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### RESEARCH ARTICLE

#### PEMPHIGUS VEGETANS OF NEUMANN: A CASE REPORT OF A RARE VARIANT OF PEMPHIGUS VULGARIS

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#### Abstract

Pemphigus vegetans is a rare variant of pemphigus vulgaris characterized by periorificial papillomatous vegetations. We present the case of a 77-year-old female patient with a history of type 2 diabetes and hypertension who presented with painful verrucous lesions on her tongue. The examination revealed a cerebriform tongue with small, hard, confluent verrucous swellings, which is a well-known sign seen in pemphigus vegetans. A biopsy revealed acantholysis and absence of keratinocyte necrosis, leading to the diagnosis of Neumann-type pemphigus vegetans. The patient was prescribed prednisone and trichloroacetic acid, which resulted in successful treatment. Management of pemphigus vegetans should be individualized based on the patient's clinical presentation and response to treatment.

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#### Introduction:-

Pemphigus vulgaris is a rare, autoimmune disorder of the skin and mucous membranes that is characterized by the production of autoantibodies, particularly IgG antibodies, against desmoglein (1). There are two subtypes : The Neumann and the Hallopeau type (2). Here we present a case of Neumann-type pemphigus vegetans in a 77-year-old female patient with a history of type 2 diabetes and hypertension. The patient presented with painful verrucous lesions on her tongue, which was characterized by a cerebriform appearance.

#### Case Report :

We present the case of a 77-year-old female patient, who was being treated for type 2 diabetes and hypertension, and presented to our clinic with painful verrucous lesions on her tongue. She reported experiencing these lesions intermittently for the past 15 years. During the patient interview, we discovered that she had been taking corticosteroids irregularly. Upon examination of the oral mucosa, we observed a cerebriform tongue (Figure 1), depapillated, pseudo-leukoplakic with heterogeneous coloration. The center of the tongue was covered with small, hard, confluent verrucous swellings. This cerebriform appearance is a well-known sign seen in pemphigus vegetans. No anomalies were detected during the rest of the dermatological examination. A tongue biopsy revealed a malpighian layer with a deep suprabasal cleavage, acantholysis, and absence of keratinocyte necrosis. The basal cells had a "tombstone" appearance. Additionally, the patient had elevated anti-intercellular substance antibodies at a titer of 1280. All these findings led us to conclude the diagnosis of pemphigus vegetans of Neumann. To treat this pathology, the patient was prescribed prednisone at a dosage of 2 mg/kg/day, with a gradual reduction in dosage, and 30% trichloroacetic acid applied every two weeks. The lesions disappeared as a result of the treatment.

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**Figures**

**Figure 1:-** Clinical image showing the cerebriiform tongue, a well-known sign in pemphigus vegetans, characterized by a typical pattern of sulci and gyri over the dorsum of the tongue, resembling the appearance of the brain's surface.

**Discussion:-**

Pemphigus vulgaris is a rare, autoimmune disorder of the skin and mucous membranes that is characterized by the production of autoantibodies, particularly IgG antibodies, against desmoglein, a transmembrane protein that is essential for maintaining intercellular adhesion in the skin and mucous membranes. These autoantibodies disrupt the normal function of desmoglein, leading to a process known as acantholysis, where the keratinocytes in the skin and mucous membranes separate from each other, resulting in the formation of blisters and erosions (1). Pemphigus vegetans, which is a rare variant of pemphigus vulgaris, was first described by Neumann in 1876. He can be differentiated into two subtypes based on their clinical occurrence, course, response to treatment, and prognosis : The Neumann type, which is characterized by periorificial papillomatous vegetations, and the Hallopeau type, which is characterized by pustular lesions that evolve into vegetations and preferentially affect the intertriginous areas. This subtype typically has a benign course with few relapses (2, 3). The cerebriiform tongue, which is observed in the clinical appearance of our patient (Figure 1), is a well-known sign in pemphigus vegetans, described as the "Premalatha sign." This distinctive feature is characterized by a typical pattern of sulci and gyri over the dorsum of the tongue, resembling the appearance of the brain's surface. He can be used as a clinical clue for its diagnosis pemphigus vegetans, especially when combined with other clinical features and histopathological findings (4). Based on the physical examination, we diagnosed our patient with Neumann-type pemphigus vegetans, and a skin biopsy was performed to confirm the diagnosis. The treatment of pemphigus vegetans is similar to pemphigus vulgaris and typically involves the use of systemic corticosteroids. However, response to oral corticosteroid treatment may be variable, and disease remission may not always be achieved (5). In our case, we opted to initiate

treatment with oral corticosteroids and switch to a systemic approach, if necessary, but this was not required, as our patient responded well to the initial therapy. In addition to corticosteroids, the use of immunosuppressants may be beneficial in improving disease remission rates and reducing the need for long-term corticosteroid therapy (5, 6). In some cases, other medications such as dapsone and retinoids have also been used successfully as adjunctive therapy (7). It is important to note that the management of pemphigus vegetans should be individualized based on the patient's clinical presentation, disease severity, and response to treatment.

**Conclusion:-**

In conclusion, we present a case of a 77-year-old female patient with painful verrucous lesions on her tongue, who was diagnosed with Neumann-type pemphigus vegetans based on clinical, histopathological, and laboratory findings. The patient responded well to oral corticosteroid treatment and the application of 30% trichloroacetic acid. This case highlights the importance of recognizing the cerebriform tongue appearance as a clinical clue for the diagnosis of pemphigus vegetans and individualizing the management of the disease based on the patient's clinical presentation, disease severity, and response to treatment. Further research is needed to identify alternative treatment options for patients who do not respond to conventional therapies.

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