Our Dermatology Online

Case Letter

Pemphigus or pemphigoid dilemma: A case letter on this persisting confusion

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Sir,

From our residency days, we have been learning about the distinct clinical and histopathological features that differentiate pemphigus and pemphigoid. Pemphigus is characterized by fragile flaccid intraepidermal blisters while pemphigoid by tense intradermal blisters that are not easily ruptured. The differentiation between the two can be challenging even for an experienced physician especially when presented in the late stage with generalized skin and mucosal lesions. At times histopathology may be misleading due to epidermal regeneration in older lesions and spongiotic blisters in early lesions as in our case where the patient presented with overlapping clinical features and equivocal histopathology and final eye opener was as always immuno-fluorescence.

A lady (61y) presented with pruritic large tense blisters, predominantly over legs arms and trunk. Many lesions were crusted and erosions were non healing. Many lesions showed smaller new vesicles in border like string of beads. She had multiple painful oral erosions covered with whitish slough with bad odor Scalp face palms and soles were spared. No wheals were observed. Bulla spread sign was positive and Nikolskys sign was negative, making it difficult to differenciate between pemphigus and pemphigoid clinically. She is a known diabetic, hypertensive, hypothyroid on medication since past 10 years. Her routine blood urine and stool examinations were WNL. RFT and LFT and Peripheral smear were also normal. USG abdomen Xray chest did not reveal any abnormality except grade 1 fatty liver. Tzank smear showed few atypical acantholytic cells and mixed inflammatory infiltrates. Histopathology showed epidermal spongiosis with intraepidermal blister and few eosinophils (Fig. 1a). There were no sub-epidermal blisters. These equivocal findings posed more difficulty in arriving at final diagnosis. Direct immunofluorescence showed linear staining of basement membrane zone with Ig G and C3 Salt split skin showed a roof pattern, a finding consistent with bullous pemphigoid (Fig. 1b). Patient was started on Methotrexate injections 15 mg weekly, topical potent steroids and antibiotics. Her regular medications were continued. After 4 weeks there was not much improvement and was getting new lesions so later switched to systemic steroids with insulin for diabetes and oral hypoglycemics were stopped. Eventhough she was not on gliptins which is known to cause bullous lesions, possibility of newer nongliptins as cause of blister was also thought of as she was nonresponsive for initial treatment were oral hypolglecemics were continued with methotrexate. Patient responded well she was in complete remission in 4 weeks of steroid in tapering dose with addition of potent topical steroids. Later switched to oral hypoglycemics and she remained in remission ruling out possibility of non-gliptin oral hypoglycemics as cause of blister.

Pemphigus and Pemphigoid are two auto immune blistering diseases with distinct clinical histopathological and immunological features which makes it easy to differentiate if the presentations are typical [1]. Even treatment and prognosis are different for the two conditions Hence it is mandatory to differentiate between the two [2]. There may be overlapping clinical and histopathological features if either of disease is early or late. Early lesions of pemphigoid show eosinophilic spongiosis with epidermal blisters [3]. Late lesions may

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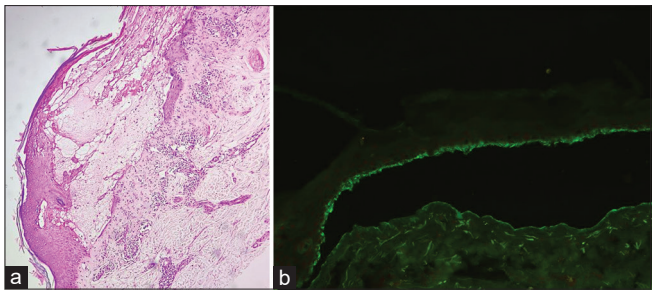

Figure 1: (a) Histopathololological examination vesicle x10 magnification showing intraepidermal blister with eosinophils. Dermis few lymphocytic infiltrates and no sub epidermal blisters. (b) Direct Immunofluorescence ofsalt split perilesional skin showing linear staining of Ig G and C3 at BMZ.

be misleading as to site of blister due to epidermal regeneration. Biopsy of well-formed pemphigoid showing epidermal blister also reported [4]. Our patient was on Metformin, Voglibose, Telmesartan, Amlodipine and Eltroxin. None of these were known to produce blistering rash [5]. Though our patient responded well after stopping voglibose and Metformin she did not relapse after restarting the same ruling out the possibility of new antidiabetic drug in the list for drug induced bullous pemphigoid.

This case highlights the importance of a thorough evaluation and early consideration of all diagnostic tools, particularly immunofluorescence for accurate diagnosis and timely intervention of auto-immune blistering diseases.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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