeutic measures. There are three case reports of successfully use PUVA in the HES and in all these cases the disease was limited to the skin[4]. A favourable experience with the use of alpha interferon in two patients with HES resistant to conventional treatment is reported[5].

HES with a variable prognosis be considered in patients presenting with obscure eosinophilia.

References

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