

Brunsting-Perry Pemphigoid Mimicking Erosive Pustular Dermatitis of the Scalp

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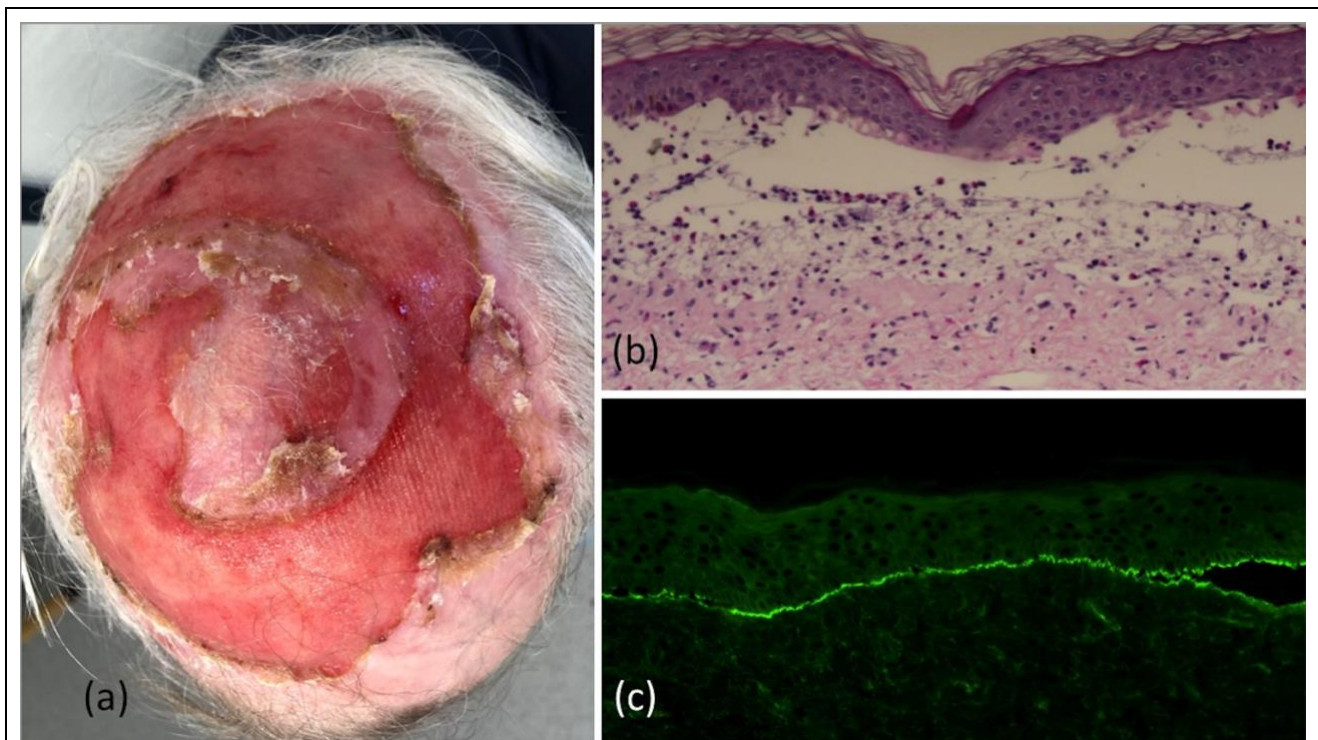


Figure 1: A patient with asymptomatic sharply demarcated crusty erosions and alopecia on the vertex. (A): Hematoxylin-eosin stain revealed intact epidermis and subepidermal blistering; (B): Direct immunofluorescence showed linear immunofluorescence with anti-IgG (n-serrated); (C): deposits along the basement membrane.

Clinical Image

A 74-year-old male with a history of actinic keratoses presented with a 1-year history of progressive asymptomatic crusty erosions and alopecia on the frontoparietal scalp region (Figure 1a). There was no history for frank pustulation or blistering. Clinical picture and a previous histology suggested erosive pustular dermatosis of the scalp (EPDS). However, new biopsies obtained from the erosion's margin including intact epidermis confirmed subepidermal blistering (Figure 1b) and linear immunofluorescence with anti-IgG (n-serrated, Figure 1c) and C3 deposits along the basement membrane. Anti-BP180 NC16A IgG was elevated in the patient's serum [37U/ml (<20)]. There was no evidence for mucosal involvement. Brunsting-Perry pemphigoid (BPP) was diagnosed. Treatment with systemic tapered corticosteroids and doxycycline 200mg/d resulted in gradual improvement of his condition.

EPDS is as rare condition most frequently affecting the vertex of the elderly with a history of actinic damage or trauma in the affected sites. Erosions, crusts, and scarring alopecia most commonly dominate the clinical picture of EPDS, whereas frank pustulation is infrequently observed. The histopathology of EPDS is unspecific. A variety of other skin conditions has to be considered as differential diagnosis, including non-melanoma skin cancer, infections, pemphigus, and dissecting folliculitis [1]. However, BPP represents a very rare differential diagnosis of EPDS. BPP is a rare pemphigoid variant clinically characterized by cicatrizing lesions predominantly on the head and neck. Non-predominant mucous membrane involvement may appear during the course of BPP, generally after skin manifestations. Positivity of direct immunofluorescence (DIF) and anti-BP180 NC16A/230 serum autoantibodies confirm diagnosis of BPP [2]. In the absence of frank blistering, it is difficult to differentiate clinically BPP from EPDS. Hence, DIF and autoantibody testing should always be performed when EPDS is considered in order to exclude bullous autoimmune dermatoses such as BPP.

Keywords: Bullous autoimmune disorders; Autoantibodies; Direct immunofluorescence; Bullous pemphigoid

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