原文題目(出處):	Oral Lesions: The Clue to Diagnosis of Pemphigus
	Vulgaris(Hindawi Publishing Corporation Case Reports in
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內文:

Introduction

- 1. most common variant of Pemphigus group
- 2. autoimmune diseases characterized by cutaneous or mucosal blistering
- 3. 40-50 years old
- 4. blisters and painful erosion
- 5. buccal mucosa, lips, and soft palate
- 6. Diagnosis: clinical manifestations and biopsy.

<u>Case Report of Pemphigus Vulgaris at Multiple Intraoral Sites, with No Involvement of Skin</u>

Age: 55 years old

Gender: male

Chief complaint: Painful non-healing ulcers on the left buccal mucosa and left posterolateral border of tongue four months ago

Present Illness:

- 1. burning sensation at both sites for the past six months
- 2. blister which appeared and burst rapidly on the buccal mucosa, after which ulcerations appeared on both the sites

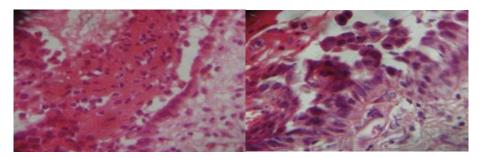
Oral Examination:



1. $2 \text{ cm} \times 2 \text{ cm}$ ovoid shallow ulcer with sloping margins along the line of occlusion of 35 to 37 on the left buccal mucosa

- 2. $1 \text{ cm} \times 1 \text{ cm}$ ovoid ulcer with yellow crusted surface on the left postero-lateral border of the tongue
- →Suspect: Pemphigus, Pemphigoid, Bullous Lichen Planus

Histopathologic Report:



- 1. ulcerated stratified squamous epithelium exhibiting suprabasal split
- 2. round acantholytic (Tzanck) cells with hyperchromatic nuclei within the split
- 3. basal cells attached to the underlying connective tissue, below the split
- 4. dense inflammatory cell infiltrate consisting mainly of plasma cells in the connective tissue

Direct Immunofluorescence Study:

Deposits of IgG and C3 (complement) in a fish-net pattern along the spinous intercellular zone

→ Diagnosis confirmed: Pemphigus Vulgaris

Discussion:

- 1. Intraepithelial blister formation
 - blisters in the epithelium where the patients IgG autoantibodies produced in response to triggering factors target two structured proteins of desmosomes: Desmogleins 1 and 3
 - new Pemphigus antigen Desmoglein 4 and other non-Desmoglein antigens like human α -9-acetylcholine receptor that regulates keratinocyte adhesion and keratinocyte annexin like molecules binding acetylcholine termed pemphaxin and catenin
 - thin separation at the desmosomal region triggers the acantholysis and suprabasal spilt

2. Epidemiology

- 40~50 years old
- Female
- Initial oral manifestation (more than 50%)
- Duration of oral lesion: 3 months~1 years

3. Treatment

- Oral, intralesional, topical corticosteroids
- Corticosteroids along with adjuvants (methotrexate, cyclophosphamide)
- Cholinergic agonists : reverse the acantholysis

4. Our Case

- Oral lesion (blister, and then ulceration): thin roof of the blister ruptures easily and forms an erosion or ulcer in the area
- Pain and burning sensation : most common symptoms
- Multiple intraoral sites : as many reports showed
- Buccal mucosa and tongue: buccal mucosa more common
- Treatment:
 - (i) 100 mg dexamethasone for 3 days with 500 mg of cyclophosphamide, two more cycles of this regime at intervals of 4 weeks each
 - (ii)30 mg Wysolone tablets during the interim 4-week period

題號	題目
1	下列有關天疱瘡(pemphigus)的敘述,何項錯誤?
	(A)與 Nikolsky sign 無關
	(B)和表皮細胞間的 desmosomes 有關
	(C)和表皮基底層細胞間的 hemidesmosome 無關
	(D)組織學變化常見表皮層呈現不同程度的水泡
答案(A)	出處:Oral and Maxillofacial Pathology, third edition
題號	題目
2	下列有關尋常性天疱瘡 (pemphigus vulgaris) 的敘述,何項錯誤?
	(A) 為自體免疫性疾病
	(B) 不會藉由垂直感染給胎兒
	(C) 使用類固醇治療時需量高時間短(high dose, short course)
	(D) 嘴唇上的潰瘍較容易結痂
答案(B)	出處:Oral and Maxillofacial Pathology, third edition