Case Report

Psoriasis in hyper IgE syndrome – a case report

Javad Ghaffari (MD) [*1]
Saeed Abedian- Kenari (PhD) [2]
Maryam Ghasemi (MD) [3]
Farzad Gohardehi (MD) [4]

1- Department of Pediatrics, Mazandaran University of Medical Sciences, Sari, Iran.
2- Department of Immunology, Mazandaran University of Medical Sciences, Sari, Iran.
3- Department of Pathology, Mazandaran University of Medical Sciences, Sari, Iran.
4- Mazandaran University of Medical Sciences, Sari, Iran.

*** Correspondence:**
Javad Ghaffari, Department of Pediatrics, Division of Clinical Immunology and Allergy, Boali Hospital, Sari, Iran.

E-mail: javadneg@yahoo.com
Tel: 0098 151 2234506
Fax: 0098 151 2234506

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Abstract

Background: Hyper IgE syndrome (HIES) is a rare primary immune deficiency, described as Job`s syndrome characterized by increased serum levels of IgE, eczema, recurrent cutaneous and pulmonary infections. In this paper, we presented a case of Hyper IgE syndrome.

Case Presentation: A 16-year-old Iranian boy presented with a one year history of skin lesions in knees and elbows was diagnosed of psoriasis disease. He had a history of recurrent infections including otitis media, pneumonia, diarrea and skin infection. Laboratory results showed increased level of total IgE and normal in other immunoglobulin. Histologic finding showed hyperkeratosis, parakeratosis of acanthotic epidermis with regular elongation of rete ridges diagnose psoriasis disorder.

Conclusion: In conclusion, this is the first case of hyper IgE patient with psoriasis disorder. We addressed the important laboratory findings and actual theories explaining possible association between hyper IgE immunoglobulinemia and psoriasis disorder.

Keywords: Hyper IgE syndrome, Psoriasis, Immune deficiency

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Hyper IgE syndrome (HIES) is a rare primary immune deficiency with multiorgan clinical manifestation, described as Job`s syndrome in 1966 (autosomal dominant and recessive) characterized by increased serum levels of IgE, eczema, recurrent cutaneous and pulmonary infections, although sporadic form of HIES was reported (1). Previous studies found that the signal transducer and activator of transcription (stat3-AD HIES) cause classic HIES but recent studies have show that the dedicator of cytokine sis 8 (Dock8, ARHIES) and or the tyrosine kinase 2 (Tyk2, ARHIES) probable causes HIES (2, 3). The incidence of HIES is about 1:1000000. Both males and females are involved equally (1-3). Pathogenesis and etiology of HIES is not clear but change in Th1/Th2 ratio, chemokine and defect in TH17 are involved in HIES. The clinical manifestation include; course facial, short stature, osteoporosis, fractures, scoliosis, craniosynostosis, hyperflexible joint, eczema, delay shedding of primary teeth, and recurrent bacterial infections (skin abscesses, pneumonia with pneumatoceles (4).

Some cases had malignancy (such as lymphoma and leukemia), arterial malformation (such as coronary artery aneurysms), viral infections (such as molluscum contagiosum and herpes simplex), neurological symptoms (such as partial facial paralysis, hemiplegia and central nervous system hemorrhage) (1, 5, 6). Arnold Chiari malformations (7), ophthalmologic (such as xantholasma, giant chalazia and strabismus) (8). Laboratory abnormalities in HIES are eosinophilia and elevated serum IgE. Increased IgE (often over 2000 IU/UL) may decrease and even reach normal values in adulthood (6). In HIES, serum IgG, IgA and IgM levels are typically normal. The prevalence of psoriasis in children ranged from 0% to 2.1%, and in adults it varied from 0.91% to 8.5%, therefore, psoriasis is a chronic, immune-mediated skin disorder that affects 1-3% of the general population worldwide (9).

Although, the skin manifestations such as dermatitis, molluscum contagiosum and furunculosis were seen in HIES however, based on our knowledge, there was not any report of psoriasis in HIES till now. Therefore, in this paper, we report a boy with HIES associated psoriasis disease which is a rare presentation.

Case presentation

A 16-year-old Iranian boy presented with a one year history of skin lesions in knees and elbows was diagnosed of psoriasis disease (figure1). He was born NVD, the second child, birth weight of 2 kg from related parents, low Apgar intubated for 3 days because of respiratory distress. The first (boy) and the third (girl) child were normal. After 6 months, he had recurrent vomiting, cough and wheezing. He had recurrent hospital admissions because of persistent pneumonia.

Also, he had skin abscess, to form furuncle and folicolitis. He also had history of one time left knee septic arthritic, hearing loss due to recurrent otitis media, and delayed shedding in primary teeth. He had normal response to polysaccharide pneumococcal vaccine. Endoscopy showed esophagitis. Lung CT scan (HRCT) showed diffuse bilateral reticular injection often in the middle and upper lobes. Spirometry showed restrictive and obstructive patterns (FVC=65%, FEV1 =35%, FEV1/FVC=77%, MEF=42%). Echocardiography was normal. Kyphosis, pigeon chest, clubbing, short stature and FTT associated with bone age 8 were seen. Likewise, our patient had delayed shedding of primary teeth, kyphosis and two times extremities' fractures.

Chronic dermatitis, facial features include frontal bossing, broad nose and prominent lower lip (coarse facials) observed in our case psoriasis lesions were in typical locations such as below both knees and the exterior of elbows.

The lesions were white-colored scaling papule. We confirm the psoriasis lesions by biopsy. Skin prick test was negative.

We could not identify the genetic and molecular defects. Laboratory tests are shown in table1. Our patient underwent treatment with inhaled corticosteroid, bronchodilator, antibiotic prophylaxis and sometimes intravenous immune globulin IVIG).

Table 1. laboratory test results in our HIES

Total IgG=1250(mg/dl)	WBC=12000 per microliter
IgG1 = 721(mg/dl)	EOS=600per microliter
IgG2= 237(mg/dl)	NBT=96%
IgG3= 46(mg/dl)	CD3=59%
IgG4= 8(mg/dl)	CD4=32%
IgA = 110(mg/dl)	CD8=27%
IgM=244(mg/dl)	CD19=10%
CH50=135 U/mL	Isohemagglutinin Anti- A antibody =1/64
IgE=1435(mg/dl)	Isohemegglutinin Anti-B antibody= negative

Ig=Immunoglobulin

Thyroid function tests, sweat test and alpha-antitrypsin were normal.

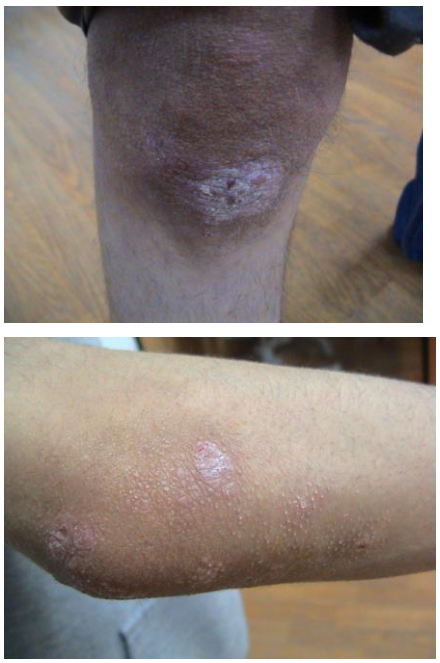

Figure 1. Psoriasis lesions on the elbow and knee in HIES

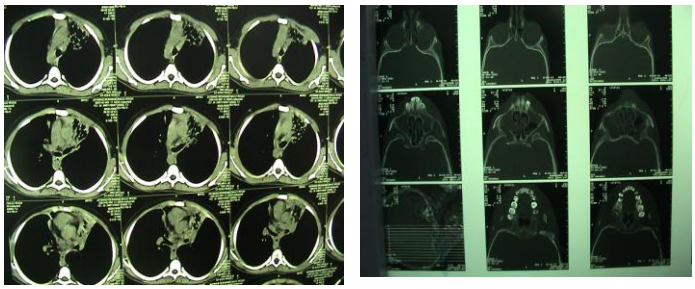

Figure 2. Bronchiectasis (left) and pan sinusitis (right) in HIES

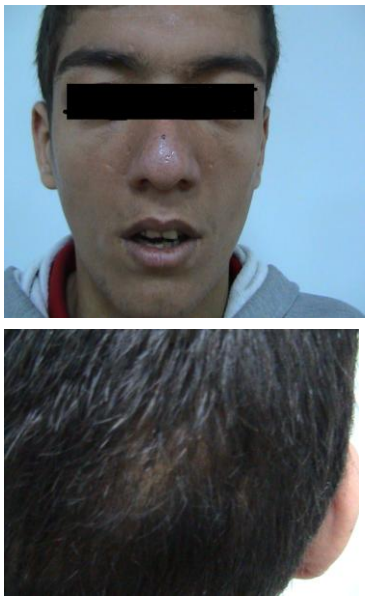

Figure 3. Coarse facial (left) and alopecia areata (right) in HIES

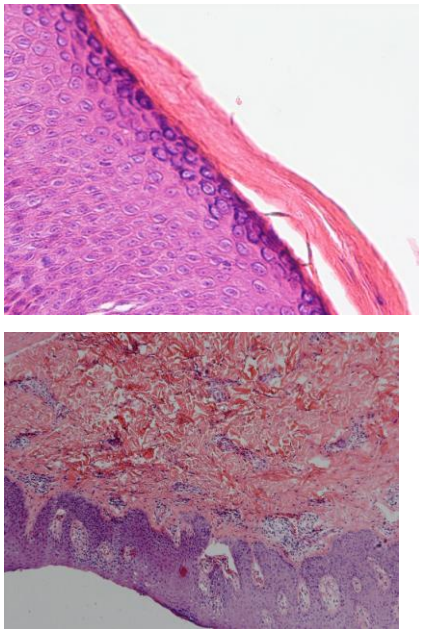

Figure 4. H and E stained slide showed hyperkeratosis, parakeratosis of acanthotic epidermis with regular elongation of rete ridges.

Discussion

HIES is a multi-system disorder with a extensive range of clinical manifestations, affects dentition, skeleton, connective tissue and the immune system, characterized by recurrent infection, dermatitis and elevated serum IgE levels (usually above 2000 IU/mL). After decades, some researchers found another rare manifestation such as coronary artery aneurysms in HIES which were not seen in our patient although he had a normal echocardiography (5). As our patient, recurrent respiratory tract infections (sinusitis, pneumonia) and otitis media were commonly caused by capsular bacterial such as staphylococcus aureus. Bronchiectasis and pneumotacele frequently complicate recurrent pneumonia and even in some reports it is pathognomonic in this group of patients which was also seen in our patient (10).

The same other reports that dermatitis is relatively common in HIES and it may be seen in early infancy, this case had recurrent staphylococcal skin infection (such as cold abscess) and diffuse eczematous rash that is intensely pruritic. In laboratory tests, we observed peripheral eosinophilia and elevated serum IgE levels (two most consistent laboratory findings) but the other classes of immunoglobulin were normal. One of the unusual manifestation of HIES is psoriasis. To our knowledge, this association has been reported very rarely such as psoriasiform and psoriasis in HIES after drug consumption (11, 12). Therefore, we report here an uncommon clinical manifestation in a patient with HIES who had psoriasis. Since psoriasis has an autoimmune pathogenesis, therefore, this disorder can occurr in immunodeficiency disease. We suggest our patient with classical form [skeletal, connective tissue, pulmonary abnormalities, boils, recurrent infections (bacterial and mucocutaneous candidiasis) and eczema] had AD form. Although, ARHIES lacks the somatic features and has marked viral infections and neurologic complications. Therapeutic options for HIES are prevention and treatment of infection. However, there is no specific treatment for HIES at present. Anecdote reports suggest that some patients have fewer infections with regular intravenous immunoglobulin (IVIG) (13). The role of bone marrow transplantation remains unclear (14).

In conclusion, HIES is a rare immunodeficiency disease that affect cutaneous such as eczema, psoriasiform and psoriasis. However, whether psoriasis lesion is independent or related to HIES still requires further studies.

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Conflict of interest: There is no conflict of interest in this paper.

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