AUTOIMMUNE BULLOUS DISEASES

PARANEOPLASTIC PEMPHIGUS SECONDARY TO HODKING LYMPHOMA

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Background: Paraneoplastic pemphigus is an autoimmune blistering disease associated with an occult or previously diagnosed tumor, the most frequently reported associated malignancies are lymphomatoid and hematologic, B-cell lymphoma, chronic lymphocytic leukemia, Castleman's disease, Waldenstrom macroglobulinemia, and thymoma. The clinical, histological, and immunological features of paraneoplastic pemphigus have been clearly defined. It is characterized by polymorphic skin lesions and a chronic mucositis often refractory to conventional treatments.

Observation: We describe a case of 19-year-old male patient, who came to our department with a 1 month history of 6 kg weight loss, erosive mucositis and flaccid blisters disseminated, as well as fever and nocturnal diaphoresis. Previously, the patient was treated with multiple antibiotics, antivirals and analgesics without improvement. Physical examination revealed disseminated flaccid blisters and several maculo papular erythematosus plaques, as well as a severe erosive mucositis. Treatment with prednisone 1 mg/kg, topical mometasone and ophthalmic lubrication was started without improvement at 1 week. A contrast-enhanced chest CT scan showed anterior mediastinal mass and cervical lymph nodes. Leukopenia, neutropenia and lymphopenia was noted. Skin biopsy reported suprabasal blister containing acantholytic cells. A mediastinal lymph node biopsy showed Reed-Stemberg cells, a hallmark of Hodking Lymphoma.
During the 26 day of hospital stay, the patient developed a progressive respiratory failure leading to death.

Key message: Paraneoplastic pemphigus is a rare condition, usually triggered by lymphoproliferative neoplasia. The diagnosis depends on the correlation between the clinical and histopathologic evaluations. Our case represents a classic presentation of paraneoplastic pemphigus and suggest the importance of a comprehensive investigative work up and an early diagnosis. Currently, the treatment of this disease is difficult and often ineffective. The prognosis is poor, and the mortality rate is high.