

原文題目(出處)：	Oral mucous membrane pemphigoid in a 6-year-old boy: diagnosis, treatment and 4 years follow-up. Int J Paediatr Dent 2010;20:76-9
原文作者姓名：	Mostafa MI, Hassib NF, Nemat AH
通訊作者學校：	Human Genetics and Genome Research Division, Department of Oro-dental Genetics, and 2Medical Division, Oro-dental Research Department, National Research Center, Cairo, Egypt
報告者姓名(組別)：	intern H 組洪欣儀
報告日期：	2010.05.10

內文：

Introduction

1. Mucous membrane pemphigoid (MMP) is a chronic immune-mediated subepithelial vesiculobullous disease of adults. Children are rarely affected.
2. “Oral pemphigoid (OP)” is used when lesions are confined to the mouth, lesions usually take the form of desquamative gingivitis.
3. An association between MMP and HLA-DQB1*03014,5.

Case report

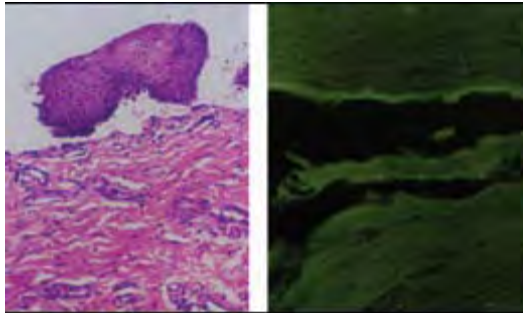
1. Age: 6 Gender:boy

Symptom: gingival bleeding, pain, eating difficulty, and peeling of the gums

- His parents had first noted redness of the gums at the age of 3 with the symptoms that led to the referral developing at the age of 5.
 - 7 months prior to referral to the department, a diagnosis of chronic periodontitis was made but he did not respond to the treatment.
2. The diagnosis was established as OP based on the clinical, histological, and immunofluorescence findings.
 - Clinical examination revealed generalized redness of the gingiva in both upper and lower jaws (Fig. 1a, b).



- Application of gentle lateral pressure on the gingiva using a piece of cotton caused peeling of the gums, indicating a positive Nikolsky's sign. Desquamative gingivitis was suspected.
- Histological findings showed separation of the epithelium from the underlying connective tissue (Fig. 1c).
- Direct immunofluorescence showed linear deposition of IgG and C3 at the mucosal-submucosal junction (Fig. 1d).
- The patient had the genotype HLADQB1*0501/0301, 0309, 0313.



3. Therapy-1(2 weeks)(U+L: Betamethasone):

- Occlusive topical steroid therapy (5 mg **betamethasone cream** b.d.) vacuum-formed trays with spacers at the gingival portion · An artificial space to allow the eruption of the permanent incisors (Fig. 1e).



- 0.2% chlorhexidine mouth wash twice/day to avoid candidosis and calculus formation.
- Avoid highly acidic foods and hard foods.
- Scaling and oral hygiene measures were performed.

After 2 weeks a small improvement was noted.

4. Therapy-2(5 weeks)(L:Betamethasone U:Fluocinonide)

- Continuing betamethasone for the lower jaw but changing to **0.05% fluocinonide ointment** for the **upper** one jaw.

Five weeks later, the patient complained of bleeding, pain, and peeling of the gums of the lower jaw only and clinical examination confirmed that there was a greater improvement in the upper gingival mucosa (Fig. 2a).



5. Therapy-3(5 weeks)(U+L: Fluocinonide)

6. After that period (12 weeks), the signs and symptoms had resolved except for the redness of the gingiva (Fig. 2b).
7. The patient was **reviewed monthly** and signs and symptoms **recurred after 6 months**. Again they responded to **2 weeks** topical treatment with fluocinonide.
8. During the **4 years** of follow-up (Fig. 2c,d), signs and symptoms recurred at approximately **six monthly intervals**, although there was a remission for **1 year gap** before the last recurrence. Patient was also reviewed every **6 months** for **extra-oral examination** particularly ophthalmological.

Comment

1. Desquamative gingivitis is a non-specific clinical manifestation of several diseases, the most common being MMP, lichen planus, and pemphigus vulgaris .
2. DIF shows intercellular epithelial antibodies in pemphigus vulgaris whereas lichen planus shows fibrin staining of the basement membrane.
3. Several studies indicated that **HLADQB1*0301 alleles** confer a predisposition to all subgroups of MMP in Caucasian patients.
4. It may have a role in T cell recognition of basement membrane antigens and the subsequent B cell production of anti-basement membrane zone autoantibodies.
5. It has been suggested that OP and ocular pemphigoid are part of a spectrum of a single disorder and that they have the same genetic predisposition, thus it is interesting to note that the patient we describe has this allele.
6. **Topical corticosteroids** are the treatment of choice for MMP, especially for localized oral lesions, although **immunosuppressants and dapsone** have also been used.
7. In this case, we found **fluocinonide** to be more effective than betamethasone. Yet, similar reported cases are very few and the true benefits of such drugs are therefore not yet clear.
8. Intervals between relapses may lengthen as the child becomes older and more attentive to oral hygiene and aware of the stimulating factors. At the same time,

- shedding and eruption mechanisms could be among the predisposing factors.
9. In conclusion, MMP should be considered in the differential diagnosis of chronic desquamative gingivitis in childhood. Occlusive therapy with topical fluocinonide until symptoms resolve is recommended, with repeated courses if symptoms recur. This approach has fewer side effects than systemic corticosteroids or dapsone, particularly in childhood. We also recommend periodic ophthalmologic examination in affected patients.

What this paper adds

- Recording a very rare case of childhood OP.
- Using occlusive topical steroid therapy in the treatment of childhood OP.

Why this paper is important to paediatric dentists

- Oral pemphigoid that is common in old age can also affect children and must be considered in the differential diagnosis of chronic desquamative gingivitis in childhood.
- Adding a new clinical experience for OP cases, as these cases are extremely rare.

題號	題目
1	以下除了哪一種疾病外，會有 Desquamative gingivitis? (A) Mucous membrane pemphigoid (MMP) (B) Lichen planus (C) Pemphigus vulgaris (D) Erythema Multiforme
答案 (D)	出處：International Journal of Paediatric Dentistry 2010; 20: 76-79
題號	題目
2	以下何者與 OP(Oral pemphigoid)無關? (A) Desquamative gingivitis (B) PTEN (C) HLA-DQB1 (D) IgG and C3
答案 (B)	出處：International Journal of Paediatric Dentistry 2010; 20: 76-79