



AUTOIMMUNE BULLOUS DISEASES

PARANEOPLASTIC PEMPHIGUS: CLINICOPATHOLOGICAL PROFILE AND IMMUNOFLUORESCENCE STUDIES OF 8 PATIENTS.

Bm Cunha⁽¹⁾ - Rgr Carvalho⁽¹⁾ - Cg Santi⁽¹⁾ - D Miyamoto⁽¹⁾ - V Aoki⁽¹⁾ - Cw Maruta⁽¹⁾ - J Cury-martins⁽¹⁾ - M Menta Simonsen Nico⁽¹⁾ - Sv Lourenço⁽¹⁾

Hospital Of The University Of São Paulo Medical School, Dermatology, São Paulo, Brazil⁽¹⁾

Background: Paraneoplastic pemphigus (PNP) is a rare life-threatening autoimmune syndrome that may precede or occur concomitantly with a benign or malignant neoplasm. PNP often presents with polymorphic mucocutaneous lesions. It may resemble pemphigus, pemphigoid, erythema multiforme, graft-versus-host-disease and lichen planus, which poses a challenge to the diagnosis.

Observation: Eight patients (5 female; 3 male) were diagnosed with PNP from 2009 to 2018 with a mean age of 46 years old (32-57 years) at presentation. Lymphoproliferative disorders were the most common underlying neoplasms (3 patients had non-Hodgkin lymphoma (NHL) and 1 chronic lymphocytic leukemia), followed by thymoma (2/8), inflammatory myofibroblastic tumor (1/8) and Brenner ovarian tumor (1/8). PNP lesions preceded the diagnosis of the neoplasm in 3 out of 8 patients (1 thymoma, 1 NHL, and 1 inflammatory myofibroblastic tumor), whereas 5 had already been diagnosed with a tumor (2 NHL, 1 thymoma, 1 chronic lymphocytic leukemia, and 1 Brenner ovarian tumor), one of which developed lesions during recurrence. All patients developed mucocutaneous lesions involving oral mucosa. The most common histopathological features were lichenoid dermatitis (8/8), acantholysis (4/8) and vacuolar degeneration (4/8). Immunofluorescence (IF) studies demonstrated immune complex deposition in all patients. Direct IF displayed IgG (5/8) and C3 (3/8) between epidermal keratinocytes, and IgG (1/8) and C3 (3/8) at the basement membrane zone. Indirect IF exhibited intraepidermal intercellular deposition on 3/8 patients using human foreskin and on 6/8 patients using murine bladder as substrate.

Key Message: PNP presents a wide range of clinical features with prominent and recalcitrant mucosal involvement that may prompt earlier recognition and enable the diagnosis of an occult neoplasm. Adequate treatment of the tumor and PNP lesions has a significant impact on patient's prognosis and quality of life.

