# Atypical Presentation of Oral Erythema Multiforme - A Case Report

Samjhana Gautam,[1] Senchhema Limbu,[2] Manisha Malla,[3] Parajeeta Dikshit[4]

*[1]Post Graduate Resident, [2,4]Professor, [3]Assistant Professor*

*[1-4]Department of Pedodontics and Preventive Dentistry, Kantipur Dental College and Teaching Hospital, Kathmandu, Nepal*

## ABSTRACT

Erythema multiforme (EM) is an acute, self-limiting, sometimes recurrent mucocutaneous inflammatory disorder recognized as hypersensitivity reaction primarily triggered by certain infections and drugs. It is often seen as confusing disease entity in children characterized by eruptive lesions of oral mucous membrane. Although oral involvement can occur in association with lesions of skin and other mucosal surfaces, the diagnosis is a little more challenging when the disease is completely limited to oral mucosa. Here, we report a typical case of oral EM that is confined in lip in a 7-year-old male child; with an acute onset of crusted ulcer with hemorrhagic encrustation, pain and swelling of lower lip since 2-days. Based upon patient's detailed history of prodromal sign and symptoms, disease progression, clinical examination of wound and hematologic investigation clinical diagnosis of erythema multiforme was made. The case was treated with topical corticosteroid and topical antiviral cream for a week showcasing resolution of lesion.

**Keywords:** *Erythema multiforme, oral mucosa, crusted ulcer, lip.*

## INTRODUCTION

Erythema multiforme (EM) is an acute, self-limited, inflammatory disease manifesting on skin and often other mucous membranes inclusive of oral mucosa.[1] It is considered as type-IV hypersensitivity reaction typically preceded by viral infections, especially herpes-simplex virus (HSV), or certain medications, though many cases are idiopathic.[2] EM is classified into major and minor forms based on the nature and distribution of skin lesions and the degree of mucosal involvement.[1] A subset of EM may affect only oral mucosa without involvement in skin and other mucosa.[3]

Oral lesions present as an acute, vesiculobullous stomatitis of oral mucosa often involving vermillion of lips causing cracking, bleeding, swelling, and crusting but heal without scarring.[4,5]

### Correspondence

*Dr. Senchhema Limbu*
*Professor*
*Departments of Pedodontics and Preventive Dentistry*
*Kantipur Dental College and Teaching Hospital, Kathmandu, Nepal*
*E-mail: senchhe@hotmail.com*

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## CASE REPORT

A 7-year-old male child was referred to the Department of Pedodontics and Preventive Dentistry at Kantipur Dental College, Kathmandu, from Shahid Gangalal National Heart Centre, Kathmandu, with a chief complaint of a wound on lower lip, that had been spontaneously bleeding and forming clots since last two days. The patient had a medical history of Cyanotic Heart Disease, including Situs Solitus, Levocardia, and Tetralogy of Fallot and had been admitted to Shahid Gangalal National Heart Centre for cardiac surgery, which was scheduled 4 days following the referral. The primary reason for referral was difficulty in performing oropharyngeal intubation for the surgery due to the lip lesion.

The lip wound was associated with mild pain, tingling sensation and swelling causing difficulty in eating. One week priorly, the patient had experienced moderate-grade fever of 100°F. His parents reported that two days earlier, he had dry, crusted lips with small bumps on the lower lip, which he removed unintentionally, leading to bleeding and hemorrhagic crusts.

On examination, bilateral submandibular lymph nodes were palpable, tender, and ranged from soft to firm. Extraoral examination showed severe ulceration with bloody crusts, cracks and edema on the lower lip (Figure 1). Upon gentle removal of clot with normal saline soaked gauze, the lip revealed a central target-like lesion with hemorrhagic crusting (Figure 2). The lesion was painful to touch and bled easily with slight manipulation. Intraoral examination revealed absence of any lesions on other surrounding oral mucosa. Patient's history along with the clinical examination indicated absence of lesions on the hands, skin, eyes, or genitals. Drug history revealed no changes in medication or use of any chemicals that could explain the condition. The patient was oriented to time, place, and person, with a normal body temperature of 98°F and pulse rate of 82 beats per minute. Thus, a provisional diagnosis of traumatic ulcer of the lower lip was made. The child was prescribed a topical analgesic antiseptic agent (Deltagel -combination of chlorhexidine, lignocaine, and metronidazole), for symptomatic relief and advised to avoid manipulating the wound while maintaining oral hygiene.

His mother also reported that the child's mouth would often be full of blood clots during sleep, and application of topical antiseptic cream had not led to improvement (Figure 3). Two days later, the patient returned back with an unhealed, aggravated wound with severe hemorrhagic encrustation (Figure 4). The case was further consulted with multidisciplinary approach and hematological investigations were carried on including Bleeding Time, Clotting Time, Prothrombin Time, INR, APTT, CBC, Hb and ESR. The hematological investigations showed no significant findings, with normal ESR.

Based on clinical examination, history of recent fever, dry lips with papules and a central target lesion, along with the patient's immunocompromised status, a clinical diagnosis of Herpes virus-associated erythema multiforme (HAEM) was made.

Re-debridement of the wound was performed under topical local anesthesia with normal saline; revealing an irregular ulceration of the lower lip with spontaneous bleeding (Figure 5). The patient was prescribed topical corticosteroid (Triamcinolone acetonide 1% gel), topical antiviral cream (Acyclovir), and multivitamins for a week. The patient was advised to follow a protein-rich diet, maintain hydration, and seek early consultation with an oral physician if prodromal symptoms appeared.

After three days, the lesions showed significant improvement (Figure 7) and complete resolution was achieved after one week (Figure 8). There is no reoccurrence of the lesion till 4-months follow-up.

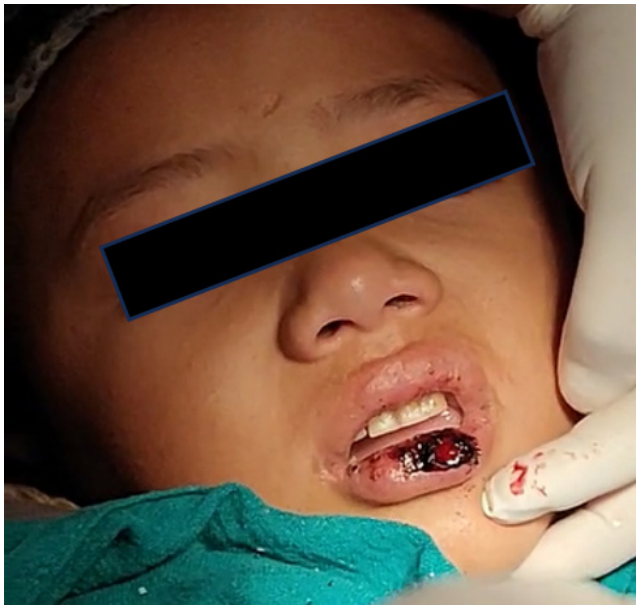

**Figure 1:** Ulceration with hemorrhagic crust on lower lip

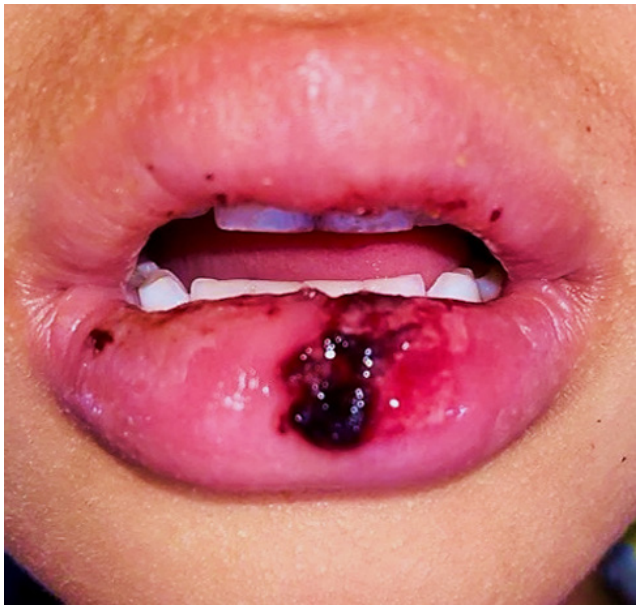

**Figure 2:** Central target lesion on lip seen after gentle removal of clot

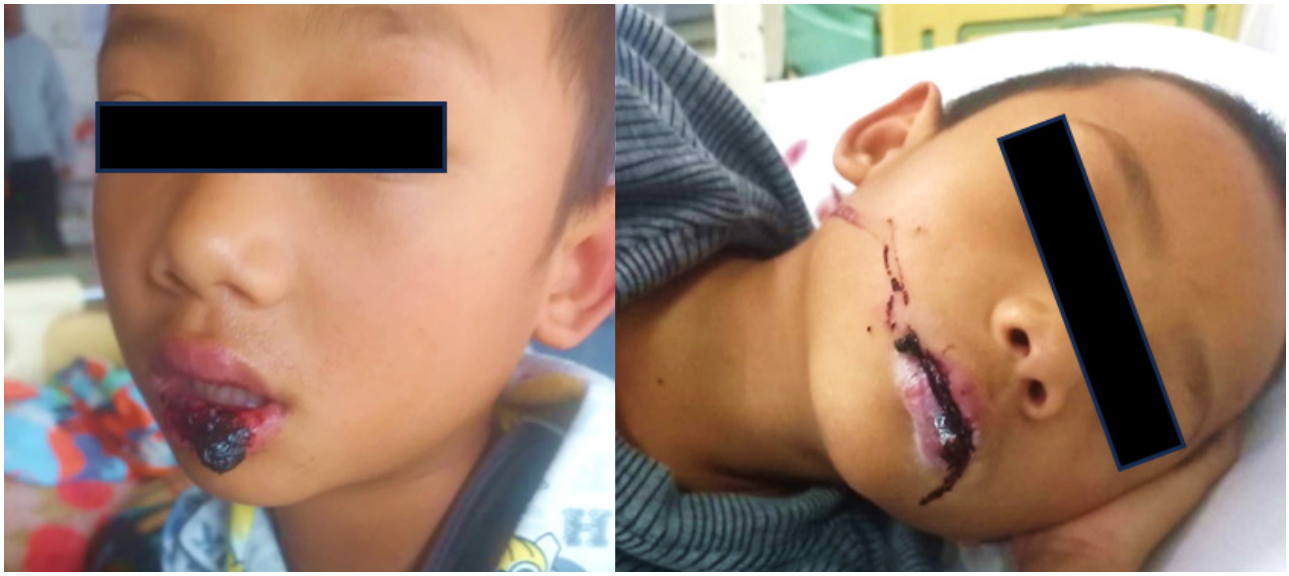

**Figure 3:** Pictures of unhealed lesion at admitted hospital bed

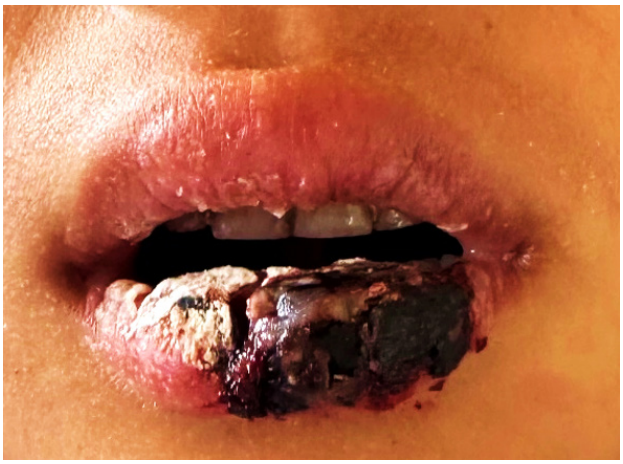

**Figure 4:** Patient returned back with unhealed wound after 2 days

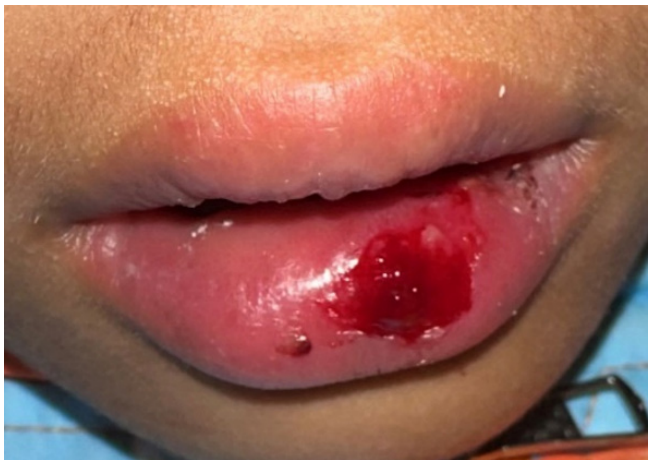

**Figure 5:** Re-debridement under topical local anesthesia with normal saline

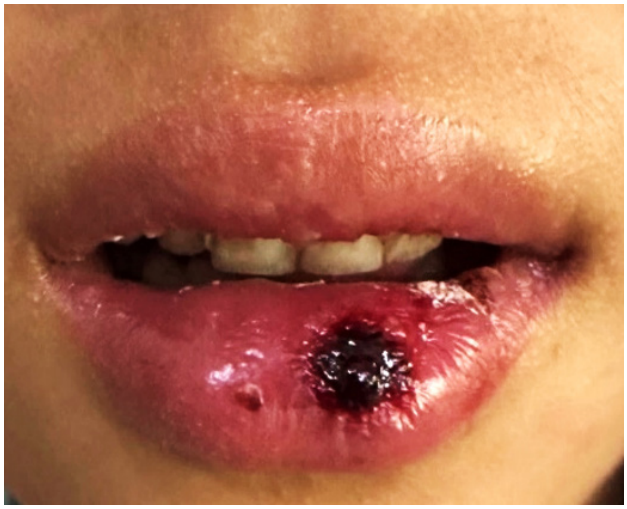

**Figure 6:** After 30 minutes of debridement

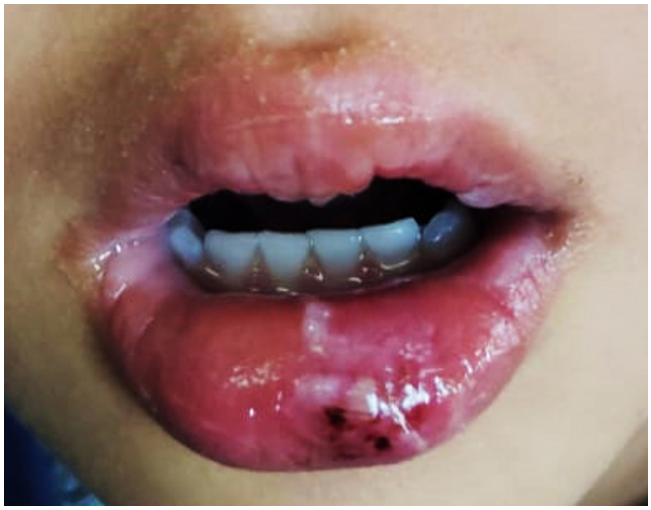

**Figure 7:** Healing wound after 3 days (Under topical medications)

**Figure 8:** Completely healed lesion after 1 week

## DISCUSSION

Erythema multiforme (EM) is an acute mucocutaneous hypersensitivity reaction characterized by a skin eruption, with or without oral or other mucous membrane lesions.[1] It can be induced by certain infections, drug intake, autoimmune diseases, and malignancies.[4] HSV infection is considered as one of the most common predisposing factors of EM with lip and oral mocosa involvement and has been identified in up to 70% of the EM diagnosed.[6]

According to Sokumbi et al in herpes associated erythema multiforme prodromal symptoms of malaise, fever, and myalgias are present a week or more before the onset of EM in general.[7] Most commonly affected sites are lips (36%), buccal mucosa (31%), tongue (22%) and labial mucosa (19%).[3] Oral features are distinctive, with the formation of blisters on lips and oral mucosa which eventually break and coalesce to form erosive and ulcerative lesions, followed by greyish pseudo membrane. Gingival desquamation, hemorrhagic crusted lip lesions, and a positive Nikolsky's sign is also seen.[5] Acccording to study by Hasan S et al.[5] swollen lips along with typical blood tinged crusted lesions are the hallmark of EM, as observed in our case. Target lesions may be seen on the lip but rarely on the intraoral mucosa.[8]

The incidence of EM is postulated to be far less than 1%, but possibly greater than 0.01%. It occurs predominantly in young adults, with a slight female preponderance and without racial predilection.[7] The peak age at presentation is 20–40 years although as many as 20% of cases are children.[1] Shklar et al.[9] mentioned in their study that Erythema Multiforme is often seen in children as a confusing disease entity characterized by eruptive lesions of the oral mucous membrane. Although the oral involvement may occur in association with lesions of the skin and other mucosal surfaces, the disease may be entirely confined to the oral mucosa, rendering diagnosis somewhat more difficult.[9] In our case the lesion was confined only in the lower lip and labial mucosa similar to the cases reported by Weston et al.[6]

Diagnosis is based primarily on clinical manifestations. The character and distribution of oral lesions are fairly typical. Examination of skin lesion facilitates diagnosis if oral lesions are suggestive of possible erythema multiforme. According to Gungor T et al. laboratory tests are of no diagnostic value except for an increased erythrocyte sedimentation rate in severe cases, lacks specificity and are helpful only in ruling out other possible diseases.[10] The detection of intralesional HSV-DNA through PCR, along with HSV genes and specific antibodies (IgG and IgM) in serology serves in confirming the diagnosis of EM associated with HSV.[3]

Shklar G et al.[9] proposed that histopathologic appearance is nonspecific with spongiosis, chronic inflammation, and subepithelial vesicle formation. Hydropic degeneration occurs in the stratum germinativum and spinosum.

Inflammatory infiltration within connective tissue includes eosinophils, lymphocytes, plasma cells, and histiocytes.[9]

Primary herpetic gingivostomatitis, aphthous ulcers, toxic epidermal necrolysis, pemphigus vulgaris, subepithelial immune disorders (pemphigoid and others), primary herpes, Behcet's disease, bullous or ulcerative lichen planus, diphtheria, systemic lupus erythematosus, acute ulcerative necrotizing gingivitis, cyclic neutropenia, and allergies have all been included in the differential diagnosis for oral lesions of erythema multiforme.[4]

The first step in management of EM is identification of etiological agent. Soares A et al recommended treating acute EM with oral mucosal involvement with topical corticosteroids, oral antiseptics solutions or oral antihistamines as a first line of treatment.[2] If mucosal involvement is severe with poor oral intake, hospitalization for fluid and electrolyte replacement and systemic corticosteroid with tapering dose is recommended.[2] Herpes associated erythema multiforme is often effectively managed with oral Acyclovir (200 mg, five times daily for 5 days), only if started within first few days. For recurrent cases, a continuous low dose of oral acyclovir (10mg/kg) for 6-12 months period is necessary.[3,4] Reports show that in cases where systemic corticosteroids and acyclovir fail, drugs such as Thalidomide, Dapsone, Levamisole, and Azathioprine can be used.[3] The present case was managed succesfully with topical corticosteroid and topical antiviral ointment application three times daily that lead to complete resolution of the lesion in a week, without any recurrence in 4 months duration; similar to as by Soares A et al.[2]

The limitation of this case report is that the diagnosis was entirely based on detailed history, clinical presentation and medical condition of patient. Oral biopsy of lesion and PCR for HSV-DNA was not feasible as the patient was referred case and was undergoing treatment in cardiac hospital where he had to undergo cardiac surgery after 4-days.

## CONCLUSIONS

Erythema Multiforme is encountered as an acute, rare, self-limiting inflammatory mucocutaneus condition sometimes limited to oral mucosa. Even though it is rare, in cases of acute stomatitis, erythema multiforme should always be taken into consideration. In the present case the condition only involved lips and taking into account of viral etiology the case was managed successfully with topical corticosteroid and antiviral cream. While erythema multiforme usually affects young adults, in cases of acute stomatitis pediatric dentists should always take erythema multiforme into consideration.

## ACKNOWLEDGEMENTS

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**Conflict of Interest:** None

JNAPD

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