

Case report

Unusual coexistence of molluscum contagiosum and verruca plana in a hyper-IgE syndrome

Xiao-Bing Lei, MD, Song-Mei Geng, MD, Wei-Hui Zeng, MD, Sheng-Shun Tan, Jun-Min Wang, and Sheng-Xiang Xiao, MD

From the Department of Dermatology, the Second Hospital of Xi'an Jiaotong University, Xi'an, Shaanxi, China

Correspondence

Xiao-Bing Lei
Department of Dermatology
Second Hospital of Xi'an Jiaotong University
157 Xi Wu Road
Xi'an
Shaanxi 710004
China
E-mail: leixb1968@163.com

Case Report

A 16-year-old boy presented with multiple painless papular lesions of 4 years' duration scattered extensively over the body. He also had grouped painless, nonpruritic, flat-topped papular lesions of 2 years' duration on the dorsa surface of the hands (Fig. 1).

The patient had suffered from hyper-IgE syndrome for 16 years since he was born. He reported that he had a history of severe eczema, recurrent pneumonia with pneumatoceles, skin abscesses and elevated IgE (IgE level: 28,149 IU/ml; reference range: 0–100 IU/ml), fulfilling all the criteria for the diagnosis of hyper-IgE syndrome.¹ In the past, he had

been treated with topical and systemic corticosteroids, topical moisturizer, antibiotics, antihistamines, and low-dose interferon- γ for eczema, skin infections, and pruritis. All these treatments had produced only short-term local regression with little resolution of his dermatitis and pruritis. Siblings, a brother and two sisters, of the patient and classmates in close contact with this patient had none of molluscum contagiosum or verruca plana on enquiry.

Physical examination revealed dry skin and dermatitis occurring on the face, neck (Fig. 2), trunk, bilateral axillae, groins, penis, perineum, and limbs, where there were also scattered multiple, firm, dome-shaped papules of 3–6 mm in diameter. Some of the papules had a central umbilication.



Figure 1 Multiple flat-topped papules on the dorsa surface of the hands



Figure 2 Multiple papular lesions scattered on the neck

1199



Figure 3 Acanthotic and cup-shaped lesion. In the cytoplasm of the prickle cells are molluscum bodies (hematoxylin and eosin, original magnification $\times 100$)

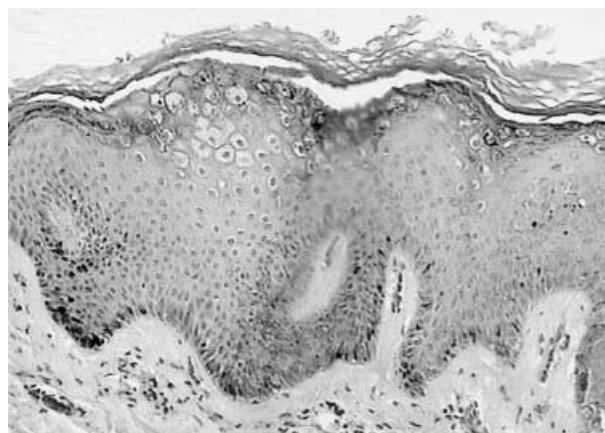


Figure 4 Lesion demonstrating a basket-weave pattern of hyperkeratosis in the stratum, slight acanthosis, and characteristic vacuolated cells in the upper spinous layer (hematoxylin and eosin, original magnification $\times 100$)

On the dorsa of the hands were 2–5-mm flat-topped, grouped papules, which were slightly erythematous.

On the left forearm was a cold abscess (1 $\times$ 1 cm in diameter). The culture of the pus in the skin abscess revealed methicillin-resistant *Staphylococcal aureus*.

Laboratory investigations revealed hypereosinophilia (8.1% of $4-10 \times 10^9/l$). T-lymphocyte subsets levels were as follows: CD3: 1450/ μl , CD4: 185/ μl , and CD8: 840/ μl . Purified protein derivative (PPD test) was negative (the patient was vaccinated with Bacillus Calmette–Guerin when he was 1 year old). Tetanus toxoid (TT) and mumps skin test were also negative. Neutrophil chemotaxis was normal. Testing for antibody to HIV was negative.

Histopathology of the dome-shaped papules on the neck showed acanthosis and cup-shaped. In the cytoplasm of the prickle cells, numerous molluscum bodies were formed (Fig. 3). A skin biopsy taken from a flat-topped lesion on the dorsa of the left hand revealed a basket-weave pattern of hyperkeratosis in the stratum, slight acanthosis, and characteristic vacuolated cells in the upper spinous layer (Fig. 4).

A diagnosis of molluscum contagiosum (MC) and verruca plana was made. The MC lesions on the neck were removed by curettage without complication. The remaining MC lesions and the verruca plana were treated with topical tretinoin cream (0.1%) for 5 weeks, but the treatment did not achieve its therapeutic effect.

Discussion

Hyper-IgE syndrome is a primary immunodeficiency disease. To date, approximately 200 cases have been reported worldwide. It is a multisystem disorder characterized by eczema, recurrent staphylococcal infections, pneumatoceles, reduced neutrophil chemotaxis, and variably impaired T-cell func-

tion. The most significant feature of this syndrome is that the elevated serum levels of IgE are uniformly found.^{2,3} In this report, we have described a patient with hyper-IgE syndrome co-existing with molluscum contagiosum (MC) and verruca plana. The coexistence of these disorders is rare and we suggest that this is the first such case in the literature.

The underlying cause of hyper-IgE syndrome remains unknown. There is no clearly defined defect of either T- or B-cell function. Reduced neutrophil chemotaxis is often described, and variable T-cell defects have been demonstrated in some patients. It has been hypothesized that hyper-IgE syndrome is associated with a Th1/Th2 imbalance.⁴ Most patients have a normal T-lymphocyte response to mitogens. Some studies have revealed that patients have defective antibody production;⁵ decreased production or responsiveness to cytokines;⁴ and abnormalities in some T-lymphocyte subgroups.⁶ Our patient's PPD, TT and mumps skin test were negative, a result that may be explained by the presence of an impaired delayed hypersensitivity response of the skin to antigens. These abnormalities render such patients susceptible to recurrent bacterial infections as well as to opportunistic infections. Various types of viral infections in patients with hyper-IgE syndrome, HSV,⁷ HIV,⁸ HPV,⁹ and MCV¹⁰ have been reported. In our case, the patient had two complications: MC and verruca plana. This is the first description of two unusual complications in a patient with hyper-IgE syndrome. Patients with defects in their cell-mediated immunity have an increased frequency of HPV and MCV infection, especially when the CD4+ cell counts are low.^{11,12} Two children with HIV infection suffered from extended MC and had low CD4+ cell count. After intensified antiretroviral therapy, their CD4+ cell count dramatically increased and their MC nearly completely disappeared.¹³ So it is well established that CD4+ cells

play a major role in protecting against MCV infection. In this report, we have described a patient with a low CD4⁺ cell count. It can be concluded that CD4⁺ cell studies should be considered in patients with hyper-IgE syndrome who are not only susceptible to recurrent bacterial infections, but are also susceptible to virus infections.

References

- Grimbacher B, Holland SM, Gallin JI, *et al.* Hyper IgE syndrome with recurrent infection – an autosomal dominant multisystem disorder. *N Engl J Med* 1999; **340**: 692–702.
- Buckley RH, Wray BB, Belmaker EZ. Extreme hyperimmunoglobulinemia E and undue susceptibility to infection. *Pediatrics* 1972; **49**: 59–69.
- Davis SD, Schaller J, Wedgwood RJ. Job's syndrome: Recurrent, 'cold', staphylococcal abscesses. *Lancet* 1966; **1**: 1013–1015.
- Chehimi J, Elder M, Greene J, *et al.* Cytokine and chemokine dysregulation in hyper-IgE syndrome. *Clin Immunol* 2001; **100**: 49–56.
- Vercelli D, Jabara HH, Cunningham-Rundles C, *et al.* Regulation of immunoglobulin (Ig) E synthesis in the hyper-IgE syndrome. *J Clin Invest* 1990; **85**: 1666–1671.
- Geha RS, Reinherz E, Leung D, *et al.* Deficiency of suppressor T cells in the hyperimmunoglobulin E syndrome. *J Clin Invest* 1981; **68**: 783–791.
- Hershko K, Hershko AY, Leibovici V, *et al.* Herpes simplex virus infection in a hyper-IgE patients: appearance of unusual mass lesions. *Acta Derm Venereol* 2002; **82**: 204–205.
- Lin RY, Smith JK. Hyper-IgE and human immunodeficiency virus infection. *Ann Allergy* 1988; **61**: 269–272.
- Sehrt P, Zielen S, Gerein V, *et al.* Hyper-IgE syndrome – two case reports of unusual complications. *Immun Infekt* 1994; **22**: 156–157.
- Martins MN, Tullu MS, Mahajan SA. Molluscum contagiosum and Job's syndrome. *J Postgrad Med* 2001; **47**: 268–269.
- Reynaud-mendel B, Janier M, Gerbaca J, *et al.* Dermatologic findings in HIV-1-infected patients: a prospective study with emphasis on CD4⁺ cell count. *Dermatology* 1996; **192**: 325–328.
- Sun XW, Ellerbrock TV, Lungu O, *et al.* Human papillomavirus infection in human immunodeficiency virus-sero-positive women. *Obstet Gynecol* 1995; **85**: 680–686.
- Horneff G, Wahn V. Mollusca contagiosa in HIV-infected children receiving optimal antiretroviral therapy. *Klin Padiatr* 2000; **212**: 83–84.