AUTOIMMUNE BULLOUS DISEASES

PARANEOPLASTEIC PEMPHIGUS AS THE FIRST MANIFESTATION OF T-CELL LYMPHOBLASTIC LYMPHOMA IN A CHILD

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We report a child with paraneoplasteic pemphigus (PNP) as the first manifestation of T-cell lymphoblastic lymphoma. A 6-year-old Chinese boy had a 6-month history of lid erythema, congested and oedematous conjunctivae, accompanied by a 4-month history of interrupted fever, cough, rash and mouth ulcer.

Physical examination showed inhalatory three-concave sign, tachypnea, a little rales in both lungs, extensive oral pseudomembranes, lip crusting, lid erythema, and congested and oedematous conjunctivae. Erythematous macules were present on his buccal mucosa and palate. There were erythematous maculopapules with scale over large areas of the cheeks, extensor aspect of fingers, hands, elbows, knees, feet and toes. Lymph nodes slightly enlarged.Pulmonary CT showed cloud flocculent high density lesions in the right middle lobe.

Skin biopsy showed hyperkeratosis, mild vacuolar degeneration of basal cells, and few scattered necrotic keratinocytes in epidermis. Besides, mononuclear cell infiltration was also seen in the upper dermis and follicle epithelial. Direct immunofluorescence (DIF) in perilesional skin showed negative result as well as the indirect IF analysis. Lymphoma node biopsy showed non-Hodgkin T-cell lymphoblastic lymphoma. Bone marrow biopsy showed normal. Thus the diagnosis of PNP was made, which was secondary to non-Hodgkin T-cell lymphoblastic lymphoma.

Due to his parents rejection of treatment, the patient died after followed- up for one year.