



AUTOIMMUNE BULLOUS DISEASES

THIRTY-SIX YEAR CLINIC-THERAPEUTIC EXPERIENCE IN PEMPHIGUS: A RETROSPECTIVE STUDY OF 277 CASES.

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Background: Pemphigus comprises a group of autoimmune bullous diseases that affect the skin and mucous membranes, with an in-hospital mortality of 4.5 - 6% in Venezuela. Currently, there is no registered epidemiological data.

Objective: To describe the clinical and epidemiological characteristics of patients with pemphigus at a hospital based specialized clinic.

Materials and methods: retrospective, longitudinal and descriptive study. We collected data of clinical and epidemiologic features from medical records of the bullous diseases section of the Dermatology Department from Hospital Universitario de Caracas between January 1982 and July 2018.

Results: 277 medical records were included. There was a female predominance, n=163 (58.8%); most of patients came from the Venezuelan middle region n=91 (51.99%); for most varieties of pemphigus mean age of onset was 45.81, and 22.3 years for IgA pemphigus and Fogo Selvagem (p = 0.00094), phototypes III and IV were prevailed (78.70%), most frequent diagnosis was pemphigus vulgaris n=178 (64.26%). Lesions usually initiated on skin=148 (54.36%) with moderate severity=130 (46.93%) p = 0.05, except to pemphigus vulgaris where n=74 (41.49% p=0) started on mucosa. Monotherapy with prednisone 42.2% was the most common choice of treatment. Average for cessation of new blister formation was of the disease was 25.77 DE±17,18 days. Complications were mostly infectious (67.50%); 61% (n=169) required at least one hospitalization, with an average of 44.28 days. A total 21.1% failed to complete treatment (p=0.14), and 25,19% (n=33) died during follow-up. In-hospital mortality by 2017 was 15.78%. The data obtained in this report is similar to literature. However, in-hospital mortality has increased due to lack of medical supplies that currently experienced our country.

Conclusions: Pemphigus is a rare and potentially fatal disease, therefore is it is important to keep epidemiological data updated and improve public health policies to reduce morbidity and mortality from pemphigus in Venezuela.

