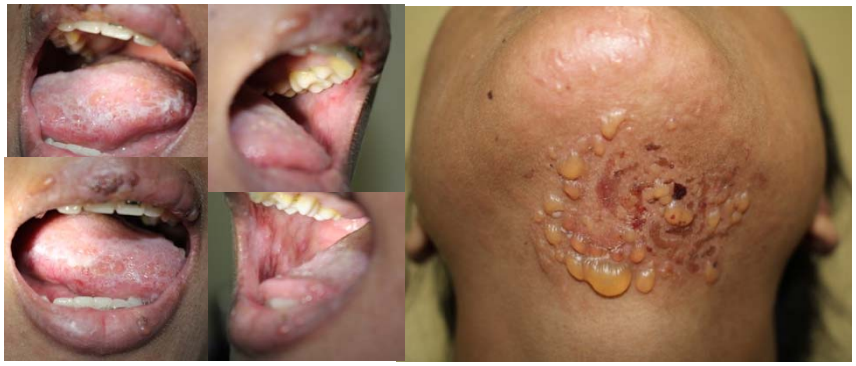
Case 1.2
A 24 year-old Thai woman from Bangkok

Chief complaint: Multiple blistering eruption on face, trunk and oral mucosa for 1 month



(Fig. 1.2.1)

Present illness: Mild pruritic, blistering eruption starting on both ears and spreading to chin, lip, oral mucosa, trunk and some area are healed without scar 1 month prior to visit

Underlying disease: SLE diagnosis since 2010

Current medications: Prednisolone (5) 1 tab EOD, ASA (81) 1 tab oral OD pc, Hydroxychloroquine (200) 1 tab oral OD on Tuesday and Thursday

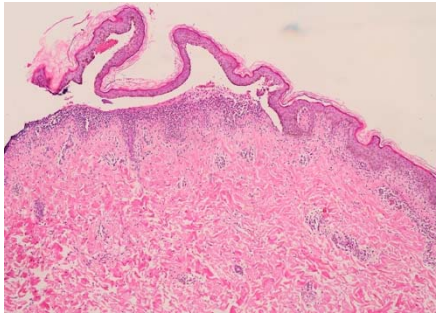
Past history: Otherwise in good health and never had similar rash

Family history: No family history of autoimmune disease

Dermatological examination: (Fig 1.2.1)
- Multiple tense, clear vesicles and bullae on erythematous edematous plaque and normal skin on both ears, vermillion border, chin and trunk
- Multiple tense vesicles on labial mucosa, some areas of crusting and erosions
- Multiple, mild tender, non blanchable erythematous macules on both palms
- Diffuse area of hair thinning, hair pull test: Positive

Physical examination: Systemic examination other than skin lesions revealed no abnormality.

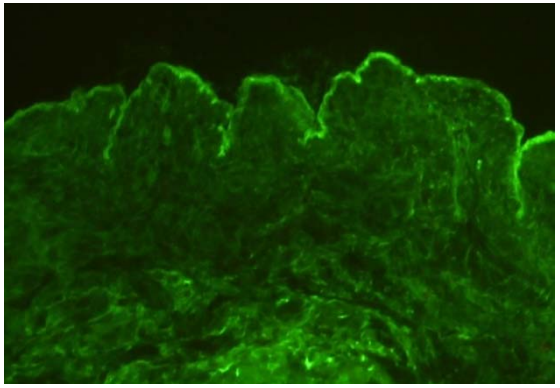
Histopathology: (S17 - 27499C, Abdomen) (Fig.1.2.2)



(Fig. 1.2.2)

- Subepidermal blister with numerous neutrophils and nuclear dusts in the papillary dermis
- Numerous neutrophils and nuclear dusts line along dermoepidermal junction with subepidermal cleft

Direct immunofluorescense:


(Fig1.2.3)

- Homogeneous granular deposition of IgM along dermoepidermal junction (DEJ)
- IgM deposition at superficial blood vessels
- Mixed linear and granular deposition of IgA along DEJ
- Linear deposition of C3 along DEJ, and auto-split tissue show linear deposition on the base

Laboratory investigations:

- CBC: Hct 36.3%, WBC 7,270 cells/µL (N 67.1%, L 26.4%, Mono 6.1%, Eo 0.1%, Baso 0.3%), Platelets 340,000 cells/µL
- BUN/Cr: 9/0.63 mg/dL
- ESR: 38 mm/hr
- UA: Negative for protein and dysmorphic RBC
- ANA: Positive coarse speckled titer≥ 1:1280
- Anti-smith, Anti-nRNP/sm, Anti-SS-A(Ro60KDa), Anti-SS-A(Ro52KDa) and Anti-Nucleosome: All are positive
- Low level of C3, C4 and CH50

Diagnosis: Bullous SLE

Treatment:
Supportive treatment: Photo protection
Specific treatment:

- Oral dapsone 100 mg alternate day
- Oral prednisolone 40 mg/day
- 2% fusidic acid ointment apply on erosions twice daily

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Discussion:

Bullous systemic lupus erythematosus (BSLE) is an uncommon blistering eruption that can occur in patients with systemic lupus erythematosus (SLE). Between 59 and 85 % of SLE patients will have skin manifestations of their disease, but less than 5 % will develop bullous disease.[1-3] The reported incidence of BSLE is approximately 2–3% of all subepidermal autoimmune bullous skin diseases.[4]

BSLE mainly affects young women, frequently of African descent, usually in their second to fourth decades, as observed in patients with SLE[5-7] however, it is also seen in other races, ages, and in males.[1, 3]

Typical clinical features are subepidermal, transient, tense, vesiculobullous eruption, which normally heals without scarring or milia. The bullae can affect any area of the body, including the mucosa, but have a predilection for the trunk, upper extremities,

neck, face, and vermillion border.[5-7] Bullae may appear on erythematous or normal skin. It is important to remember that all cases of BSLE require a diagnosis of SLE. The diagnostic criteria of BSLE was first described by Camisa and Sharma in 1983[2] and were revised in 1988.[1] are as follows;

1. diagnosis of SLE based on ACR criteria
2. vesicles and/or bullae
3. histopathologic features similar to DH
4. DIF with IgG and/or IgM and often IgA at the BMZ
5. IIF testing that can be negative or positive for circulating autoantibodies against the BMZ via the salt-split skin technique.

Our patient had 4/5 criteria which justify the diagnosis BSLE.

Subepidermal vesicles with predominant neutrophil infiltration are the classic histopathologic finding. Distinctive immunopathologic features are multiple immunoreactants (IgG, IgA, IgM and complement) along the basement membrane zone (BMZ)[8] as in our patient that had linear and granular deposition of multiple immunoglobulin along DEJ.

DIF studies on salt-split skin samples from BSLE patients show immune reactant deposition along the dermal side of the blister cavity, below the lamina densa in the BMZ, where type VII collagen is found. The major target antigen in patients with BSLE is the anchoring fibril component type VII collagen in the dermis, however other autoantibodies such as bullous pemphigoid (BP) Ag1, laminin 5, laminin 6 and BPAg 2 can be found.[9]

BSLE may not necessarily correspond to exacerbation of systemic disease, like in our patient where exacerbation was limited to the skin. Some studies showed no clinical or laboratory evidence that indicated flaring of disease.[8] However, some case reports suggest that blistering paralleled internal involvement, particularly lupus nephritis.[10] Other reported systemic associations, although not frequently documented, are hematologic, neurologic, arthritis and serositis.[11-13]

Dapsone is the mainstay of treatment in BSLE, with a dramatic response observed in the first 24–48 hours. This is especially true for isolated BSLE.[14] Our patient received dapsone for treatment and lesions rapidly improve within 72 hours. The conventional treatment for SLE with systemic corticosteroids and antimalarials has revealed minimal improvement for BSLE in the literature.[14] Other immunosuppressants, such as methotrexate, azathioprine, cyclophosphamide, mycophenolate mofetil, have been used in patients with BSLE with modest results.[14] Rituximab, a chimeric monoclonal antibody that reacts with CD20, has shown promising results for refractory BSLE in case reports.[12]

References:

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