



AUTOIMMUNE BULLOUS DISEASES

PEMPHIGUS VEGETANS: 10 CASES ACROSS THE SPECTRUM IN TUNISIA.

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Introduction: Pemphigus vegetans (PVg) is a rare vegetative variant of pemphigus. Two subtypes are recognized: the Neumann type and the Hallopeau type.

Objective: The present study aims to describe the aspects of PVg in Tunisia.

Methods: A monocentric retrospective cohort study was conducted including the cases of PVg.

Results: The study cohort consisted of ten patients with a sex ratio of 0.42. The average age at diagnosis was 49.18 years + 15.62. Seven cases occurred in the context of a pemphigus vulgaris (PV) and three in superficial pemphigus (PS). Intertriginous areas were affected in four cases associated with an involvement of the lips in one case. The lips were exclusively affected in one case. In the five others, neither the folds nor the lips were involved. Four patients were of the Neumann subtype, three were consistent with the Hallopeau subtype and three exhibited unclassified forms. The first patient was a 51-year-old man with a 25-year follow-up for PS who presented with multiple vegetating tumors of the scalp, forehead, and back with no skin erosions. Another atypical form consisted of bilateral vegetating lesions of the hallux in a 33-year-old man occurring during the follow up of a previously diagnosed PV. The last case was a 52-year-old patient with a seven-year history of PS who exhibited extensive vegetating and hyperkeratotic plaques of the back. In all three cases, the histology showed features of PVg.

Conclusion: Our case series of PVg illustrated the polymorphous aspects of this variant occurring as classically described in the context of PV but also in patients with PS. We also find that PVg can develop at the onset of the disease or years after. Finally, patients can present with the classical Hallopeau or Neuman forms but also with unclassified presentations.

