Co-existence of Pemphigus Foliaceous and Psoriasis: Is There a Common Pathogenetic Link?

Dear Editor,

There are case reports of co-existence of psoriasis and autoimmune bullous disorders. Most reports of autoimmune bullous disease associated with psoriasis are that of bullous pemphigoid.[1] Few reports of coexistent psoriasis and pemphigus foliaceous are there.[2-5] On literature search, we could not find any such reports from India. Herein we report a case of coexisting psoriasis and pemphigus foliaceous.

A 47-year-old man presented to us with flaccid vesicles and crusted erosions over face and upper trunk for one month. He was a known case of chronic plaque psoriasis for last three years with a remitting and relapsing course. He was taking weekly methotrexate 7.5 mg off and on during disease flare up. For last 1 month he had developed fluid filled lesions over face and upper trunk that soon evolved into crusted erosions. Patient denied history of drug intake for any other chronic illness. General, physical and systemic examinations were within normal limits. Dermatological examination revealed flaccid vesicles and bullae with a thin rim of erythema present over trunk. Secondary lesions were in the form of erosions with adherent superficial crusting over chest and face [Figure 1]. Hyperkeratotic plaques with adherent white scales consistent with psoriasis were present over bilateral elbows and shins [Figure 2]. There was no mucosal involvement. Complete blood count and routine biochemical parameters were within normal range. Skin biopsy from the erosive lesions showed acantholysis of the upper epidermis with moderate perivascular infiltrate of lymphocytes and eosinophils [Figure 3]. The direct immunofluorescence demonstrated intercellular deposition of complement (C3) and IgG in the superficial layers of epidermis [Figure 4]. Histopathology study of scaly plaques revealed parakeratotic hyperkeratosis with regular elongation of rete ridges with perivascular lymphocytic infiltration suggestive of psoriasis [Figure 5]. On the basis of clinical features and histopathological finding the scaly plaques were diagnosed as plaque psoriasis and bullous lesions as pemphigus foliaceous. Considering presence of both the diseases, patient was started on methotrexate 7.5 mg weekly once and mycophenolate mofetil 2000 mg daily orally in divided dose. As bullous lesions continued to appear with this treatment, patient was given intravenous dexamethasone 8 mg per day for 7 days and lesions responded. Patient was discharged with 30 mg prednisolone

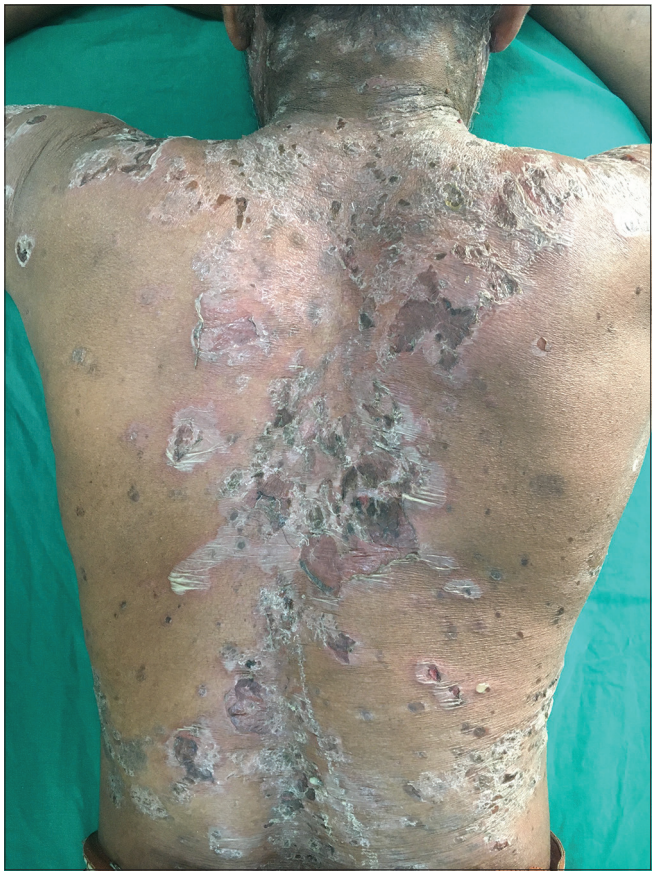

Figure 1: Flaccid vesicle and bullae with crusted erosion over trunk

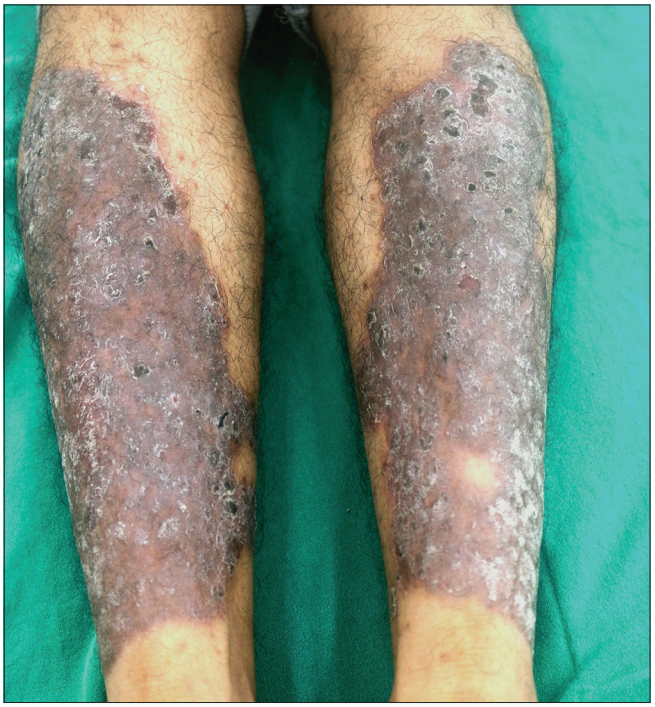

Figure 2: Well defined erythematous to hyperpigmented indurated scaly plaques over bilateral shins

Table 1: Summary of recent case reports of psoriasis and pemphigus foliaceous

Authors	Year of publication	Age (years)/ gender	Time lapse between psoriasis & pemphigus foliaceous	Treatment given
Giomi *et al*.[3]	2004	77/female	7 years	Methyl prednisolone
Kurtzman *et al*.[4]	2015	62/female	Several months	Systemic corticosteroid, azathioprine
Sanz-Bueno *et al*.[5]	2017	60/male	Not available	Adalimumab
Zhang *et al*.[2]	2018	79/male	4 years	Topical steroid , topical tacrolimus
	2018	74/male	30 years	Topical steroid, Systemic methotrexate
	2018	54/male	5 years	Systemic prednisone, methotrexate
	2018	58/female	30 years	Systemic prednisone, cyclosporine
Our study	2019	47/male	3 years	Systemic prednisolone, methotrexate, mycophenolate mofetil

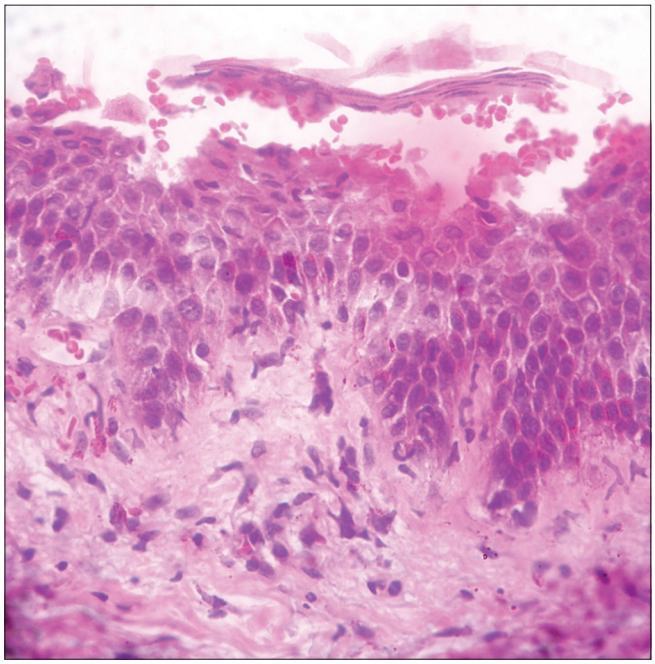

Figure 3: Intra-epidermal cleft with loss of superficial epidermis with acantholytic cells (H & E, 40×)

along with methotrexate 7.5 mg weekly once and mycophenolate mofetil 2000 mg daily. Prednisolone was gradually tapered and stopped subsequently over a period of two months with continuation of rest two medications. Patient did not show any flare up of psoriasis after stoppage of prednisolone and both the diseases were well controlled till last follow up.

A summary of the previously reported cases is given in Table 1.

Psoriasis is considered as a T lymphocyte mediated chronic inflammatory disorder with abnormal cytokine and chemokine profile causing excessive keratinocyte proliferation. In contrast, pemphigus is a disorder of humoral immune system where antibodies are produced against desmogleins that are expressed in skin and mucosa. Psoriasis being a common dermatosis, coexistence of both the diseases could be a chance association. Considering greater prevalence of psoriasis in pemphigus compared to controls as observed in a large scale retrospective

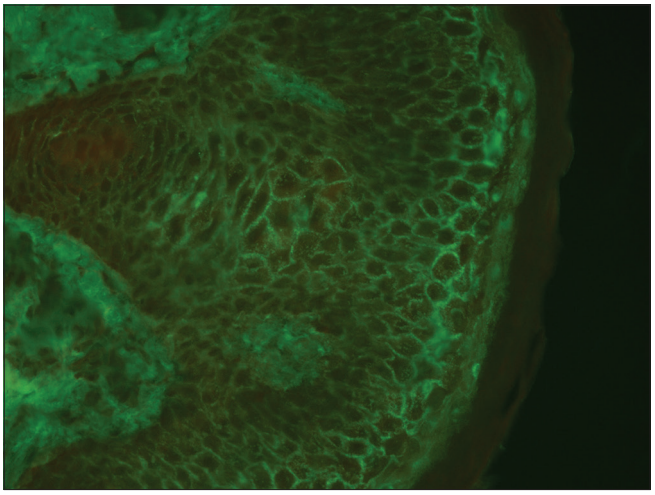

Figure 4: Direct immune fluorescence of perilesional skin shows intercellular deposits of Ig G (40×)

study[6] and the previously reported cases of simultaneous occurrence of both diseases, following hypothesis can be postulated for their co-occurrence.

1. Plasminogen activation hypothesis: plasmionogen activation system has been postulated to be responsible for acantholysis in pemphigus. It has been seen that psoriatic skin lesions have an elevated level of tissue plasminogen activator[3]
2. Common HLA antigen: Possible role of human leucocyte antigen HLA DRB1 alleles has been reported in many studies, both in psoriasis as well as pemphigus foliaceous. So a common HLA genotype and abnormal T lymphocyte activation can produce lesions of both psoriasis and pemphigus[4]
3. Decreased suppressor T lymphocyte function: Some studies have reported reduced suppressor T cell function in psoriasis that leads to increased activity of humoral immune system and production of antibodies[7]
4. Pathogenesis of psoriasis involves both auto-inflammation and autoimmunity. Research has established links between auto-inflammatory diseases and disorders of autoimmunity. Raised IL 1 β in the former can cause activation of adaptive immunity, thus triggering autoimmunity[8]
5. Role of NLRP 1 inflammosome: Variations in NLRP 1 gene are associated with increased susceptibility for psoriasis as well as pemphigus foliaceous.[9,10]This gene

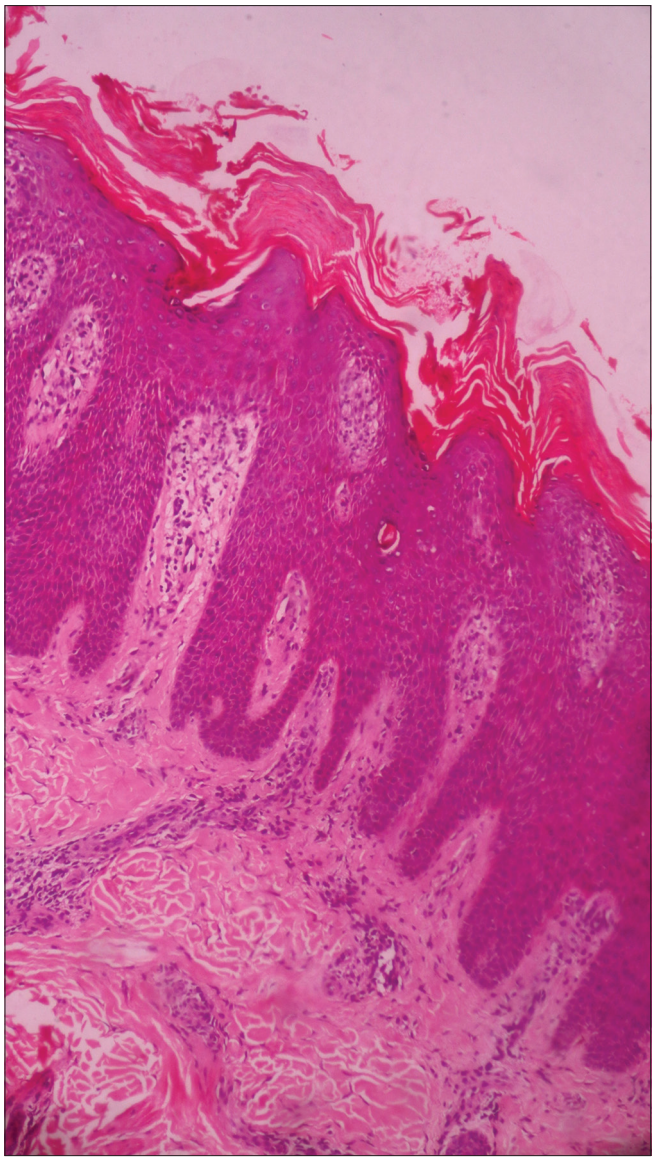

Figure 5: Regular elongation of rete ridges with para keratosis suggestive of psoriasis (H & E, 10×)

encoding proteins of the inflammosome could be a common link in the pathogenesis of both the entities.

Systemic corticosteroids are contraindicated in plaque psoriasis, on the contrary they are the first line of treatment for pemphigus. Hence, coexistence of both the dermatoses as in the index case poses a challenge in management. Addition of a steroid sparing agent along with gradual tapering of systemic corticosteroid will be of help. Further molecular studies in both the diseases are required to confirm any common pathogenetic link.

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Conflicts of interest

There are no conflicts of interest.

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