



DERMATOPATHOLOGY

PEMPHIGUS VEGETANS LIMITED TO THE FACE INITIALLY MISDIAGNOSED AS SQUAMOUS CELL CARCINOMA.

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Background: Pemphigus vegetans (PVeg) is an infrequent pemphigus variant (1-2%). First described by Neumann as severe papillomatous vegetations (1.876); while the Hallopeau subtype is predominantly pustular and subsides more easily with treatment.

Mostly, the lesions are multifocal and mainly localized on the inguinal folds, intertriginous areas, periorificial areas and oral mucosa. To date there are only four facial cases reported of PVeg.

At histopathologic examination, acantholysis with neutrophilic and eosinophilic pustules in the epidermis. Direct Immunofluorescence (DIF) is positive for IgG and C3 in an intercellular pattern. Indirect reactivity to desmoglein (Dsg) 3, Dsg1 and desmocollins (Dscs).

Oral agents such as captopril, enalapril, penicillamine and, less commonly, physical or chemical factors have been implicated in the development of PVeg.

Exceptional manifestations may be misdiagnosed as neoplasia resulting in delayed diagnosis and treatment. Herein, we report a lesion on the face of PVeg initially misdiagnosed as squamous cell carcinoma (SCC).

Observation: A sixty-three-year-old man presented a recurrent lesion of the right frontal area.

Initially he was diagnosed in another center as a SCC with a punch biopsy.

The excisional surgery with free-margins was informed as Warty acantholytic dyskeratoma on a severe photo-damaged-skin without neoplasia. Six months later the lesion recurred in the same location. Physical exam: verrucous well-delimited hypertrophic 2 x 2,5 cm plaque with an eroded surface. Dermatoscopy: irregular vessels with keratotic plugs on a coppery-yellowish background. Re-examined at our center, the plaques showed severe acantholysis. Two 4 millimeters punch were performed (one on the lesion; other perilesional for DIF). The histopathologic examination revealed intense acantholysis with positivity for IgG and C3, in an intercellular granular pattern, diagnosing PVeg.

Key message: PVeg may mimic SCC and Hailey-Hailey disease. Follow-up and raised awareness in an unexpected evolution plus repeated biopsies by an expert may aid the





diagnosis in these difficult cases.

