



AUTOIMMUNE BULLOUS DISEASES

A CASE OF PARANEOPLASTIC PEMPHIGUS IN AN ADULT FILIPINO MALE

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Background: Paraneoplastic pemphigus is a rare blistering disease with a complex pathogenesis, distinct from classic forms of pemphigus. It is associated with a known or occult neoplasm, and refractory to therapy. Severe oral and conjunctival erosions are prominent features, with blisters and lichenoid eruptions. We report a 51-year old Filipino male with intractable stomatitis, extensive blisters and lichenoid plaques, associated with progressive weight loss and weakness. Diagnosis was based on clinical and histopathologic evaluation, with direct immunofluorescence.

Observation: A 51-year old Filipino male presented with intractable stomatitis, symblepharon, generalized cutaneous blisters and lichenoid plaques, associated with progressive weight loss and weakness. The patient was subsequently admitted and treated for anemia, multiple electrolyte imbalance and end-stage renal disease. He was given intravenous hydrocortisone and a high-potent topical steroid with antibiotic for the cutaneous lesions, which resulted in considerable clinical improvement. Additionally, he was co-managed by otolaryngology and ophthalmology specialists for stomatitis and symblepharon. Investigations to identify an underlying neoplastic condition were done on outpatient basis. Histopathology of skin punch biopsy showed suprabasilar, acantholytic intraepidermal vesiculobullous dermatitis; compatible with pemphigus variants. Direct immunofluorescence was consistent with paraneoplastic pemphigus.

Key message: Intractable stomatitis with extensive blisters has various differential diagnoses. Although rare, paraneoplastic pemphigus must be considered in middle-aged individuals, especially when associated with progressive weight loss and weakness—even before an associated neoplasm is identified. Clinical evaluation, histopathology and direct immunofluorescence are essential in diagnosing paraneoplastic pemphigus. We emphasize the importance of early diagnosis in order to initiate appropriate treatment and prevent severe, life-threatening complications.

