



AUTOIMMUNE BULLOUS DISEASES

## THE FIRST CASE REPORTED OF PEMPHIGUS HERPETIFORM IN PREGNANCY

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Background: Pemphigus herpetiformis is a rare variant of pemphigus that clinically resembles dermatitis herpetiformis. Clinically, is characterized by erythematous urticarial plaques and tense vesicles that present in a herpetiform arrangement.

Observation: A 36-year-old female patient presented ulceration of the oral mucosa, tongue and lips (figure 01). During investigation, she was at the 10th week of gestation. High digestive endoscopy demonstrated discrete lacerations in the region of the esophagus. Biopsy suggested herpes virus infection. Treatment with aciclovir was initiated, without improvement.

Due to the clinical features of a bullous with mucosal involvement, hypotheses of pemphigus vulgaris, epidermolysis bullosa acquired variant pemphigus mucosae, cicatricial pemphigus and herpesvirus infection were made.

Due to the atypical setting and diagnostic difficulties, we initiated prednisone 1mg/kg/day. No immunosuppression was found, including HIV.

In the second trimester of gestation, the patient presented erythematous herpetiform eruption. (figure 02)

Biopsy showed suprabasal acantholytic dermatitis with infiltrate composed of lymphocytes and eosinophils (figures 03). Immunofluorescence was negative, because she was in treatment with prednisone.

At 26 weeks of pregnancy, the fetus presented acute fetal distress. The newborn evolved with early sepsis and died.

Azathioprine was associated with the use of prednisone with complete resolution.

One week after this initial improvement, she developed urinary focus sepsis and a new oral mucosal eruption. Azathioprine was suspended. She was treated with cefepime. ELISA showing anti-desmoglein antibodies 3.

Analyzing the clinical presentation, level of cleavage with eosinophils and ELISA, we conclude that is a case of pemphigus herpetiformis.

The patient presented complete improvement of the sepsis. Initiated dapsone. Patient maintains complete remission. (figure 04)

Key Message: We report the first case report of a patient with pemphigus herpetiformis





initiated during pregnancy. In addition, we have a herpetiform pemphigus that by initially affecting mucous membranes, made our report even rarer and more complex.

