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Hyper-Immunoglobulin E Syndrome in a Neonate: A Case Report Bir yenido?anda Hiperimmunglobulin E sendromu: Olgu sunumu

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Abstract

Hyper-immunoglobulin E syndrome (Job syndrome) is a rare primary immunodeficiency with variable presentation, characterized by recurrent infections, facial dimorphism, eczema, scoliosis, joint hyper-extensibility, pathologic fractures, very high IgE (>2000 IU/mL), severe eosinophilia and variable impaired T cell function. We present a case of Hyperimmunoglobulin E syndrome in neonate with review of the literature. J Microbiol Infect Dis 2013; 3(3): 144-146.

Index terms— hyper-immunoglobulin E syndrome, recurrent infections, neonate.

1 Introduction yper-immunoglobulin (Ig) E syndrome (HIES or Job

Syndrome) is a rare primary immunodeficiency generally characterized by recurrent infections such as staphylococcal cold skin abscesses and pneumonia, eczema, scoliosis, joint hyperextensibility, pathologic fractures, a typical facial appearance, craniosynostosis, very high IgE, severe eosinophilia, and variable impaired T cell functions. The mechanisms responsible for hyperproduction of IgE and eosinophils in patients with HIES are presently unknown. Generally the onset of HIES occurs in children and elderly individuals. 1,2 HIES may have variable presentation, and laboratory values in different age groups. 3,4 Author ? ? ?: Department of Pediatrics, Dicle University, Medical School, Diyarbakir, Turkey. e-mail: ilyasyolbas@hotmail.com Author ?: Department of Dermatology, Dicle University, Medical School, Diyarbakir, Turkey. Author ¥: Diyarbakir Children's Hospital, Diyarbakir, Turkey.

We present a 15-days old newborn with HIES whose only have staphylococcal cold skin abscesses eosinophilia and high immunoglobulin E levels.

A fifteen-days-old male neonate born at 40 weeks of gestation by normal spontaneous vaginal birth to a 24 years-old mother without history of significant disease such as eczema or HIES in the family. The antenatal ultrasonography was normal. The patient was admitted to Dicle University Hospital at fifteenth day of his life, because of cold abscess that appeared 5 days before admission. On physical examination there was a 2x3 cm swelling compatible with cold abscesses in the anterior right knee area, right supraclavicular area, lateral right chest area and anterior left ankle area. He also had a characteristic facial appearance such a broad nasal bridge, cheilitis, thickened skin, and deepset eyes with a prominent chin and forehead (Figure **??**

2 Case Report

3 Discussion

HIES is a multi-system disorder with a wide range of clinical phenotypes and signs, including skeletal, connective tissue, and vascular abnormalities. 3 Most of patients with HIES suffer from recurrent staphylococcal infections

of skin and lungs. 4 Generally recurrent pyogenic pneumonias start in early childhood, and the most common infecting organisms are Staphylococcal aureus, Haemophilus influenzae and Streptococcus pneumoniae, Also mucocutaneous candidiasis is common in HIES. 4 Musculoskeletal abnormalities of HIES are scoliosis, osteopenia, minimal trauma fractures, hyperextensibility and degenerative joint disease. 3,5 The patients with HIES may have problem with development of their teeth. 6 Our case had multiple cold skin abscesses in the various regions of body but had no other stigmata of HIES at this age. Characteristic facial appearance of HIES include broad nasal bridge, cheilitis, thickened skin, and deep-set eyes with a prominent chin and forehead3. Our case had the similar characteristic facial appearance such as broad nasal bridge, cheilitis, thickened skin, and deep-set eyes with a prominent chin and forehead.

The two most consistent laboratory abnormalities in HIES are eosinophilia and elevated serum IgE. Over time, the serum IgE may decline in adults or may increase in newborn. 3 Demirci at al 7 found that IgE level of a two-month-old patient with HIES was 75.3 IU/ml (Range: 15-32 IU/ml), But in the same patients' they found IgE level 13,000 IU/ml after eight months. The patients with HIES have normal serum IgM, IgG, and IgA levels. 3 The diagnosis of HIES is usually made based on characteristic facial appearance and clinical features associated with high serum IgE level and eosinophilia. 5 Our patient had some of the characteristic features and laboratory findings. However definitive diagnosis is made on genetic basis such as STAT3.

Management of HIES currently revolve around prevention and treatment of infections. There is no cure for HIES at present. Therapy includes drainage of cutaneous abscesses followed by intravenous antibiotic therapy directed against mostly staphylococcal aureus. Prophylactic antibiotics and specific treatment is based on organ involvement. Immunoglobulin replacement therapy and some other treatments such as IFN-g, IFNa, histamine-2 antagonists, and cyclosporine have been tried, which seem to be useful in the management of patients with HIES. 2,8 Prophylactic antibiotic or antifungal prophylaxis (e.g., trimethoprim-sulfamethoxazole or fluconazole) should be recommended in the patients with HIES with recurrent sinopulmonary, cutaneous infections, mucocutaneous candidiasis and invasive fungal infections. 8 In conclusion, HIES may present with some features in the newborn baby. Recognition of leading signs of the disease will provide early diagnosis and prophylactic measures. [1]

Figure 1: H

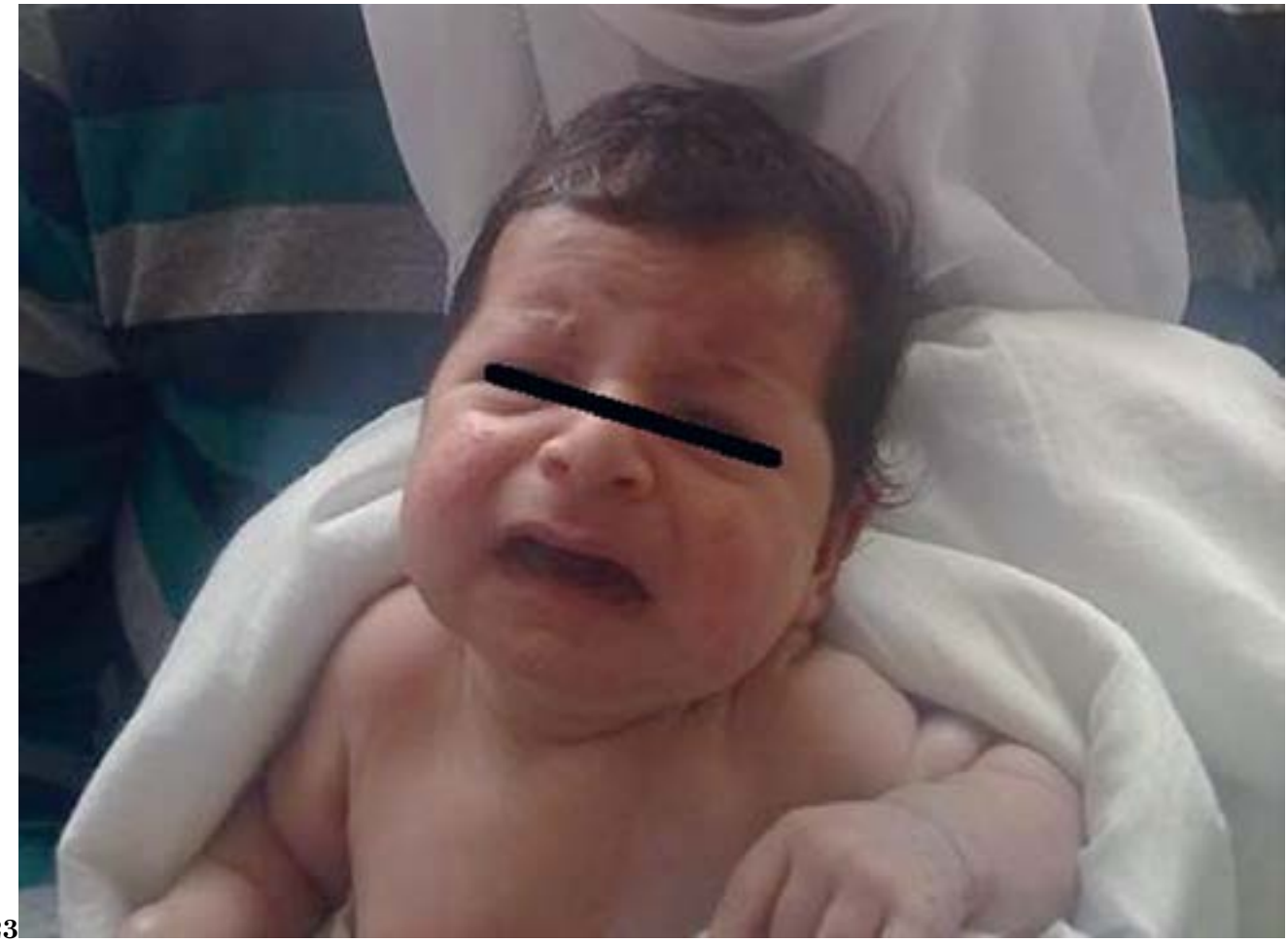
123 

Figure 2: Figure 1 :Figure 2 :Figure 3 :

Figure 3:

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