ORAL ZINC IN JOB'S SYNDROME

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A 26-year-old man presented with coarse facies, recurrent cutaneous "cold abscesses", upper respiratory tract infection and candidiasis since infancy. Laboratory investigations showed extremely high serum IgE (2701 IU/1) level and pus culture from an abscess grew coagulase positive staphylococcus and Pseudomonas aeruginosa. Besides surgical drainage of the abscess and a short course of cephalexin, zinc sulphate (220 mg/day) was administered orally for six months. All lesions healed within a fortnight and clinical remission was observed for one year on follow up.

Key words: Job's syndrome, Zinc, Immunodeficiency

Introduction

Job's syndrome is a disorder of neutrophil chemotaxis characterized by development of recurrent cutaneous 'cold abscesses' -like lesions by staphylococci, eczema, sino-pulmonary infection, mucocutaneous candidiasis and hyperimmunoglobulinemia E.[1-3] Nevertheless, deep or visceral infection and bacteremia are rare. Familial trait and occurrence with atopic diathesis have been reported.[3] We report a case of Job's syndrome where good therapeutic response was seen with oral zinc besides a short course of cephalexin.

Case Report

A 26 - year-old man presented with recurrent wide spread cutaneous abscesses, throat infection and oral candidiasis since infancy. His parents, siblings and children were unaffected. Physical examination revealed coarse facies with large and broad nose, prominent cheeks, dark coarse hair over scalp and face. Multiple, large (10-30 cms), soft, fluctuating cutaneous abscesses with minimal inflammatory signs were seen over face, axillae, shoulders,upper arms and interscapular area. Healed lesions showed multiple, hyperpigmented, irregular scars of 10-40 cm size distributed over neck, chest, axillae,back,abdomen and inguinal area. Systemic examination suggested chronic rhinopharyngitis only.

Routine laboratoy and biochemical tests, VDRL test,ELISA tests for HIV I and 2 and skiagrams of chest/paranasal sinuses and ultrasonography of abdomen did not show any abnormality. Serum IgE level was extremely high (2701 IU/1) and pus culture from an abscess grew coagulase positive staphylococci and *Pseudomonas aeruginosa.* Histopathology of a recent skin lesion showed subcutaneous abscess. The abscesses were drained under the cover of cephalexin. Additionally, zinc sulphate (220 mg/day) was administered orally, for six months. The abscesses healed in a fortnight and didn't recur during one year follow-up.

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Discussion

An inflammatory response usually initiates within 2-4 hours of the local infection. In Job's syndrome inflammatory response is delayed mainly because of the defective chemotaxis. Under such circumstances micro-organisms continue to grow unhampered resulting in extensive 'cold abscess',[1,3] with minimal signs of inflammation.

High serum IgE found in Job's syndrome can directly interact with an antigen or sensitized leucocytes to inhibit release of the chemical mediators.[2]Nevertheless, the liberation of histamine may inhibit lysosomal enzymes and neutrophil chemotaxis directly[4-5], or indirectly by increase in cyclic AMP within the leucocytes.[6] Defective cell motility, impaired pseudopodia formation and abnormal microtubule formation are other intrinsic factors likely to impair the chemotaxis.[4-7] Defective neutrophil chemotaxis can be f;ound in Down's syndro;me, acrodermatitis enteropathica, atopic eczema, protein energy malnutrition, immunological and connective tissue disorders. These diseases often evidence low serum zinc levels and high susceptibility for bacterial infections of the skin and mucus membranes. Zinc deficiency is known to cause thymolymphoid suppression and elevation of serum free cortisol levels in these disorders.[8] Positive role of zinc in correcting the defects of chemotaxis, phagocytosis, acrodermatitis enteropathica and wound healing is well recognized. The favourable effect of zinc may be attributed to its effects o;n cellular fluidity, membrane stability, cytoskeleton, membrane receptors, calcium metabolism and secretory events of the phagocytic cells.

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