



AUTOIMMUNE BULLOUS DISEASES

OLD MAN WITH HEAD AND NECK SCARRING

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Back ground: Brunsting–Perry pemphigoid is a clinical subtype of cicatricial pemphigoid. It is characterized by subepidermal blisters with erosions and hemorrhagic crusts which heal leading to atrophic scars. The lesions are characteristically confined to the head, neck and upper trunk. Middle-aged and elderly populations are commonly affected.

Observation: A 75 year old male presented with ulceration over the jaw area, under the chin and occipital scalp for 1 year duration. On examination there were multiple erythematous atrophic plaques on above distribution with foci of ulcerations. Patient couldn't recall any blistering episodes. All the biochemical investigations including ANA, VDRL and retroviral studies were negative. Initial biopsy revealed predominant plasma cell infiltrate in the superficial dermis leading to a diagnostic dilemma.

Three months later he presented with tensed blisters over the upper chest, face, neck and dorsum of the hand with ulceration in some. He continued to have atrophic scarring over the lower jaw area and under the chin. Repeat biopsy of a new lesion revealed subepidermal blister containing numerous eosinophils and scattered lymphocytes. The plasma cell rich infiltrate was not noted in this biopsy. Direct immunofluorescence from the perilesional skin showed linear deposits of IgA, IgG and C3 along the basement membrane. Salt split test and indirect immunofluorescence test could not be done due to unavailability. Patient responded well to oral and topical steroid treatment.

Key message: Though rare, Brunsting perry type cicatricial pemphigoid should be considered as a differential diagnosis when an elderly person presents with blisters and atrophic scarring, specially over the head neck and upper trunk area.

This case with typical skin lesions compatible with the above diagnosis is reported due to the rarity of the disease and as proper treatment can lead to good recovery.

