COEXISTENCE OF PEMPHIGUS VULGARIS AND VITILIGO VULGARIS IN A PATIENT OF CHRONIC LIVER DISEASE- A POINTER TOWARDS COMMON AUTOIMMUNE AETIOLOGY

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PRESENTATION OF CASE

A 56-year-old gentleman with stable vitiligo vulgaris for the past 20 years presented with flaccid vesicles and raw, painful erosions on various parts of body. These lesions erupted on and off for the past 2 years and healed with post-inflammatory hyperpigmentation. Current exacerbation of the symptoms and erosions in oral cavity was present for the past 1 month. Patient noticed that these fluid-filled lesions arose only on the pre-existing white patches on skin. There was no history suggestive of any underlying connective tissue disorder. He was on homeopathic treatment for vitiligo for the past 20 years and was also being treated for diabetes for the past 5 years. The patient was recently diagnosed with chronic liver disease (HCV related viral load of 3,41,700 IU/mL) along with portal hypertension with grade III oesophageal varices. He was on ribavirin for the same for the past 2 months.

On examination, well-defined depigmented macules of varying sizes were seen on face, bilateral ears, lower lip, neck, trunk and bilateral palms. Some of the macules showed perilesional pigmentation, but no leukotrichia was present. Crusted plaques were noticed on many of these vitiliginous lesions on the photoexposed parts. In the subsequent visits, the patient developed vesicles, flaccid bullae, few turbid bullae and crusted plaques on vitiliginous lesions as well as normal skin over the face and the body predominantly on lower lip, bilateral external auditory canal, back, upper arms, chest and abdomen (Figure - 1,2). Perilesional Nikolsky's sign was positive, but distant (direct) Nikolsky's was negative. Oral cavity revealed superficial erosions in bilateral buccal mucosa. Gingivae and vermilion border of lip were not involved.

Haematological and biochemical investigations were normal, but for the liver function tests, which were deranged.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of pemphigus vulgaris, phototoxic reaction, paraneoplastic pemphigus and bullous lupus erythematosus were considered.

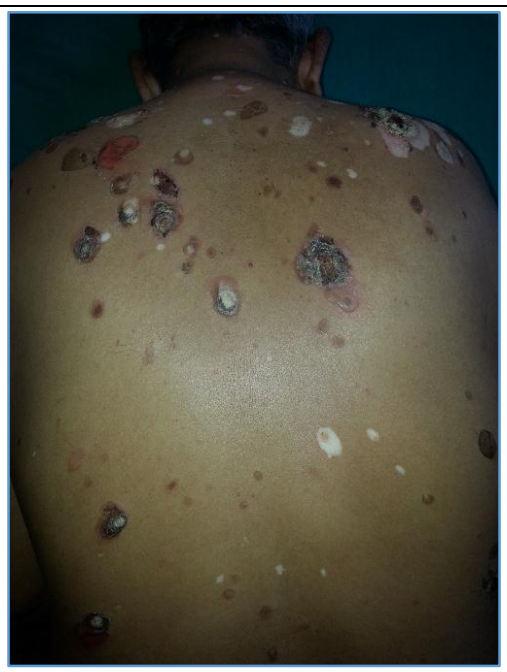

Figure 1. Depigmented Macules and Crusted Plaques (Both on Depigmented Macules and Normal Skin) Over the Back. A Flaccid Bulla is also Present

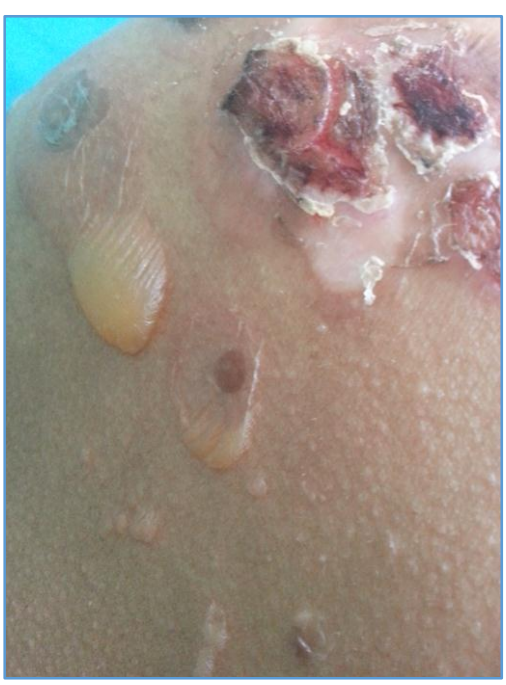

Figure 2. Flaccid Bulla Over Normal Skin and Crusted Plaque Over Depigmented Macule

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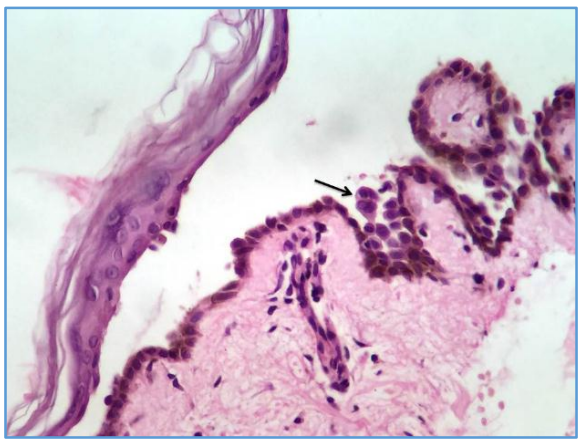

Figure 3. Suprabasal Separation of Epidermis with Few Acantholytic Cells (Arrow) and Occasional Erythrocytes. H and E, 400x

CLINICAL DIAGNOSIS

A clinical diagnosis of pemphigus vulgaris was made after reviewing the skin biopsy report and immunofluorescence report.

PATHOLOGICAL DISCUSSION

Skin biopsy sent from a newly-erupted vesicle for Histopathological Examination (HPE) showed thinned out epidermis with a large blister cavity and suprabasal separation with numerous acantholytic cells.

The blister cavity contained occasional pus cells and Red Blood Cells (RBCs) and mild perivascular lymphocytic infiltrate in the underlying dermis (Figure 3). Direct Immunofluorescence (DIF) was positive (3+) for IgG and IgM (1+), which were deposited on the surface of keratinocytes in and around the lesions. It was negative for IgA, C3, C1q. These findings were consistent with pemphigus vulgaris. Hence, a diagnosis of vitiligo with pemphigus was made.

The mechanism of occurrence of pemphigus vulgaris in a case of vitiligo vulgaris in a patient of hepatitis-C induced chronic liver disease is not known, but a common auto-immune aetiology might be responsible. Human Leucocyte Antigen (HLA) DR4-DQ1 haplotype predisposes to both vitiligo and pemphigus.[1,2] Also, in vitiligo, due to decreased levels of acetylcholine esterase, Acetylcholine (ACh) activity is raised. Cholinergic receptors regulate adhesion between keratinocytes. Due to persistently high levels of Ach, there is possible down regulation of cholinergic receptors, which may result in loss of adhesion between keratinocytes leading to pemphigus. This can explain the development of pemphigus lesions on vitiliginous lesions.[1,2] Tumour Necrosis Factor-α (TNF-α) has been found to be a common mediator for both vitiligo and pemphigus.[1,2] All these hypotheses may explain development of pemphigus in our patient of vitiligo vulgaris.

Case reports of development of pemphigus vulgaris in a patient of vitiligo and Hashimoto's thyroiditis, colocalisation of mucosal vitiligo and oral pemphigus vulgaris and association of alopecia areata, vitiligo and pemphigus vulgaris have been previously reported.[1,2,3,4] But, coexistence of vitiligo, pemphigus vulgaris and hepatitis-C induced chronic liver disease in the same patient is extremely rare and cannot be a mere coincidence. This case further strengthens the common autoimmune hypothesis.

DISCUSSION OF MANAGEMENT

Patient was started on oral steroids (prednisolone - 1 mg/kilogram body weight/day). The patient showed good initial response to prednisolone, but was lost to follow up.

FINAL DIAGNOSIS

A diagnosis of vitiligo with pemphigus vulgaris was made.

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