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Atopic dermatitis like lesions in Hyperimmunoglobuline E syndrome

Afaf Khouna[1], Hazim Aburabie[2], Noufissa.Benajiba[3], Siham.Dikhaye[4], Nada Zizi[4]
[1, 2, 4] Dermatology Departement, University Mohammed First CHU Mohammed VI, Oujda, Morocco
[4] Laboratory of Epidemiological, Clinical Reaserch and Public Health, University Mohammed First CHU Mohammed VI, Oujda, Morocco
[3] Perdiatric Departement Faculty of Medecine Oujd, University Mohammed First CHU Mohammed VI, Oujda, Morocco

Abstract
Hyperimmunoglobulin E syndrome (HIES) is a recurrent multisystemic infectious syndrome, with reccurent cutaneous and non cutaneous infections, associated with atopic dermatiti slikelesion.We report a case of 10 years old girl who had recurent pulmonary, sinus and cutaneous infections associated with atopic dermatitis like lesion and hight level of Immunoglobulin E.

Keywords: Atopic dermatitis, Hyperimmunoglobulin E-infections

Introduction
Hyperimmunoglobulin E syndrome (HIES) is a recurrent multisystemic infectious syndrome, linked to a rare primary immunological disorder [1]. It manifests at birth or during early childhood, by severe and recurrent infections of the skin, sinus and the pulmonary tract, associated with chronic eczematiform dermatosis [2]. This clinical triad is associated with biological signs such as hyperimmunoglobulinemia E (> 2000 IU / mL) with moderate eosinophilia [1].

We report the case of a 10-year-old girl, from a consanguineous marriage, followed for repeated infections, with chronic cutaneous lesions of atopic dermatitis within the framework of a SHIE.

Observation
She was a 10-year-old girl, the youngest of four siblings, born of a 1st degree consanguineou smarriage, having a history of recurrent infections: pulmonary, cutaneous type of abscesses, otitis, and sinusitis complicated by a perforation of the nasal septum. Having as family history, a sister followed for refractory iron deficiencyanemia, and a cousin with the same symptomatology who died at the age of 24 years. She was admitted to the pediatric department for the etiological assessment of repeated infections, and chronic skin lesions. The general examination found a pale patient, with facial dysmorphia associating aflattening of the base of the nose, hypertelorism and a Dennie Morgan fold (Figure 1). The dermatological examination found a patient with phototype IV, with diffuse xerosis of the skin and scratching lesions, as well as axillary, inguinal lichenified lesions at the elbow and popliteal folds (Figure 2). In addition, oval ulceration wa sobserved during scarring in the leftankle, following a cold abscess (Figure 3).The blood count showed a normal white bloodcell count of 4400 / mm3, with a normal eposinophillevel of 60 / mm3 PNE, hypochromic microcyticanemia of 9 g/dl of hemoglobinwith a very high ImmunoglobulinE level of 9165. IU / ml and a normal level of other immunoglobulins.

Discussion
HIES was described in 1966, associating cutaneous and pulmonary staphylococcal infections on a background of eczematous dermatitis, in a context of elevation of total IgE in patients with facial abnormalities. This disease has been called Job-Buckley Syndrome or HIES. Its genetic determinism was established in 2007 for the autosomal dominant form and in 2009 with a mutation of the STAT3 gene, leading to an intensification of the production of immunoglobulin E by B lymphocytes. The recessiveformislinked to the DOCK8 gene [2].

The average age of diagnosis is 6.8 years with extremes between 0 and 30 years. It is a very rare disease, clinically manifested by severe and recurrent skin infections, cold cutaneous abscess,lung and sinus infections, with chronic eczema [1]. The dominant form of HIES is characterised by a skin rash at birth or infancy, with papules and pustules affecting the face and scalp, wrongly taken as new born acne [2]. HIES can be revealed by atopic dermatit is whichis an inflammatory condition affecting about 15% of children, characterized by eczematoid skin lesions, cutaneousxerosis, and pruritus, affecting folds and "bastion zones" [3]. Lichenification predominates over erythema and edema, unlike classical atopic dermatitis [4], whichis the case in our patient. The minor signs are infraorbital pigmentation, subpermalfolds (Dennie-Morgan sign) [4], also present in our patient.

Staphylococcal infections are usually superficial in atopic dermatitis, whereas in SHIE infections are profound with deep abscess in the form of fluctuating, non inflammatory masses [3], whichour patient had developed in herleft ankle and was undergoinghealing. Skin biopsies usually show an eosinophilic infiltrate, and bacterial culture often highlights Staphylococcus aureus.

Other cutaneous manifestations are represented by chronic cutaneo-mucous candidiasis, present in 60% of cases [3].

The discovery of eczematous lesions with high level of immunoglobulin E and/or recurrent infections may suggest the

presence of HIES [2].

Morphological malformations are often present, such as facial asymmetry with hemihypertrophy, flattening of the base of the nose, prominent forehead with delayedloss of temporary teeth [3].

In our patient, the association of recurrent pulmonary, sinus and skin infections, atopic dermatitis like, facial dysmorphism and elevated IgE levels were sufficient to support the diagnosis of SHIE.

Therapeutic strategies are mainly aimed at the treatment and prevention of cutaneous and pulmonary infections and their complications. The occurrence of cutaneous or pulmonary abscesses imposes prolonged antibiotic treatments targeting the most commonly implicated organisms: staphylococci, pneumococci and haemophilus, associated with antifungalprophylaxis. Special attention should be paid to skin care. Bonemarrow transplantation improves immunodeficiency without affecting other dominant HIES abnormalities [2].

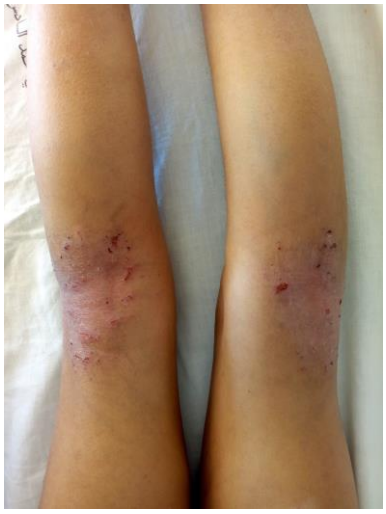

Fig 2: lichenified lesions of poplitealfossae

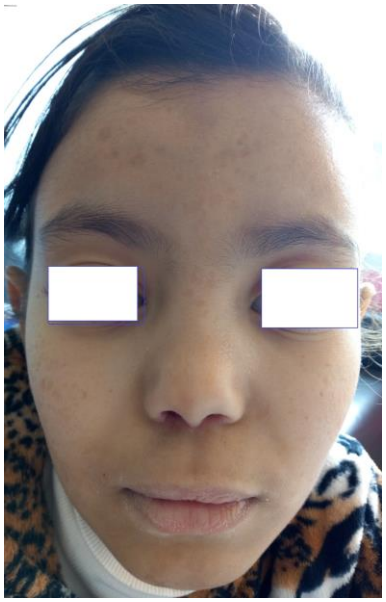

Fig 1: Hypertelorism and flattening of the base of the nose

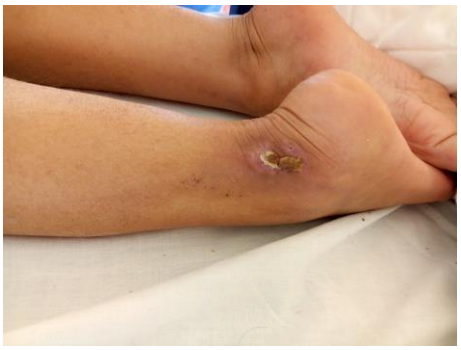

Fig 3: Cold abscess of the leftanklehealing

Conclusion

Atopic dermatitis is a chronic condition, widespread, the association with other signs, particularly reccurent infections must push clinical and paraclinical investigations in search of a genetic syndrome including HIES. The identification of the molecules responsible for SHIE may lead to an improvement in the control of atopic dermatitis and staphylococcal infections in the future [3].

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