ERYTHEMA MULTIFORME: A CASE REPORT

A Case Report

Steffi Jose*, Prachi Patel*, Bhavin Dudhia, Roseline Patel*****

ABSTRACT

Erythema multiforme (EM) is a mucocutaneous disorder, which ranges from a mild, self-limited, cutaneous, exanthematous variant with minimal oral involvement to a progressive, fulminating, severe variant with extensive mucocutaneous epithelial necrosis (Stevens-Johnson syndrome; and toxic epidermal necrolysis). EM results from a cell-mediated immune reaction against a precipitating factor, it is characterized by ulcerations, erosions, and bleeding within the mucosa associated with encrustations and tissue tags. Dermal counterparts present with target iris lesion. Here, we report a case of EM major where there are classical oral and dermal lesions in a 40yearold male patient.

Key Words: Erythema multiforme, Mucocutaneous disorder, Target lesion, Vesiculo-bullous lesions

Received: 30-07-2018; **Review Completed:** 23-10-2018; **Accepted:** 18-01-2019

INTRODUCTION:

Ferdinand Von Hebra described erythema multiforme (EM) in the year 1866 as a self-limited and acute skin disease that is symmetrically scattered on the extremities with a typical recurring concentric pattern in the form of "target lesion".[1,4] Despite being often caused by, or at least associated with, infection or drug therapy, the pathogenic mechanism of EM remains unclear, and as a consequence there are no evidence-based, reliably effective therapies[2] EM and related disorders comprise a group of mucocutaneous disorders characterized by variable degrees of mucosal and cutaneous blistering and ulceration that occasionally can give rise to systemic upset and possibly compromise life[2]

CASE REPORT:

A 40 year old male presented with a painful mouth owing to ulcers associated with bleeding since 2 weeks.

History revealed fever 2 weeks ago, following which he noticed vesicles on the bilateral buccal mucosa [Figure 1 & 2] and labial mucosa [Figure 3 & 4]. The vesicles subsequently ruptured leaving ulcerated areas and encrustations associated with severe pain and bleeding on mastication, making it difficult to consume food. Five days after the appearance of oral lesions, he developed cutaneous lesions on the upper and lower extremities, which were preceded by itching. General physical examination unveiled multiple concentric target or iris lesions on upper and lower extremities [Figure 5

Pre- Treatment

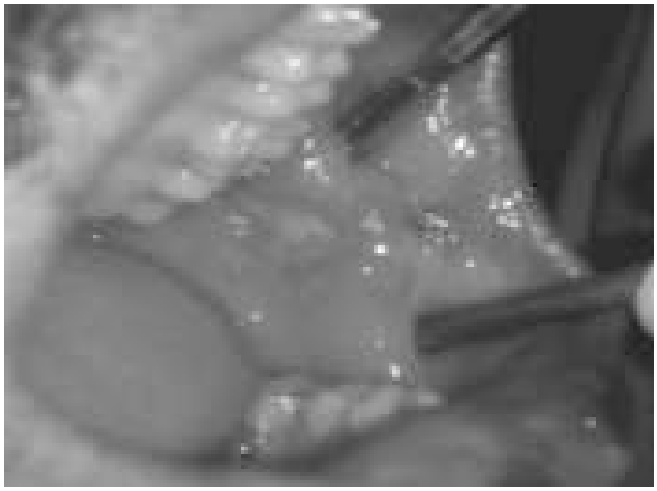

Figure 1 Shows vesicles on left buccal mucosa

Figure 2 Shows vesicles on right buccal mucosa

PG Student*, Professor & Head**, Reader***

DEPARTMENT OF ORAL MEDICINE AND RADIOLOGY AHMEDABAD DENTAL COLLEGE AND HOSPITAL

ADDRESS FOR AUTHOR CORROSPONDENCE : DR. STEFFI JOSE, TEL: +91 9429903876

Figure 3 Shows encrustations on lips

Figure 4 Shows ruptured vesicle lower labial mucosa

Figure 5 Shows concentric target lesion on left hand

Figure 6 Shows concentric target lesion on left leg

& 6].

Encrustations that bleed readily on provocation were noticed on the lips and upper and lower labial mucosa which were jagged and tender, sparing gingiva. Compromised oral hygiene, diffuse multiple ulcers, and erosions were present on the upper and lower labial mucosa, bilateral buccal mucosa extending to the retromolar area. The floors of ulcers were covered by pseudomembrane, surrounded by erythematous halo with irregular margins and a nonindurated base with bleeding on provocation.

Erythema Multiforme, Stevens-Johnson syndrome, Mucous membrane pemphigoid and pemphigus vulgaris were considered as differential diagnosis.

As patient was not ready for biopsy, cytosmear was taken. Cytosmear report showed basophilic cells and a few nucleated eosinophilic cells. Allergy test showed patient was allergic to groundnut, cabbage and housedustmite.

Considering the history, prodromal symptoms, hemorrhagic crustations involving the upper and lower lips in addition to widespread involvement of oral mucosa and symmetric distribution of the target/iris/bull eye shaped dermal lesions (pathognomic) on extremities; the condition was diagnosed as Erythema Multiforme major.

The patient was prescribed ointment Betnovate 0.1% t.i.d., along with a topical antiseptic analgesic gel (Dologel CT). Total resolution of the oral and dermal lesions with hypopigmentation in oral

Post-Treatment

Figure 7 Shows Healedhypopigmented left buccal mucosa

Figure 8 Shows Healed Hypopigmented Right Buccal Mucosa

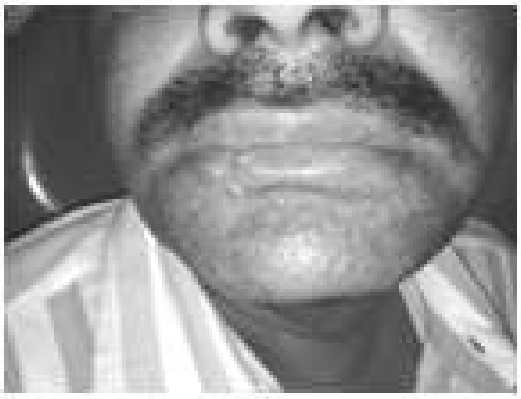

Figure 9 Shows Healed Hypopigmented Lips

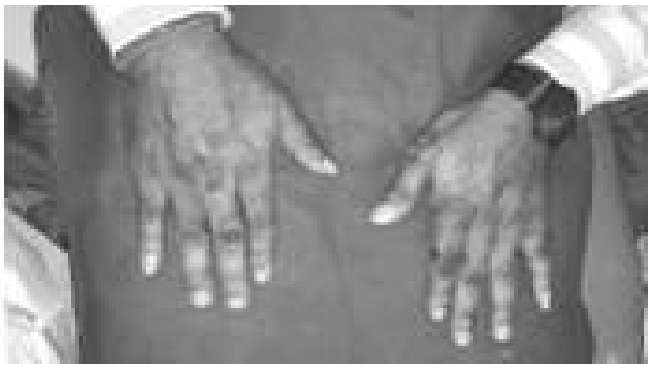

Figure 10 Shows healed hyperpigmented hands

Figure 11 Shows Healed Hyperpigmented Legs

[Figure 7, 8 & 9] and hyperpigmentation in dermal [Figure 10 & 11] lesions were attained in 1 month.

DISCUSSION:

Erythema multiforme is an acute, sometimes recurrent, mucocutaneous condition of uncertain etiopathogenesis that can follow the administration of drugs or infections.[3] Medical literature has coupled numerous factors to the development of EM. These include infections, use of certain medications, malignancy, autoimmunity, radiation, immunization, and menstruation. of these, infection represents approximately 90% cases, and the most common agent is herpes simplex virus (HSV), which is drawn in up to 70–80% of the cases[4].

Classification of EM is according to the degree of mucosal involvement and existence of dermatologic lesions [4,6]

i. Erythema Multiforme minor (EM minor) - typical targets or raised, edematous papules distributed acrally.

ii. Erythema multiforme major (EM major) - Typical targets or raised, edematous papules distributed acrally with involvement of one or more mucous membranes; epidermal detachment involves less than 10% of total body surface area (TBSA).

iii. Stevens-Johnson syndrome (SJS) - detachment below 10% of the TBSA plus widespread erythematous or purpuric macules or flat atypical targets and iv. Toxic epidermal necrolysis (TEN) - detachment above 30% of the body surface are plus widespread purpuric macules or flat atypical targets.

Typical and/or raised atypical target lesions are the hallmark of Erythema Multiforme.[2] Disease involves less than 10% of the body surface area. Lesions are often symmetric in distribution, with a predilection for the extensor surfaces of the extremities.[2] Our case was presented with typical target lesions on upper & lower extremities.

Erythema Multiforme usually affects the lingual, buccal, and/or labial mucosa, and less frequently the floor of the mouth, palate and the gingivae.

The oral manifestations of the spectrum of EM range from tender superficial erythematous and hyperkeratotic plaques to painful deep hemorrhagic bullae and erosions.[2] The oral lesions initially

manifest with edema, erythema, and erythematous macules of the lips and buccal mucosa, followed by the development of multiple vesicles and bullae that quickly rupture and result in pseudomembrane formation[2,6]. The lips tend to become swollen and show diagnostically distinctive bloody excrustations.[2] Intact vesicles are rarely observed because they rapidly breakdown to form illdefined ulcers.[2,6] Affected patients may also have trismus, dysphonia, dysarthria, and/or dysphagia.[2] Our patient had presentation of encrusted lesions that bled readily on provocation on the lips, upper and lower labial mucosa. Patient had compromised oral hygiene, diffuse multiple ulcers and erosions were also present on the upper and lower labial mucosa, bilateral buccal mucosa extending upto the retromolar area. Differential diagnosis are to be considered in the lesion confined to oral cavity are herpes, vesicullobullous lesions like pemphigus vulgaris, bullous pemphigoid. Herpetic lesions are usually smaller and well circumscribed, more common in keratinized mucosa especially in gingiva.[8] Our case did not have any gingival ulceration. Extensive irregular ulcerations in the lining non keratinized mucosa were seen in our case also showed mild ulcerations, which were typical of EM and were not feature of herpes infections. Temporal relationship between the drug intake and onset of disease excludes the possibility of any infectious etiology.

Microscopic examination of skin lesions reveals edema just below the epidermis that when mild or moderate, produces urticarial lesions; when the edema is severe, blisters are formed. Other histological features consist of dilation of blood vessels, accompanied by a perivascular infiltration composed mainly of lymphocytes, nuclear dust resulting from disintegration of neutrophils and eosinophils (leukocytoclasis), edema, acanthosisand erythrocytes extravasations. The characteristic histopathological change of EM minor is epidermal cell death, which is termed “satellite cell necrosis”, mimicking apoptotic cell death.[5] In our case cytosmear report showed basophilic cells and a few nucleated eosinophilic cells. There were no tzanck cells seen in cytology report which further excludedherpes simplex virus.

The mainstay of treatment is topical like kenacort, flucort or betnovate; and/orsystemic corticosteroids like prednisolone (0.5–1.0 mg/kg/day tapered over 7–10 days) or azathioprine, or both or other immunomodulatory drugs such as cyclophosphamide, dapsone, cyclosporine, levamisole, thalidomide or interferon-a.[2] Cyclosporine given intermittently may control recurrent EM.[2] Antiseptic, analgesic and anesthetic mouthwash containing benzydamine hydrochloride, diphenhydramine hydrochloride and diclonine are also prescribed.[1] In our case the patient was prescribed ointment Betnovate 0.1% t.i.d., along with a topical antiseptic analgesic gel (Dologel CT).

Most cases of EM are self-limited, with lesions evolving over 1-2 weeks and subsequently resolving within 2-3 weeks. Patients who form keloids may be at higher risk. Hypopigmentation or hyperpigmentation may follow resolution of lesions.[1] Our case showed healing with hypopigmentation of oral lesions and hyperpigmentation of dermal lesions on follow up in 1 month.

Recurrence is common in EM (up to one-third of cases) but is not common in Stevens - Johnson syndrome (SJS) / Toxic Epidermal Necrolysis (TEN). Failure to diagnose SJS early in the course may result in a premature discharge of the patient, with subsequent deterioration in patient's condition. Patients and parents, when appropriate, should be warned about potential long-term complications.[1]

CONCLUSION:

In conclusion, EM is a mucocutaneous disease that requires a prompt and precise diagnosis. An important step in the management of erythema multiforme is recognition and withdrawal of the causative agent. The treatment withtopical corticosteroids alongwith an antiseptic analgesic can be considered before prescribing systemic corticosteroids. By delivering apt information, educating the patient and prevention of contact with the causative agent oral physicians can play a role in preventing the recurrence of these lesions.

REFERENCES:

1. Bashir T, Arti S, Jyoti G, Monu Y, Vijay K, Naeem A. Erythema Multiforme: A Case Report. Int J Sci Stud 2014;2(7):249-251.
2. Parvinderjit S, JasbirKaur, Erythema Multiforme-Oral Variant: Case Report and Review of Literature. Indian J Otolaryngol Head Neck Surg (July 2011) 63(Suppl 1):S9–S12
3. Kamala K A, Ashok L, Annigeri R G, Herpes associated Erythema Multiforme. ContempClin Dent 2011;2:372-5.
4. Rashmitha A, Gotoor SG, Muppirala S, Raju DR. Erythema multiforme: A case report. J Indian Acad Oral Med Radiol 2017;29:153-5.
5. Oliveira, L.R. and Zucoloto, S. Erythema Multiforme Minor: A Revision. American Journal of Infectious Diseases 4(4):224-231, 2008.
6. Yu-Feng H, Hui-Wen Y, Jen-Hong Y, Erythema multiforme-case report. J Dent Sci 2006:1(2)
7. Rakhi I, Prabhu N. Etiopathogenesis of Erythema Multiforme - A Concise Review. Etiopathogenesis of Erythema Multiforme - A Concise Review.2017; 5(4)
8. BharatiPatil et al., Oral Blistering - Report of Two Cases & Literature Review. J of Clinical and Diagnostic Research. 2013 Sept, Vol-7(9): 2080-83