Brunsting-Perry Pemphigoid Successfully Treated with Intralesional Rituximab

Icim Komurcugil[1], Nermin Karaosmanoglu[2], and Ibrahim Emre Bulut[2]

[1]Ankara Eğitim Ve Araştırma Hastanesi
[2]Ankara Training and Research Hospital

August 25, 2023

Abstract

Brunsting-Perry pemphigoid (BPP) is a variant of cicatricial pemphigoid characterised by tense, pruritic subepidermal bullae which eventually heals with atophic scarring. BPP has a predilection for head and neck area with little or no mucosal involvement. Average age of onset is 58 with a 2:1 male/female ratio. Here, we report a

Brunsting-Perry Pemphigoid Successfully Treated with Intralesional Rituximab

Introduction

Brunsting-Perry pemphigoid (BPP) is a variant of cicatricial pemphigoid characterised by tense, pruritic subepidermal bullae which eventually heals with atophic scarring.[1] BPP has a predilection for head and neck area with little or no mucosal involvement.[2] Average age of onset is 58 with a 2:1 male/female ratio.[3] Here, we report a case of BPP treated with intralesional rituximab injection.

Case Report

A 70-year old male patient was referred to our clinic with 3 month history of pruritic, non-healing ulceration on his face. The lesion showed no improvement from topical antibiotics prescribed in an outpatient setting. According to our patient, potent topical corticosteroid ointments led to temporary remission only.

Dermatological examination revealed 3x3 cm erythematous plaque with central ulceration on his infra-auricular area. (Fig. 1) There was no involvement of oral, nasopharyngeal, genitourinary and ocular mucosa.

Two 4 mm punch biopsies were obtained, one from edge of the ulcer and the other from perilesional skin. Histopathology revealed subepidermal splitting and diffuse lymphocytic infiltration in the papillary dermis. (Fig. 2) Direct immunoflourescence (DIF) analysis showed strong linear IgG and C3 positivity along the basement membrane.

According to these results, our patient was diagnosed with Brunsting-Perry pemphigoid. Intralesional rituximab was administered for treatment. Intravenous hydrocortisone and pheniramine maleate were given prior to the injections, as part of the premedication protocol. Four sessions of intralesional rituximab (10 mg/ml) injections were performed in total, each with a 2-week interval. 1 mg of rituximab (0.1 ml) was injected one centimeter apart until the entire lesion was covered. A noticable improvement was noted at the end of four sessions. (Fig. 3)

Discussion

BPP is a rare, autoimmune bullous skin disorder classified as a variant of mucous membrane pemphigoid (MMP). It was first reported in 1957 by Brunsting and Perry, as a scarring disease localised to the head and

neck, with no mucosal involvement.[4]

The pathogenesis of BPP includes an attack on the target proteins of the basement membrane by autoreactive antibodies. Antibodies targeting laminin 332, type VII collagen, and BP180 have been identified up to now. Autoreactive IgG targets these proteins with subsequent deposition of C3 complement, which can be demonstrated by DIF. This autoimmune cascade results in a lymphocytic infiltrate with significant eosinophilia. This disrupts the normal structure of the basement membrane, causing a separation below the epidermis.[1]

Treatment of BPP is challenging and should be initiated as early as possible. Mild cases can be treated with potent topical or intralesional corticosteroids whereas moderate-to-severe disease requires systemic therapy with immunosuppressive drugs such as prednisone, azathioprine, mycophenolate mofetil, and cyclophosphamide. [5,6]

Rituximab is a humanised IgG1 type monoclonal antibody against CD20 antigen, which is found on the surface of mature B cells. Rituximab is indicated for the treatment of non-Hodgkin's lymphoma, chronic lymphocytic leukemia, rheumatoid arthritis, microscopic polyangiitis and granulomatosis with polyangiitis (Wegener's Granulomatosis). Rituximab has become the first-line therapy for moderate to severe pemphigus vulgaris. Intralesional rituximab has been shown to be effective in recalcitrant mucosal lesions of PV. [7] In severe resistant MMP cases, rituximab has been quite successful, in combination with systemic immunsupressive or immunmodulatory treatment.[8]

Our patient was resistant to treatment with potent corticosteroids so we decided to administer intralesional rituximab as a novel therapeutic approach. Our case is the first reported case of Brunsting-Perry pemphigoid treated successfully with intralesional rituximab.

References

1. Blum FR, Sigmon JR. Successful treatment of Brunsting-Perry pemphigoid with dupilumab. *JAAD Case Rep* . 2021 Feb 23;10:107-109. doi: 10.1016/j.jdcr.2021.02.010. PMID: 33850992; PMCID: PMC8022106.
2. Zhou S, Zou Y, Pan M. Brunsting-Perry pemphigoid transitioning from previous bullous pemphigoid. *JAAD Case Rep* . 2020 Feb 19;6(3):192-194. doi: 10.1016/j.jdcr.2019.12.018. PMID: 32149175; PMCID: PMC7033299.
3. William DJ, Dirk ME, James RT, Misha AR, Isaac MN. *Andrews' Diseases of The Skin: Clinical Dermatology,* Elsevier; 2020.
4. Brunsting LA, Perry HO. Benign pemphigold; a report of seven cases with chronic, scarring, herpetiform plaques about the head and neck.*AMA Arch Derm.* 1957 Apr;75(4):489-501. doi: 10.1001/archderm.1957.01550160015002. PMID: 13410108.
5. Kourosh AS, Yancey KB. Therapeutic approaches to patients with mucous membrane pemphigoid. *Dermatol Clin.* 2011 Oct;29(4):637-41. doi: 10.1016/j.det.2011.06.022. PMID: 21925010.
6. Eichhoff G. Brunsting-Perry Pemphigoid as Differential Diagnosis of Nonmelanoma Skin Cancer. *Cureus.* 2019 Aug 16;11(8):e5400. doi: 10.7759/cureus.5400. PMID: 31620323; PMCID: PMC6793596.
7. Vinay K, Kanwar AJ, Mittal A, Dogra S, Minz RW, Hashimoto T. Intralesional Rituximab in the Treatment of Refractory Oral Pemphigus Vulgaris. *JAMA Dermatol.* 2015 Aug;151(8):878-82. doi: 10.1001/jamadermatol.2014.3674. Erratum in: JAMA Dermatol. 2015 Jun;151(6):680. PMID: 25536513.
8. Baffa ME, Corrà A, Maglie R, Mariotti EB, Montefusco F, Pipitò C, Senatore S, Quintarelli L, Caproni M, Antiga E. Rituximab in Mucous Membrane Pemphigoid: A Monocentric Retrospective Study in 10 Patients with Severe/Refractory Disease. *J Clin Med.* 2022 Jul 15;11(14):4102. doi: 10.3390/jcm11144102. PMID: 35887866; PMCID: PMC9325091.