

A Verrucous ‘Tumour’ On The Scalp

Case History

A 67-year-old Sikh male was diagnosed with pemphigus vulgaris in 1996 when he presented with oral erosions. His disease remained controlled on long-term, low-dose prednisolone therapy. In 2004, he developed a verrucous keratotic plaque over the occipital region of his scalp and it had been gradually enlarging (Fig. 1). Biopsies were performed in 2004 and 2008 and histological features in the epidermis of the specimens consist of orthokeratosis, parakeratosis, digitated epidermal hyperplasia, extensive suprabasal clefting, and acantholysis (Fig. 2). A dense infiltrate of plasma cells and lymphocytes were seen in the dermis. Direct immunofluorescence showed intercellular IgG and C3 deposition throughout epidermis. Indirect immunofluorescence was positive with antibody titers of more than 1/160. The levels of anti-desmoglein 3 antibody were also elevated at 206 U/mL (negative if value is less than 7) and 81.4 U/mL (negative if value is less than 14) respectively.



Fig.1 Verrucous keratotic plaque measuring 5 cm x 3 cm on the occiput with areas of erosion within the lesion.

What is the diagnosis?

- A. Squamous cell carcinoma
- B. Skin metastases from a primary cancer
- C. Pemphigus vegetans
- D. Vegetating cicatricial pemphigoid
- E. Vegetating pyoderma

Discussion

Pemphigus vegetans is the least common variant of pemphigus vulgaris. The diagnosis of pemphigus vulgaris is supported by the patient's background history, presence of erosions within the lesion, histological features of suprabasal diffuse acantholysis, features on direct immunofluorescence study, and positive indirect immunofluorescence and desmoglein 1 and 3 levels. The levels of desmoglein 1 and 3, in addition, correlate with disease activity. There are 2 forms of pemphigus vegetans. Our patient has the Neumann subtype, which is typified by bullae coalescing into vegetating masses, evolving into dry, hyperkeratotic and fissured lesions. This type is often refractory to treatment. The other form is the Hallopeau subtype, consisting of a polycyclic eruption of pustules that transform into papillomas and eventually flattening into dark brown plaques. Spontaneous remission is possible for this latter subtype.

The mainstay of treatment is systemic corticosteroids. Combinations with immunosuppressants (such as mycophenolate mofetil) or etretinate have also been tried with success.¹ Other treatment modalities include utilising extracorporeal photopheresis (ECP), carbon dioxide laser and anti-CD20 (Rituximab).^{2,3} It is important to note that the underlying disease activity in pemphigus should be adequately suppressed to prevent recurrence of the lesions.

In our patient, he received a variety of topical medications for his scalp lesion, consisting of betamethasone 0.1% scalp lotion, 5-fluorouracil, and 10% salicylic acid. He also received 2 courses of treatment with liquid nitrogen, all of which resulted in minimal improvement. He subsequently experienced some decrease in lesional size with clobetasol propionate cream 0.05%.

In summary, pemphigus vegetans is uncommon, with challenges in both diagnosis and management. Although

Answer: C

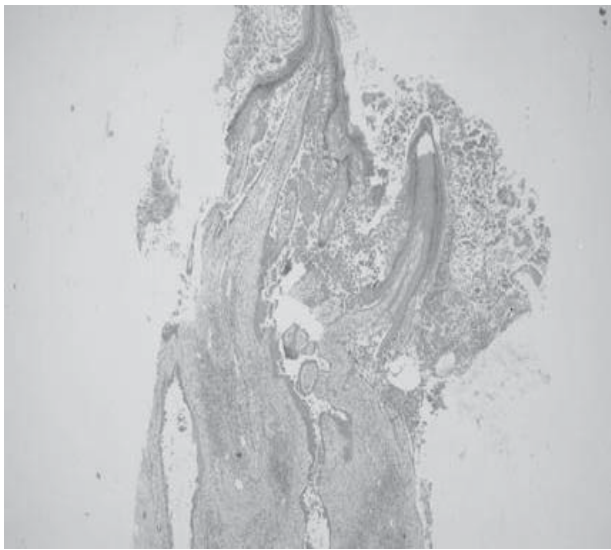


Fig. 2. Histology demonstrating orthokeratosis, digitated epidermal hyperplasia, suprabasal clefting, and a superficial perivascular dermal infiltrate. (Haematoxylin and Eosin, x 100).

it appears like a tumour, the occurrence of such a lesion in the background of pemphigus vulgaris should prompt the consideration of pemphigus vegetans and further investigations.

REFERENCES

1. Ichimiya M, Yamamoto K, Muto M. Successful treatment of pemphigus vegetans by addition of etretinate to systemic steroids. *Clin Exp Dermatol* 1998;23:178-80.
2. Lin MH, Hsu CK, Lee JY. Successful treatment of recalcitrant pemphigus vulgaris and pemphigus vegetans with etanercept and carbon dioxide laser. *Arch Dermatol* 2005;141:680-2.
3. Peterson JD, Chan LS. Effectiveness and side effects of anti-CD20 therapy for autoantibody-mediated blistering skin diseases: A comprehensive survey of 71 consecutive patients from the initial use to 2007. *Ther Clin Risk Manag* 2009;5:1-7.

Lucinda S Tan,¹ MBBS, MRCP (UK), MMed (Int Med), Hong Liang Tey,¹

MBBS, MRCP (UK), FAMS

¹National Skin Centre

Address for Correspondence: Dr Lucinda S Tan, National Skin Centre
1 Mandalay Road, Singapore 308205.
Email: symez@hotmail.com