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Case Report

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Hyperimmunglobulin E Syndrome with Severe Cold Abcesses

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Abstract

Observation: Hyperimmunoglobulin E syndrome is an immundeficiency disease characterized by recurrent pyogenic infections and periferic eosinophilia. It may be sporadic or with autosomal dominant transmission. Cold abscesses seen in patients are pathognomonic for this disease. This paper reports a 4-year-old male patient having severe cold abscesses in inguinal region, both thighs, and below umbilicus. On examination and laboratory investigation, there were pneumonia, high IgE levels and eosinophilia. Staphylococcus aureus growth occurred in culture from wound and blood and the patient was given antibiotherapy accordingly.

Introduction

Hyperimmunoglobulin E syndrome (HIES) is characterized by increased IgE levels, recurrent skin and pyogenic infections, atopy-like dermatitis, and eosinophilia in peripheral blood. It is an immunodeficiency disorder which may be sporadic and sometimes with autosomal dominant trait. Cold abscesses seen in patients are pathognomonic [**1, 2**].

Case Report

A 4-year-old male patients admitted to our clinic with complaints of recurrent high fever, cough, sputum, and wounds in inguinal region. He had wounds in abdominal and inguinal regions, and legs beginning from one year of age and being recurred for 6 times. In addition to these wounds he had cough, sputum, fever, malaise and occasionally yellow discharge from his ear. Dermatological examination revealed five abscesses which localised from inferior side of umbilicus to superior portion of corpus penis and to bilaterally lomber regions, with sharp and irregular margin, with hypertrophic scar plaque, and with a widest diameter of 2x1 cm. There were no erythema, and edema nor increased temperature around abscesses. Again, there were atrophic and hypopigmented scars that was located scarcely in both thighs, and right upper quadrant (**Figure 1**). His body temperature was 39°C.

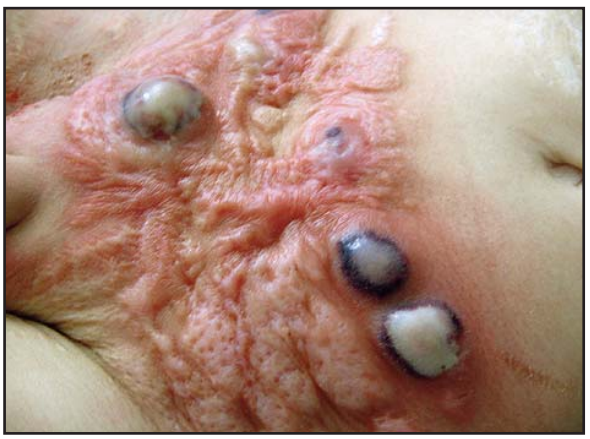

Figure 1. Five abscesses with sharp and irregular margin, with hypertrophic scar plaque

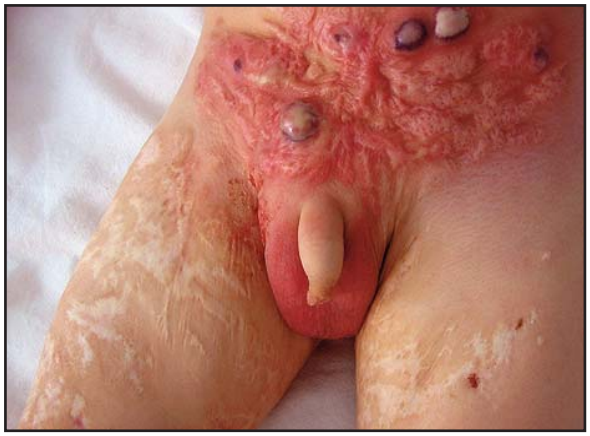

Figure 2. Five abscesses with sharp and irregular margin, with hypertrophic scar plaque

Laboratory investigation revealed that white blood cell was 36,000/mm^3 (65% neutrophil), IgE level 1900 ng/mL, ASO 1465 IU/ml, ESR 49/hr. A lot of candida spores were seen in feces. Staphylococcus aureus was cultivated in culture media from samples of blood and wound. The patient having pneumonia and chronic otitis was consulted with relevant departments. Patient had been evaluated with nitrozolium blue test and found negative.

At the beginning, the case was started systemic sultamicilline suspension and ibuprofen syrup. According to antibiogram obtained from blood culture, sultamicilline was changed by vancomycin HCl vial 140 mg 4x1. It was started dexamethasone sodium phosphate ointment, ciprofloxacin 0.3% ointment, pseudoephedrine HCl syrup for ear discharge. Nystatin suspension five times a day was given for candida spores seen in stool. Abscesses were drained surgically and treated topically bacitracine neomycine sulphate ointment two times a day, polyvinylpyrolidone iodine complex solution administerd twice daily. Hyperimmunglobulin E syndrome was diagnosed for the case in the context of recurrent otitis, pneumonia, pyogenic skin infections, IgE level increase, Staphylococcus growing in blood cultures, and eosinophilia in peripheral blood. Abscesses seen in the case were typically cold abscess.

Discussion

Hyperimmunglobulin E syndrome (HIES) is an immundeficiency syndrome characterized by an elevated IgE level, recurrent skin and systemic pyogenic infections, severe dermatitis-like atopy, peripheral eosinophilia and neutrophil chemotaxis disorder and it may have an autosomal dominant trait [**1, 2, 3, 4, 5**]. Despite its well-established clinical picture, underlying immunologic mechanism is not clear [**3**]. It is usually a sporadic disorder however autosomal dominant cases were reported [**1, 5**]. In these patients, decrease in T-cell count and function was determined. Decreased Th1 response and increased Th2 response was suggested in pathogenesis. Mutation and atopy in IL-4 receptor gene located in 16th chromosome were confirmed in HIES cases [**1**].

Skin findings of HIES usually starts in infancy and there is commonly athopy-like eczema. Lichenification is very severe [**1, 2, 3, 4, 5**]. Skin infections are frequent and usually start at infancy. *Staphylococcus* abscesses are present on the head, neck and intertriginous regions. These lesions are sensitive and painful on touching [**1, 5**].

Cold abscesses are rarely seen and pathognomic for hyperimmunglobulin E syndrome. However, they are not essential for diagnosis. Cold abscesses are big tumorlike lumps with fluctuation and cyst. They are neither hot nor with erythema without systemic symptoms. They are filled with pus and *Staphylococus aureus* growth usually occurs in cultures [**1**]. There were similar abscesses in our case and *Staphylococcus* was cultivated from the culture.

Pulmonary infections are frequent and severe in general. Bronchiectasia, bronchopleural fistula and Staphylococcal pneumonia are secondary to pneumatocells. Staphyloccoccal pneumonia associated with pneumatocell is mainstay in HIES diagnosis [**1, 6**]. Upper respiratory tract infections are also common including sinusitis, otitis media with discharge, otitis externa and mastoiditis [**1, 4**]. IgE level is frequently >2000 [**1, 2, 3, 4, 5, 6**]. Our case had pneumonia and otitis media. Staphylococcus was obtained from blood culture.

Prophylactic antibiotherapy, skin care and treatmend of existing infections are important factors for management of this disorder [**1, 6**]. Cimetidine, ascorbic acid, chromoglycate, levamizole, isotretinoin, cyclosporin A7, interferon gamma, interferon A, high dose gamma globulin (IVIG) 400 mg/kg/day for 5 days and plasmapheresis are current treatments [**1, 5**]. We report the case since this is a rare disorder and clinical findings are typical in our case.

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