HYPEREOSINOPHILIC SYNDROME: REPORT OF A CASE AND REVIEW OF LITERATURE

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Abstract- Idiopathic hypereosinophilic syndrome represents a heterogeneous group of leukoproliferative disorders associated with prolonged eosinophilia of an undetectable cause with multi organ system dysfunction. It is a rare group disorder in children, most cases are reported in adult age group. We report a child with this syndrome who along with the usual features of the syndrome also had the presentation of cardiac and neurologic complications which did not respond to treatment.

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Key words: Hypereosinophilic syndrome, cardiac complications, neurologic complications

INTRODUCTION

Idiopathic hypereosinophilic syndrome (HES) is a rare condition with organ damage secondary to high eosinophilic counts and marked eosinophilic tissue infiltration (1-3). The eosinophil count exceeds 1500 cells/mm$^3$ and persists for at least 6 month unless death intervenes. It is rare in children; most cases are reported in women aged between 20 and 50 years (1-3). The clinical presentation is generally insidious but may be acute with sudden cardiac or neurologic involvement. In this article, we report a case of HES who presented with cardiac and neurologic complications.

CASE REPORT

A 3.5 years old male child was admitted in the pediatric ward of Imam Khomeini Hospital, Tehran University of Medical Sciences. He first presented with the complaints of high grade persistent fever along with weakness, nonproductive, non spasmodic cough and tachypnea for a period of 3 months. The child was treated symptomatically as outpatient and became asymptomatic but high grade fever, abdominal distention and tachypnea recurred within a week and he was admitted.

At admission, the child appeared sick with a pulse rate of 120/min, a respiratory rate of 54/minute and a blood pressure of 95/65 mmHg. Anthropometric measurements (height and weight) were normal for sex and age. Auscultation of chest revealed bilateral crepitations and breath sounds in lower right lobe were decreased. Systolic murmur grade III/VI at tricuspid site could be heard. The spleen was palpable 3 cm below the left costal margin and liver 4 cm below right costal margin in the midclavicular line. They were not tender, soft in consistency and had a smooth surface. Other systemic examinations were normal. There was no history of oliguria, dysuria, hematuria, jaundice, skin manifestation, bleeding tendencies, joint pains, worm infestation or allergic disorders. Moreover, there was no history of tuberculosis contact and no history of pica or geophagia. The child was not consuming any medications. There was no history of a similar illness in the family.

Investigations revealed hemoglobin of 7.7 g/dl, WBC of 28000 cell/mm$^3$ (polymorph 10%, lymphocytes 12%, eosinophils 71%) and platelet
Hypereosinophilic Syndrome

A case of HES was first reported in 1968 by Hard and Anderson but no details were available (1, 4). Chusid and coworkers later used restricted definition and criteria for diagnosis of HES as persistent eosinophilia of more than 1500 cells/mm³ for at least 6 months or death before 6 months with signs or symptoms of HES, lack of evidence for any recognized cause of eosinophilia and signs and symptoms of multi-organ system involvement (1-5). Fewer than 30 cases have been reported in children below 12 years of age (3). The presentation include: sign and symptoms of multi organ involvement like as weakness, cough, dyspnea, myalgia, rash, fever, rhinitis (1, 6, 7). Any organ may be involved. The characteristic feature of HES in tissue damage related to the release of basic protein, eosinophil peroxidase, cationic protein and eosinophil derived neurotoxin (4). In NIH series hematological involvement was seen in all, pulmonary in 40%, skin in 56%, neurological in 64%, splenomegaly in 45%, hepatomegaly in 35%, cardiovascular in 54% and ocular involvement in 18% (1, 8, 9). In our case the bone marrow, liver, spleen and lung were involved with no peripheral organ damage. The organ involvement was consistent with other reports (2). Myeloproliferative diseases or acute eosinophilic leukemia are considered as differential diagnosis (1). Characteristics of lymphoproliferative include presence of hypogranular, vacuolated eosinophils, presence of Philadelphia chromosome and decreased alkaline phosphatase (1, 5-7). Acute eosinophilic leukemia is of special concern as a differential diagnosis. The conversion of idiopathic HES to

DISCUSSION

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Fig. 1. Sever eosinophilic infiltration in liver tissue.

Fig. 2. Sever eosinophilic infiltration in lung tissue.
leukemia is rare. The pulmonary infiltration with eosinophilia syndrome should be kept as a differential diagnosis, especially where there is history of recurrent wheezing or dyspnea.

A patient with HES who has no organ dysfunction or symptoms despite high AEC needs no treatment except for a close follow up at 3 to 6 monthly intervals (2, 4, 6). The symptomatic patients should be treated with prednisolone therapy until clinical improvement occurs and then the dose should be tapered (1, 4, 5, 10). Symptomatic patient non responsive to steroids should be offered chemotherapeutic agents. Common chemotherapeutic drugs used include hydroxyurea, vincristine, 6 mercaptopurine, busulfan and chlorambucil. Hydroxyurea because of absence of leukemogenic effect and oral administration is used frequently. Interferon alpha and cyclosporine have also been found to be useful in HES (11). Imatinib mesylate is a promising drug in the treatment of idiopathic HES with response seen as early as one week after starting treatment, although further data are awaited for its use in the pediatric population (11).

Long term prognosis of patients with idiopathic HES contributes to be rather poor with 40% reported mortality at 10 years (11). Although mortality of HES is high, adhesive medical treatment can result in significant clinical improvement. The most common mode of mortality in HES is damage to heart and CNS by eosinophilic infiltration. Thrombectomy in thrombo-embolism, endocardial resection in endocardial fibrosis and valve replacement in severe regurgitation of mitral and tricuspid valve can be life saving (2, 3). NIH published a follow up of 50 patients diagnosed and treated over 11 years, with variable survival rates depending on the major organ involved. Patients with cardiovascular complications had a low survival rate. In conclusion, HES represents an emergency in children and it may be complicated by vital organ involvements. Our case highlights the importance of through screening in a symptomatic patient with marked eosinophilia.

REFERENCES