Uncommon and Unusual Variants of Autoimmune Bullous Diseases

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Abstract

Background: Autoimmune blistering diseases (AIBDs) are a type of dermatosis with antibodies produced against various structural proteins of the epidermis or dermoepidermal junction. AIBDs are broadly divided into intraepidermal and subepidermal types. Apart from the common AIBDs, there is an array of uncommon AIBDs. **Objective:** To discuss uncommon variants of AIBDs so that the readers are updated about them. **Methods:** In this review, we have discussed uncommon and unusual variants like pemphigus herpetiformis, IgA pemphigus, paraneoplastic pemphigus, induced pemphigus, IgG/IgA pemphigus, oral lichenoid pigmentation in pemphigus, pemphigus acanthoma, and follicular pemphigus. Rarer variants of the pemphigoid group of disorders include anti-laminin 332 pemphigoid, mixed linear IgA/IgG pemphigoid, anti-p200 pemphigoid, Brunsting-Perry pemphigoid, IgM pemphigoid, granular C3 pemphigoid, anti-p105 pemphigoid, ORF-induced anti-laminin 332 pemphigoid, and acral purpura in dermatitis herpetiformis. **Conclusion:** This review will help in early diagnosis and treatment of uncommon and unusual variants of AIBDs.

Keywords: *Anti-laminin 332 pemphigoid, granular C3 pemphigoid, IgA pemphigus, pemphigus herpetiformis*

Introduction

Autoimmune bullous diseases (AIBDs) are broadly classified into intraepidermal (IEBD) and subepidermal AIBDs (sAIBD), based on the plane of cleavage within the skin or mucosae. Pemphigus vulgaris (PV) and pemphigus foliaceous (PF) are considered the prototypes for IEBD, whereas bullous pemphigoid (BP) is considered the prototype for sAIBDs.

Occasionally, clinicians may encounter uncommon forms of pemphigus and pemphigoid group of diseases. In this section, we have reviewed the various nonclassical forms of pemphigus and pemphigoid.

Uncommon Variants of IEBDs

Pemphigus herpetiformis (PH)

PH comprises nearly 6-7% of total pemphigus cases with a mean age range from 31 to 83 years and occasionally can occur during childhood.[1,2] The hallmark of PH is erythematous, urticarial papules and plaques with a few vesicles in a herpetiform pattern closely resembling Dermatitis Herpatiformis (DH) seen over torso and limbs.[3] Pruritus and mucus membrane involvement have been documented in 86% and less than 10% of cases, respectively.[4] Rarely, PH may transform into PV or PF and vice versa.[5-7]

Histopathological examination reveals eosinophilic and/or neutrophilic spongiosis, subcorneal pustules, intraepidermal split, and minimal acantholysis.[4] Direct and indirect immunofluorescence (DIF and IIF) will exhibit intercellular staining (ICS) of the epidermis with IgG and C3, with more prominent staining in the upper epidermis, a finding similar to PF.[8,9] Circulating antibodies desmoglein 1 (Dsg-1) and less commonly Dsg-3 have been detected in PH; rarely anti-desmocollin (Dsc) 1 and 3 and an unidentified 178-kDa protein.[1] The diagnostic criterion for PH has been shown in Table 1.[4]

PH usually responds well to dapsone monotherapy (100-300 mg daily).[1,9] Systemic corticosteroids may be added in unresponsive cases. Few patients may transit to classical forms of pemphigus, wherein they may require additional immunosuppressive drugs (azathioprine, cyclophosphamide, and mycophenolate mofetil) to induce clinical remission.

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Intravenous venous immunoglobulin (IVIG) and plasmapheresis have also been useful in resistant cases.[4,8-10] Rituximab has proven efficacious in a few cases.[11]

IgA pemphigus

IgA pemphigus is an uncommon form of pemphigus with an average age of disease of 51.5 ± 21.0 years with slightly more prevalence among females, and few cases have been reported among children.[12,13] There are two phenotypic types of IgA pemphigus—the subcorneal pustular dermatosis (SPD) type and the intraepidermal neutrophilic (IEN) type with the SPD type being more common than the IEN type (44.7% vs 39.8%).

It is characterized clinically by the presence of flaccid vesicles or pustules on normal skin or on an erythematous base, seen over flexural folds of the groin and axilla; torso and limbs can also be affected. The pustules may merge to form a circinate or annular lesion with a characteristic "sunflower-like" appearance[14] [Figure 1]. Mucosal lesions occur in about 13.2% of patients.[12]

An intraepidermal split may be detected at either subcorneal or intraepidermal levels based on the clinical phenotype. The blister cavity shows neutrophilic infiltration with less degree of acantholysis [Figure 2]. IgA staining in the intercellular spaces of the epidermis is seen in DIF and IIF [Figure 3].[15] Dsc-1 is the target antigen in the majority of IgA pemphigus of SPD type; the exact target antigen in the IEN type is not yet known, although few cases have shown IgA antibodies to Dsg-1 or Dsg-3.[16-18]

IgA pemphigus has been associated with hematologic malignancies, solid organ malignancies, psoriasis, ulcerative colitis and neurological disorders.[15,19-21] IgA pemphigus has also been reported after COVID-19 vaccination.[22]

There is a reported case of pemphigus foliaceus mimicking clinical features of IgA pemphigus, which showed no response to steroids, and azathioprine, however, had improved with dapsone.[23]

The mainstay of treatment is dapsone; oral and topical corticosteroids may be added in resistant cases.[1,15] Other agents have been tried including retinoids, mycophenolate mofetil, adalimumab, azathioprine, cyclophosphamide, plasmapheresis, and colchicine.[24-27] Successful results have also been achieved using NB-UVB.[28]

Paraneoplastic pemphigus (PNP)

Anhalt *et al.*[29] published the first reports of paraneoplastic pemphigus (PNP) in association with lymphoproliferative diseases. Paraneoplastic autoimmune multiorgan syndrome (PAMS) was later introduced, highlighting the systemic involvement in PNP. This concept describes that PNP is not just a dermatological disease but rather a syndrome

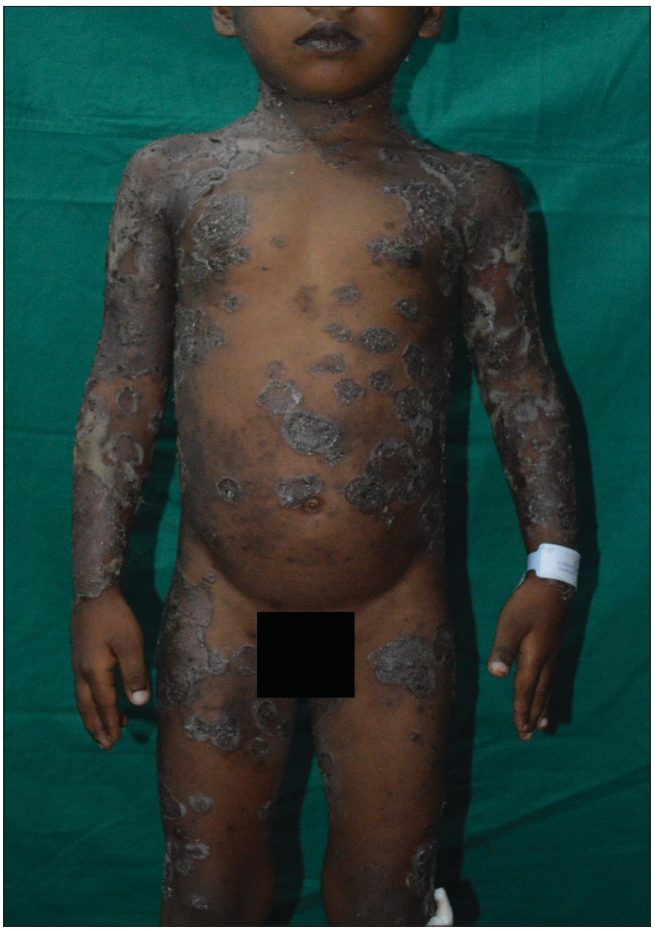

Figure 1: Multiple annular and circinate vesiculopustules over trunk in a juvenile IgA pemphigus patient

Table 1: Suggested criteria for diagnosis of Pemphigus Herpetiformis[a,4]

Clinical	Pathologic	Immunologic[c]
Itchy herpetiform skin blisters in the presence or absence of erosions.	Intraepidermal eosinophils and/or neutrophils[b]	DIF demonstrating intercellular staining with IgG +/–C3
Itchy urticarial or annular erythematous plaques in the presence or absence of erosions.	Intraepidermal split with/ without acantholysis	IIF demonstrating intercellular staining with IgG
		Demonstrating antidesmogleins (1,3) or antidesmocollins (1,2,3) or both in serum

[a]Diagnosis requires one clinical, one pathologic, and one immunologic feature. [b]Eosinophilic/neutrophilic spongiosis or pustules/ vesicles/blisters containing eosinophils or neutrophils. [c]No immunologic findings characteristic of DH, BP, or PNP. Adopted from: Costa LMC,Cappel MA, and Keeling JH. Clinical, pathologic, and immunologic features of pemphigus herpetiformis: a literature review and proposed diagnostic criteria, International Journal of Dermatology, 2019;58: 997-1007

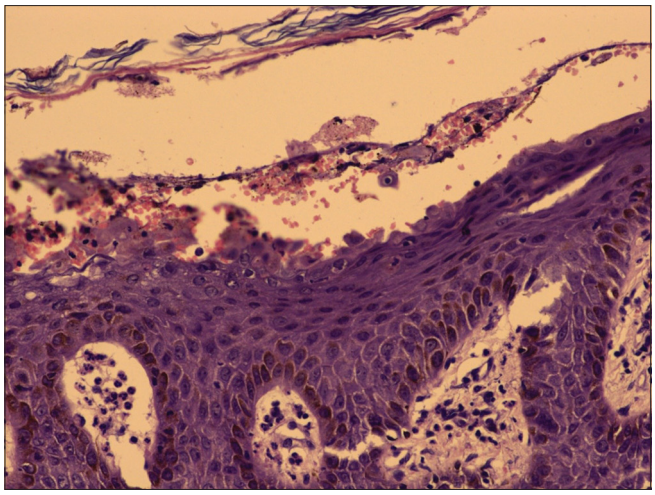

Figure 2: Histopathology of IgA pemphigus (SPD type) showing subcorneal split with neutrophilic infiltration (hematoxylin and eosin staining, x200)

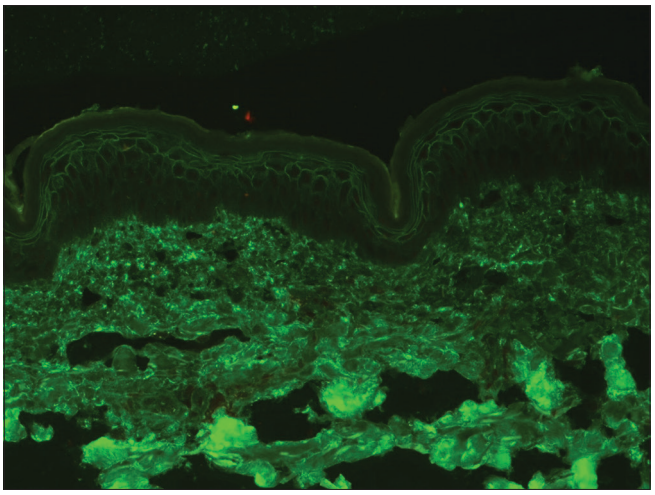

Figure 3: DIF image showing intercellular staining with IgA in IgA pemphigus (FITC, x200)

marked by both skin and systemic pathology linked with neoplasia.[30] The prevalence of PNP is around 3–5% of all pemphigus cases and seen commonly between 45 and 70 years.[31,32]

The plakins are the main target antigens in PNP with envoplakin, periplakin, and desmoplakin I and II commonly affected.[33] Less commonly, autoantibodies also develop against BP230, plectin, epiplakin, BP180, *P*-200 protein, Dsg1, Dsg3, Dsc1, Dsc2, Dsc3, and Alpha-2-Macroglobulin-like 1 (A2ML1) and transglutaminase 1.[34-37]

The early signs of PNP include painful, progressive, recalcitrant stomatitis characterized by erosions and ulcerations affecting the oropharynx and extending to the vermilion border of the lips and the tongue [Figure 4].[38,39] In addition, mucositis can also involve the pharynx, larynx, esophagus, eye, and anogenital region.[31,33]

Cutaneous lesions in PNP are extremely polymorphous. These include a) pemphigus-like [Figures 5 and 6], b) bullous pemphigoid-like, c) erythema multiforme (EM)-like, d) graft-versus-host disease (GVHD)-like, and e) lichen planus–like.[37] Atypical cases of PNP without mucosal involvement have also been described.[40,41]

Bronchiolitis obliterans can be one of the complications of PNP, leading to death in these cases.[42,43]

The various PNP associated neoplasia are lymphoproliferative malignancies, hematological malignancies, Castleman disease, thymomas, and Waldenstrom's macroglobulinemia.[37,44]

The histopathologic findings depend on the clinical presentation and include acantholysis, suprabasal split, dyskeratotic keratinocytes, vacuolation of basilar cells of the epidermis, and inflammatory cell exocytosis.[45] DIF and IIF usually show intercellular IgG and/or C3 deposits. In addition, BMZ IgG and/or C3 deposits are also reported in about 50% of cases.[31] Rat bladder is considered the optimum substrate for IIF; it is more sensitive than normal human skin (75% vs 50%).[31,46]

Serum autoantibodies are detected using ELISA, immunoprecipitation (IP), and immunoblotting (IB).[31] Recently, the BIOCHIP mosaic technique and multivariant ELISA containing envoplakin have been found to be a sensitive serological tests.[47] Various authors have proposed diagnostic criteria for PNP over the period of time.[29,48,49] The recent one by Antiga et al. has been shown in Table 2.[37]

Systemic corticosteroids are the first line treatment of PNP. Prednisolone in the dose of 0.5–1.5 mg/kg/day is a commonly used drug, often in conjunction with steroid-sparing agents such as azathioprine, mycophenolate mofetil, cyclosporine, cyclophosphamide, and methotrexate. In addition, intravenous immunoglobulins (IVIG) has also been explored as a modality.[33,37] Rituximab has given good results in PNP secondary to B-cell malignancies.[33,50]

Alemtuzumab, an anti-CD52 drug, daclizumab, a monoclonal antibody against IL-2 receptor of T cells, plasmapheresis, and thalidomide have also been tried.[31,33,37]

Drug-induced pemphigus

The pathomechanism of induced pemphigus is not described clearly. The proposed processes includes inhibiting keratinocyte aggregation by enzyme inhibition, keratinocyte disaggregation by activating plasminogen, thiol-cysteine bond formation thereby inhibiting cell adhesion, interleukin-1 alpha (IL-1α), and tumor necrosis factor-alpha (TNF-α) release to cause acantholysis in the absence of an autoimmune response,[51,52] and immunological alterations, resulting in new antigens.[53]

The various chemicals implicated to cause pemphigus are as follows.

a) Sulfhydryl (-SH) group-thiols or mercaptans:penicillamine, captopril, pyritinol, piroxicam, gold sodium thiomalate. b)

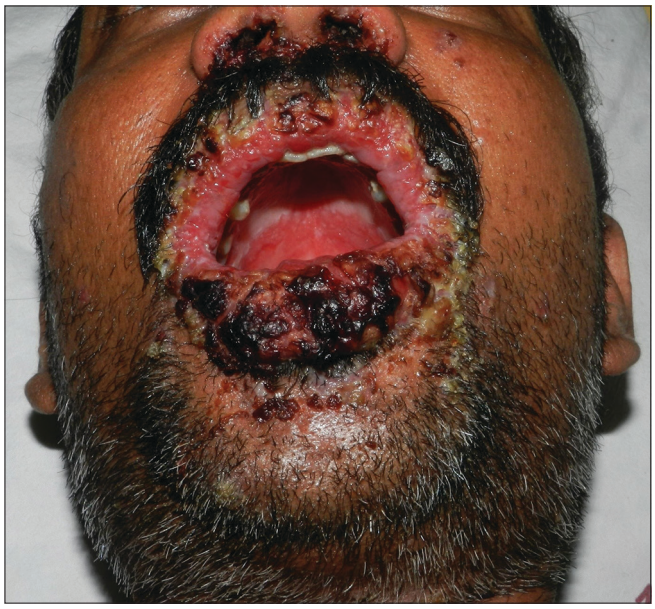

Figure 4: Recalcitrant mucositis in PNP

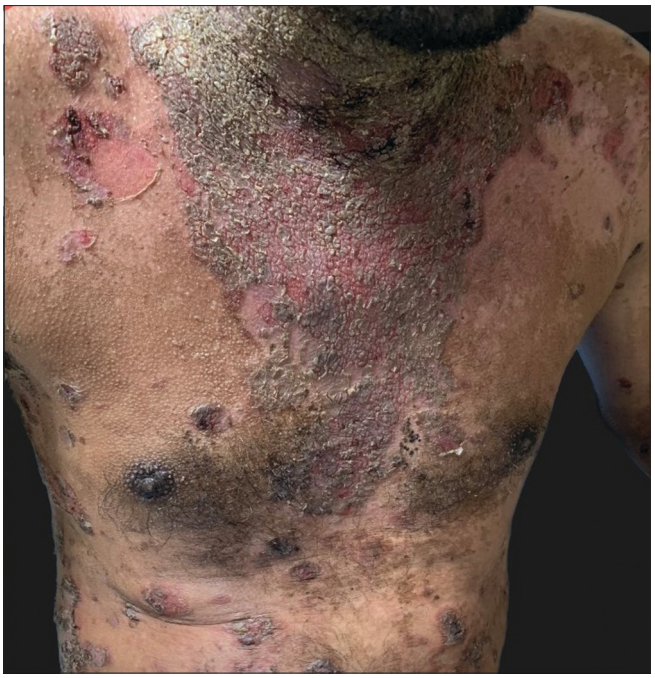

Figure 5: Erosions with crusting over the trunk in PNP

Amide group:Penicillins and carbamazepine. c)Phenolic group:Aspirin, levodopa, heroin and rifampin. d)Non-thiol non-amide non-phenolic group:Nonsteroidal anti-inflammatory drugs, calcium channel blockers, angiotensin-converting enzyme inhibitors, glibenclamide, and dipyrone.[53,54]

Along with the systemic drugs, there are also a few topical agents with the capability to trigger or cause pemphigus, a phenomenon, known as contact pemphigus.[55]

The commonest type of drug induced pemphigus is PV (38.9%), followed by PF (33.5%). The thiol group of drugs usually induces PF, whereas nonthiols usually induce PV.[51] Histologic features are the same as that of classical pemphigus. Histopathology cannot differentiate these two entities. DIF showed ICS with IgG and C3 in 93% and 83.8% of patients, respectively. IIF showed positive results in 67.3% and 68.3%, respectively.[51] ELISA may be performed to detect anti-Dsg1 and/or Dsg3 antibodies. For identifying drug-induced pemphigus Dsg immunolabeling, interferon-gamma release test or lymphocyte transformation test can be used.[56] The first line of therapy includes combining corticosteroids and an immune suppressive drug similar to the ones used in classical pemphigus along with stopping the culprit drug.[54]

IgG/IgA pemphigus

The exact nosological position of this rare entity is unclear at present. It may present as classical pemphigus (PV or PF) with vesicles, bullae, or erosions. On the other hand, arciform or annular erythema with erosions or blisters (PH or IgA pemphigus) have been reported. Mucosal involvement is less commonly seen, unlike classical PV.[57,58] Histopathological examination will reveal acantholysis (suprabasal, subcorneal, or intraepidermal) and neutrophils and eosinophils aggregation in the epidermis.[58] DIF and IIF will have intercellular staining of keratinocytes with both IgA and IgG. ELISA has also demonstrated IgG and IgA antibodies against Dsg 1 and 3.[59]

Oral lichenoid pigmentation in pemphigus

Lichen planus (LP)-like pigmentation after healing of oral pemphigus lesions has been documented in the literature. De *et al.*[60] studied 32 pemphigus patients with lichen planus–like oral pigmentation, irrespective of disease activity and treatment status. Histopathology was consistent with the diagnosis of pemphigus in 75% of cases, and DIF revealed ICS in 77.8% of cases. In addition, they also noted the higher rates of apoptosis markers such as Fas L and caspases 3 in these cases indicating the role of apoptosis. They concluded that oral LP-like lesions are a sign of active disease. However, Kianfar *et al.*[61] considered LP-like pigmentation as the post remission phase of pemphigus.

Verrucous pemphigus/pemphigus acanthomata/ postpemphigus acanthoma

Occasionally, pemphigus lesions may heal with seborrheic keratosis (SK) or verrucous lesions, which have been known as "postpemphigus acanthoma." Histopathological examination of these lesions showed intraepidermal cleft and acantholysis, in addition to hyperkeratosis, acanthosis, and papillomatosis; DIF of perilesional skin showed ICS, indicating that these lesions represent the active stage of pemphigus.[62] These lesions are also believed to be recalcitrant in nature.[63,64] Hence, post pemphigus acanthomas should be considered a potential indicator of future clinical relapse. In such cases, treatment should be titrated accordingly and close monitoring is advised.

Table 2: Criteria for diagnosis of PNP/PAMS suggested by Antiga E *et al.*[37]

Clinical criteria:

1. Chronic erosive mucositis
2. Polymorphic skin lesions including flaccid or tense blisters, erythema multiforme-like lesions, and lichenoid dermatitis.
3. Associated neoplasm comprising most frequently, but not exclusively, a lymphoproliferative or hematologic malignancy

Laboratory criteria:

Major

1. Epithelial cell membrane (and/or) staining by IIF using rat bladder
2. ELISA/immunoblotting/immunoprecipitation detection of antienvoplakin, antiperiplakin, antidesmoplakin, or anti-A2ML1 antibodies.

Minor

1. Cutaneous histopathology showing interface lichenoid dermatitis and/or acantholysis and/or necrosis of keratinocytes.
2. DIF and/or IIF showing ICS and/or linear/granular BMZ staining with IgG and C3
3. Detection of anti-Dsg antibodies and at least one of the following: ELISA/immunoblotting/immunoprecipitation detection of antidesmocollin, antiepiplakin, anti-BP180, or anti-BP230

Diagnosis of PNP/PAMS is confirmed: 2 clinical criteria + 1 major laboratory criterion or 2 clinical criteria + 2 minor laboratory criteria.

Diagnosis of PNP/PAMS is possible: 2 clinical criteria + 1 minor laboratory criterion t

If neoplasm is absent, in addition to the other 2 clinical criteria, 2 major laboratory criteria, or 1 major laboratory criterion and 2 minor laboratory criteria need to be present to make a provisional diagnosis of PNP/PAMS, and monitoring is recommended to exclude a possible occult tumor

Adopted from: Antiga e *et al*. S2k guidelines on the management of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome initiated by the European Academy of Dermatology and Venereology (EADV). J Eur Acad Dermatol Venereol. 2023;37:1118-1134

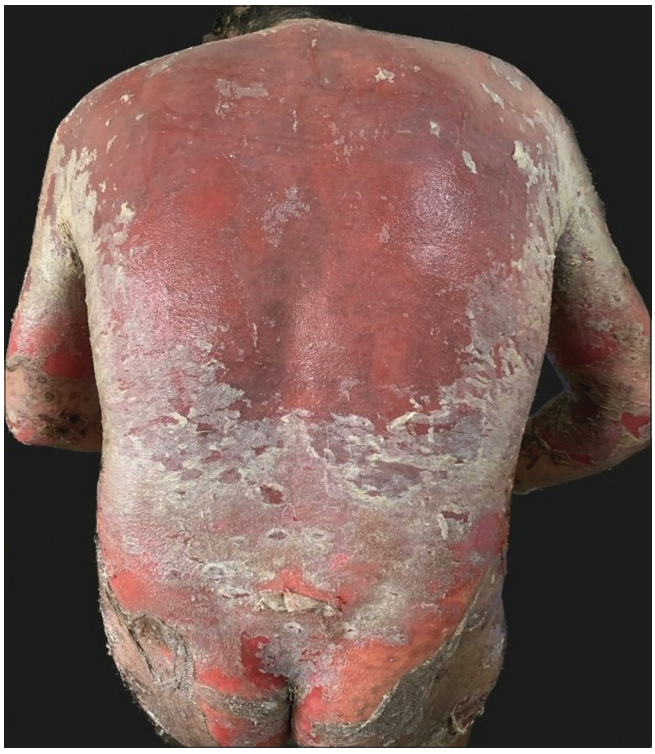

Figure 6: Extensive erosions over the back in PNP

Follicular pemphigus

Rarely, pemphigus may present with folliculocentric papulovesicles and pustules. This entity has been reported in three proven cases of pemphigus who were on oral steroids. DIF showed intercellular staining with IgG of the outer root sheath. The authors believed that this could be because of disease flare, which resulted in Koebnerization. This presentation was akin to acneiform eruption; hence, the patients were treated with doxycycline and topical benzoyl peroxide in addition to rituximab.[65]

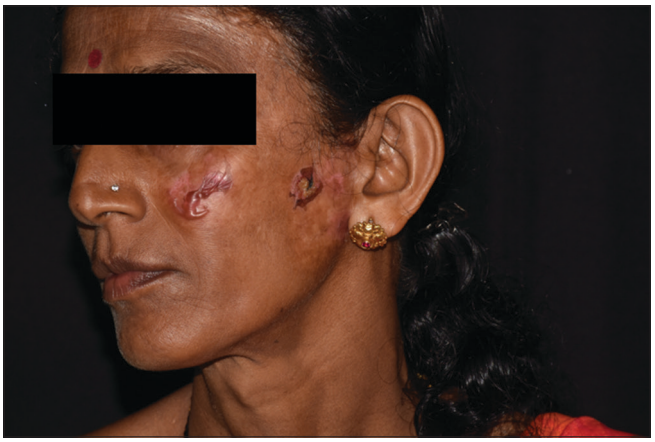

Figure 7: Bullae over the face in a patient of Brunsting-Perry pemphigoid

Dyshidrosiform pemphigus

Dyshidrosiform pemphigus presents with vesiculobullous lesions usually affecting the soles. It may later evolve into classical pemphigus with lesions elsewhere in the body. Local trauma has been proposed to be a triggering factor.[66-68]

Uncommon Variants of Subepidermal AIBDs

Anti-laminin 332 pemphigoid

Anti-laminin 332 (LAM 332) pemphigoid is a type of mucus membrane pemphigoid (MMP), in which antibodies are directed against laminin 332. It accounts for 20% of all MMP cases. Laminin 332 has 3 subunits LMα3, LMβ3, and LMγ2 with a molecular weight of 460 kDa. LAM 332 forms a primary structural element of anchoring filaments, thereby connecting lamina lucida with lamina densa.[69]

Clinically, these patients resemble classical MMP with erosions and vesiculobullous lesions affecting oral, ocular, laryngeal, pharyngeal, nasal, esophageal, and genital mucosae. In comparison with other types of MMP, LAM 332 pemphigoid may behave aggressively.[70]

DIF shows linear BMZ deposition of IgG and/or C3 and sometimes IgA with an "n" serration pattern. IIF on Salt-Split Skin (SSS) shows dermal pattern and helps to tilt the diagnosis in favor of anti-LAM 332 pemphigoid. However, circulating antibodies are present in low titers and may be detectable by IIF in only 50% of patients. Other tests that help to detect antibodies to LAM 332 include immunoblotting, ELISA, immunoprecipitation, biochip assay, and footprint assay.[71] Terra *et al*.[72] suggested criteria for diagnosis of anti-LAM 332 pemphigoid. This phenotype of MMP is associated with a high risk of internal malignancy (one in four patients), and effort should be made to detect antibodies to LAM 332 in patients with MMP.[73] Therapy for anti-LAM 332 MMP is similar to other forms of MMP and often requires a multidisciplinary approach by a dermatologist, ophthalmologist, and oncologist if there is underlying neoplasia. Medical management includes oral steroids, cyclophosphamide, azathioprine, mycophenolate mofetil, dapsone, rituximab, and IVIG. The responsiveness to drugs is variable in various reports, and the duration of therapy is prolonged to achieve remission.[74]

Mixed linear IgA/IgG bullous dermatoses

Few cases diagnosed clinically as linear IgA disease (LAD) have shown linear staining of IgG in addition to IgA both by DIF and IIF. These cases have been labeled as mixed linear IgA/IgG bullous dermatoses that demonstrate linear BMZ staining with IgA and IgG with roof/combined/floor staining pattern on IIF with SSS depending on the antigen involved and histopathology showing subepidermal split with neutrophilic and eosinophilic infiltrate.[75-77]

Sakaguchi *et al*.[78] also reported 3 patients of linear IgA/IgG bullous dermatoses wherein the immunoblot analysis showed IgA and IgG predominantly against laminin 332, in addition to type VII collagen, laminin-γ1, and BP230 and BP180 recombinant proteins.

In a study involving 213 cases of LAD, 45 (21.12%) predominantly adult patients showed linear BMZ staining with IgG in addition to IgA.[79] The treatment is similar to LAD and includes dapsone, topical and systemic steroids, MMF, azathioprine, IVIG, and rituximab.[76,80]

Anti-p200 pemphigoid

Anti-p200 pemphigoid is a novel sAIBD described by Zillikens *et al.* in 1996. Since then, various cases have been documented across the world including India. Once considered a rare condition, it is now believed that it is the common cause of dermal binding sAIBD.[81]

It is marked by the formation of antibodies against the 200-kDa protein present in the lower part of lamina lucida. Most of the patients (>90%) also show reactivity to the C-terminal of laminin γ1; hence, it is also known as laminin γ1 pemphigoid.

The affected patients are younger than those affected with BP, with the average age of onset at 65.5 years. It is indistinguishable clinically from other sAIBDs such as BP and LAD. Affected patients present with tense blisters, often on a urticated background. Involvement of palms and soles have been documented in 50% of the affected patients, whereas mucosal lesions have been described in 38.5% of the patients. Nearly half of anti-p200 pemphigoid patients in Japan have coexistent psoriasis.[82] However, this association has not been found in other parts of the world.

Histopathology of anti-p200 pemphigoid shows neutrophil-rich subepidermal blister, whereas immunofluorescence findings are similar to epidermolysis bullosa acquisita (EBA) revealing linear staining of BMZ with IgG and C3, which localizes to the floor of the split on IIF. Serration pattern analysis ('n' serration) and immunoblotting analysis should be performed to differentiate it from EBA.[81,83]

A case of IgA type of anti-p200 pemphigoid is also reported wherein they demonstrated IgA antibodies against 200-kDa antigen by DIF, IIF, and immunoblotting technique.[84]

There are no guidelines regarding the management of these cases. The treatments tried so far include high-potency topical corticosteroids, systemic corticosteroids, dapsone, azathioprine, doxycycline, cyclosporine, colchicine, and IVIG. The prognosis is better than EBA.

Brunsting-Perry pemphigoid (BPP)

Brunsting-Perry pemphigoid (BPP) is a rare type of sAIBD predominantly affecting the head and neck region. It affects older individuals with vesiculobullous lesions that heal with scarring [Figure 7]. Mucosal involvement has been seen in a few patients during the course of the disease.

Histopathology shows subepidermal blisters with mixed inflammatory infiltrates in a majority of patients (55.2%). Eosinophilic infiltrate (36.2%) and dermal fibrosis (12.1%) are the other findings in BPP.[85] DIF finding is similar to other sAIBD; IIF on SSS reveals either roof or floor staining pattern. Although the exact target antigen is not

known, antibodies to BP180 have been detected in most of the patients. Other antigens known to be involved are BP 230, laminin 332, and type VII collagen. The treatment of BPP is challenging; variable success has been reported with tetracycline, dapsone, methotrexate, and oral corticosteroids. Dupilumab has also been successfully used in one of the BPP cases.[86]

IgM pemphigoid

Pemphigoid diseases are sAIBD with IgG and C3 deposition noted along the BMZ. In addition, linear BMZ staining with immunoglobulin M (IgM) has been documented in 6-18% of patients with pemphigoid. An exclusive IgM deposition along the BMZ has been described by Boch *et al.*[87] in three adult patients presenting with erythematous papules and plaques. IIF on SSS showed a "roof" staining pattern with IgM in all three patients. Immunoblotting analysis showed reactivity against col XVII.

Granular C3 dermatosis

Granular C3 dermatosis was initially defined by Hashimoto *et al.*[88] in 2016. They reported 20 patients who clinically resembled DH but revealed granular deposition along BMZ with only C3. Histopathology showed subepidermal blister, liquefactive degeneration with lymphocytes, eosinophils, and neutrophilic infiltrations with favorable disease course and responsiveness to topical steroids and dapsone. Serological studies have shown the IgA antibodies against transglutaminase 2 (TG2) but not against transglutaminase 3 (TG3) and BP 180 antibodies by ELISA.[89]

Anti-p105 pemphigoid

Chan *et al.*[90] described a patient presenting with mucocutaneous vesicles and bullae clinically resembling PV or toxic epidermolysis necrosis (TEN). Histopathology showed a subepidermal split with neutrophilic dermal infiltrate. DIF findings were similar to other sAIBD, and IIF revealed a dermal staining pattern. IB revealed reactivity to the p105 protein located in the lower lamina lucida.[91]

Orf-induced anti–laminin 332 pemphigoid

Five patients of orf-induced sAIBD have been described recently; all of them demonstrated IgG1 and/or IgG3 autoantibodies against laminin 332. Although laminin 332 was the target antigen, mucosal involvement was not seen predominantly in these patients, unlike MMP.[92]

Acral purpura in dermatitis herpetiformis (DH)

DH classically presents as pruritic papulovesicles distributed mainly over the extensors of elbows, buttocks, and knees. Rarely, DH may present with acral petechiae, purpura, and crusts with minimal or no itching in the setting of no lesions elsewhere in the body. DIF in these patients would show classical findings of DH with papillary dermal IgA staining.[93]

Conclusion

One should be aware of the lesser-known variants of pemphigus and pemphigoid, so that the disease is diagnosed in the early stage and prompt treatment can be initiated. Furthermore, histopathological and immunological tests should be undertaken for confirmation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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