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Pemphigus Vegetans In The Scalp: A Therapeutic Challenge

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ABSTRACT

Pemphigus vegetans is a rare variant of *pemphigus vulgaris* characterized by vegetating plaques in the flexures. A 49 – year - old male patient with *pemphigus vulgaris* developed in the third year of his disease a hypertrophic plaque in the scalp. Lesion´s biopsy has revealed acanthosis, papillomatosis of the epidermis and suprabasal clefts; a diagnosis consistent with *Pemphigus vegetans* was made. The patient was then put on shaving, mechanical debridement, clobetasol propionate and sulfamethoxazole – trimethoprim after the isolation of *Staphylococcus* aureus in biopsy fragment of the scalp lesion. After increasing azathioprine dose and starting with papain dressings on the long – standing lesion it was observed a slow and progressive clearing. A chronic infectious process linked to *Pemphigus vegetans* might explain the refractoriness to multiple therapeutic approaches. Papain has shown to be an effective and inexpensive alternative.

KEYWORDS

Pemphigus, scalp, Papain, Focal infection, Prednisone, Immunosuppressive agents

INTRODUCTION

Pemphigus vegetans [PV] is a rare variant of *pemphigus vulgaris* [1-2%], which affects intertriginous areas (mainly axillae and groins) and mucous membranes. The Neumann subtype is characterized by flaccid bullae/erosions and Hallopeau type by pustules. Both clinical forms develop into vegetating plaques. The histopathology reveals suprabasal acantholysis, epidermal hyperplasia, acanthosis, papillomatosis and intraepidermal abscesses. Corticosteroids are the first line of treatment. The significance of scalp involvement for the course of pemphigus remains controversial. Scalp disease is common in patients with pemphigus vulgaris, but it is not classic for pemphigus vegetans. According to our literature review on the subject, six cases of PV in the scalp have been published between 1984 and 2020 (Table 1). We report a case of a very recalcitrant scalp lesion aiming to analyze the therapeutic difficulties of this condition.

CASE REPORT

Male patient, 49 years old was hospitalized in 2011 with erosions and blisters on the scalp, face, trunk, arms, perianal area, groins, and oral mucosa. Skin histopathological examination confirmed the clinical hypothesis of *Pemphigus vulgaris*. Prednisone (2 mg/kg/day) was started and during the prednisone decrease several relapses occurred leading to the adjuvant therapies: twenty-eight pulses of cyclophosphamide and four cycles of immunoglobulin. In 2014 erythematous, boggy, crusted, vegetating and malodorous scalp plaque was observed (Figure 1); the skin biopsy revealed epidermal hyperkeratosis, papillomatosis and suprabasal clefts confirming the clinical impression of PV (Figure 2). At the same time vegetating lesions developed in the lip's commissures and groins. Two fragments were obtained through scalp lesion biopsy for bacteriological and mycological examinations. Fungal culture was negative; *Staphylococcus aureus* (CA MRSA) was isolated and treated for twelve months with oral sulfamethoxazole – trimethoprim (TMP – 6 mg/Kg/day). Since then the patient has been treated with the following schemes in sequence: shaving+ electrocurettage + silver sulfadiazine 1%; physiological saline 0,9%+ mechanical debridement + trichloroacetic acid 90% (TCA) daily; clobetasol propionate 0,05 cream. In 2019 azathioprine [3mg/kg/day] was added and papain 10% gel once a day has since been applied as the only topical therapy.

Table 1: Pemphigus vegetans of the scalp: reported cases (1984 – 2020).

				TREATMENT			
Author	**Nºof cases**	**Gender**	**Age**	**Topical**	**Systemic**	**Response**	**Follow-up**
Ahmed[1]	1 scalp, face groins	M	51	NR	Prednisone	rapid clearing	six years
Danopoulou[2]	1 scalp	F	54	clobetasol propionate	prednisolone mycophenolate	free of recurrent disease	four months
Rackett[3]	1 scalp, axillae	M	46	wet dressings keratolytic shampoos	prednisone,azathioprine, intralesional corticoid injection, dapsone, methylprednisolone pulse therapy, methotrexate	control	NR
Dhamija [4]	1 scalp, mouth nose, glans	M	45	NR	dexamethasone pulse therapy	80% clearing of scalp lesion	NR
Lehrhoff[5]	1 scalp	M	58	mometasone	prednisone, methotrexate	improvement	NR
Mori[6]	1 scalp	M	42	NR	prednisone, azathioprine	almost complete	two years

Abbreviations: NR – not reported / M – male / F – female

Figure 1: Crusted vegetating and suppurative plaque

DISCUSSION

In three described cases, scalp lesions were an isolated manifestation of PV; the case with the longest follow – up (six years) presented rapid involution without relapse (Table 1). Our patient evolving also for six years has had multiple relapses in addition to refractoriness of the lesion on the scalp.

Corticosteroid injections have been used in the treatment of many dermatoses. The indication for our case should be the persistence of the lesion despite systemic therapy [7], but the moisty and weepy conditions made it impossible.

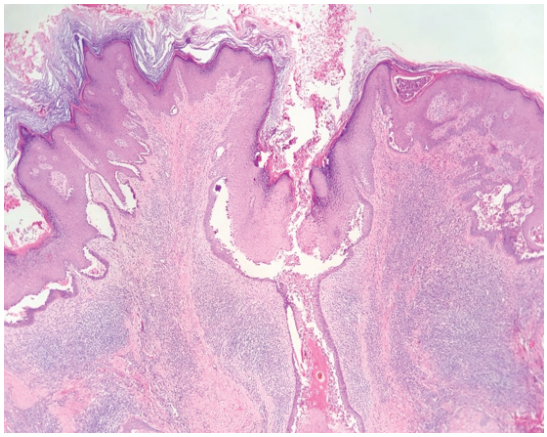

Figure 2: Hyperkeratosis, mild papillomatosis, prominent acanthosis and acantholysis with epidermal and follicular suprabasal clefts (HE, 40X)

Although shaving and mechanical debridement could be considered an aggressive approach, they were the best option for the eradication of a chronic supportive focus.

As treatment with SMZ – TMP has brought temporary benefit while administered and decolonization is only carried out in the absence of any skin lesions, we have faced a dilemma.

TCA 90% is indicated for the therapy of chronic ulcers, as a caustic and healing agent, favoring necrosis of the epidermis followed by epithelial renewal [8]. Papain 10% gel is indicated for the treatment of wounds of various etiologies [9,10]; it is extracted from the latex of the papaya green fruit (*Carica papaya*), and forms a mixture of proteolytic enzymes and peroxidases, leading to the proteolysis of the devitalized tissue [9,10].

Clobetasol 0.05% cream was employed when the secondary

infection improved; patients on systemic corticosteroid may not need high potency corticoid because systemic therapy should exert sufficient anti-inflammatory effects. In the present case this was not considered by the evolution of the disease itself. As papain is a low-cost medication and had led to slow but progressive improvement, we have decided to keep it. While the biologic agents that can be used for treating pemphigus are increasing, most of the world´s patients can afford only corticosteroid and some immunosuppressive agents. They are still unaffordable in the Brazilian Unified Health System which was the setting of this study.

CONCLUSION

We speculate that the lesion on the scalp was complicated by chronic infection linked to *Staphylococcus aureus* and favored by the state of immunosuppression. Papain, an enzyme of vegetal origin is widely used for the treatment of wounds; especially because of its inflammatory and healing properties has shown promising results (Figures 3 & 4).

Figure 3: A 6 years follow up: crusting and scaling in the previously vegetating area

Figure 4: A 6 years follow up: crusting and scaling in the previously vegetating area

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