CASE REPORT

ERYTHEMA MULTIFORME

Authors: (1) Nupur Agarwal, (2) Puneet Gupta (3) Sunil R. Panat
(1) Senior Lecturer, Department of Oral Medicine and Radiology, Institute Of Dental Sciences, Bareilly, (U.P)
(2) Senior Lecturer, Department of Public Health Dentistry, Institute Of Dental Sciences, Bareilly, (U.P)
(3) Principal, Professor and Head, department Of Oral Medicine and Radiology, Institute Of Dental Sciences, Bareilly, (U.P)

Correspondence: Dr. Nupur Agarwal, Senior Lecturer, Department of Oral Medicine and Radiology, Institute Of Dental Sciences, Bareilly, (U.P), Contact No. 8937946494 e-mail: rupun48@gmail.com

Abstract: A Female patient aged 35 years reported to the department of oral medicine and radiology with the chief complaint of pain and ulcers in the mouth since 3 months. On extra oral examination skin lesions i.e. vesicles on the chest, Axilla And Dorsal Surface Of Hand were evident. Lesions on the hand were resembling the target lesion. Lips were crusted and showed bleeding. Intra-Oral Examination showed the presence of Mixed red and white, diffused large, irregular lesions on the buccal mucosa of the both right and left side, palate and tongue with the necrosed tissue. Considering the history and clinical examination provisional diagnosis of Erythema Multiforme was given.

Key words: Target Lesion, Topical Steroid, Erythema Multiforme

Introduction

Erythema multiforme was initially described in 1866 by Ferdinand Von Hebra as an acute self-limited skin disease, symmetrically distributed on the extremities with typical and often recurrent concentric “target lesion”. Erythema multiforme is a reactive mucocutaneous disorder that comprises variants ranging from a selflimited, mild, exanthematous, cutaneous variant with minimal oral involvement (EM minor) to a progressive, fulminating, severe variant with extensive mucocutaneous epithelial necrosis (Stevens-Johnson syndrome: SJS; and toxic epidermal necrolysis).

CASE REPORT:

A Female Patient aged 35 years reported to the department of oral medicine and radiology with the chief complaint of pain and ulcers in the mouth since 3 months. History of Presenting Illness revealed that patient was suffering from ulcers and pain in the mouth since 3 months. patient had difficulty even on swallowing. Ulcers were painful and bleeds when they ruptures. Pain was sudden in onset, severe in intensity, continuous in nature and was of lancinating type. Patient was also suffering from burning sensation and skin lesions on the chest and axilla. Initially lesions start as the vesicles which used to ruptures in 2-3 days to form the ulcer which bleeds on palpation. Lesion was tender on palpation and was not scrapable. Nicolky's sign was negative. Generalized stains and calculus is present.

Considering the history and clinical examination provisional diagnosis of Erythema Multiforme was given. Pemphigus Vulgaris, Bullous Lichen Planus, Herpes Zoster And Herpes Simplex were included in the Differential Diagnosis.

Complete blood picture and cytosmear and skin biopsy were performed, all the blood counts were under the normal range. Cytosmear revealed numerous acantholytic cells which appear to be cytomorphologically normal with interspreaded neutrophils. Histopathological impression is of acute intraepithelial vesiculobullous lesion. Skin biopsy confirmed the diagnosis of erythema multiforme.

Local Application of kenakort 2 times daily and application of gentian violet, oral dose of corticosteroid i.e. prednisolone 30 mg twice a day for one week and Antiseptic analgesic and anaesthetic mouth wash containing benzydamine hydrochloride, diphenhydramine hydrochloride and diclonine were prescribed to the patient. Patient responded well to the treatment, after one week there was 70% reduction in the lesions, patient was further advised to continue with the Local Application of kenakort 2 times daily and local application of gentian violet. Systemic dose was reduced to 20 mg twice a day.

DISCUSSION:

Erythema multiforme was initially described in...
Erythema Multiforme (EM) is an acute mucocutaneous hypersensitivity reaction characterised by a skin eruption, with or without oral or other mucous membrane lesions. Occasionally EM may involve the mouth alone. It can be classified into the types EM minor: Typical target or raised, edematous papules distributed acrally. EM major: Typical target or raised, edematous papules distributed acrally with involvement of one or more mucous membrane, epidermal detachment involves less than 10% of total body surface area. SJS/TEN: widespread blisters predominant on the trunk and face, presenting with erythematous or pruritic macule and one or more mucous membrane erosions, epidermal detachment is less than 10% for SJS and 30% for TEN. 

50% of cases are idiopathic. Most notably causes are infectious agents and drugs. Infectious causes are more common in children and herpes simplex infection is most common cause in young adults. Erythema multiforme has been reported to be triggered by numerous agents, particularly viruses, especially herpes simplex virus (HSV) but other herpesviruses (varicella-zoster virus, cytomegalovirus, Epstein-Barr virus), adenoviruses, enteroviruses (Coxsackie virus B5, echoviruses), have all been implicated. Drugs most commonly implicated are Allopurinol, Barbiturates, Carbamazepine, NSAIDs, Penicillin, Phenytoin Sulphonamides. Other causes are Malignancy, Hormonal, Collagen vascular disease, Immunological disorder, Radiotherapy.

Erythema Multiforme presents with Sudden onset of rapidly progressive, symmetrical cutaneous and/or mucocutaneous lesion. Symmetrically distributed erythematous expanding macule or papule evolve into classic iris or target lesion with bright red borders and central petechiae vesicles or purpura. Lesion may coalesce and become generalised. Lesions shows Centripetal spread. Burning sensation is noticed in affected areas. Rash favors palm and soles, dorsum of the hands, and extensor surface of extremities and face. Non-specific prodomal symptom such as fever malaise myalgia, arthralgia, headache, sorethroat, cough, nausea, vomiting may appear 1-14 days before the skin lesions develop. Skin lesions have been classified as typical targets', raised atypical targets' and erythematous macules with or without blisters'. Typical targets' are defined as individual lesions less than 3 cm diameter with a regular round shape, a well-defined border, and two concentric palpable, oedematous rings, paler than the centre disc'. These lesions are most common in EM minor and milder forms of EM major in a symmetrical distribution on the extensor surfaces of the extremities. Raised atypical targets' appear similar to target lesions and are palpable erythematous lesions with a rounded shape but poorly defined borders and a dark central area, which may erode and become necrotic. These lesions are most common in severe EM major or in SJS. Flat atypical targets' as their name suggests are not palpable and they form ill-defined erythematous areas with a tendency to central blister formation. These lesions are most common in SJS. Erythematous or purpuric macules with or without blister formation' are of variable size and may become confluent. These lesions are most common in SJS and TEN. 

In case of SJS/TEN Fever is common. Skin findings may be similar to EM but are more severe. Inflammatory vesiculobullous lesion often with hemorrhage and necrosis are typical. Most commonly mouth, lips, and bulbar conjuctivae and genital mucosae. Nasopharynx, respiratory tract, GI tract, ad genitourinary tract are sometime affected. Genital involvement consists of hemorrhagic, bullous inflammation urinary retention and dysuria. Eye involvement occurs in 85% of cases. These ranges from hyperemia to extensive pseudomembrane formation.

Oral lesion appears along with skin lesion in 70% of the cases. Oral lesions start as bullae on an erythematous base, but intact bullae are rarely seen by the clinician because they break rapidly into irregular ulcers. EM lesion are larger, irregular, deeper and often bleeds. Lesion may occur anywhere on the oral mucosa but involvement of lip is prominent and gingival involvement is rare. In full blown cases lips are extensively eroded and larger portion of the oral mucosa are denuded of the epithelium. Pt. cannot eat or even swallow and drools blood tinged saliva. With in 2 or 3 days the labial lesion begins to crust. Healing
Differential Diagnosis includes Pemphigus Vulgaris, Bullous Lichen Planus, Herpes Zoster, Herpes Simplex, Drug Eruptions, Bahcet Syndrome, Urticaria and Herpetic Gingivostomatitis.

Histopathological Characteristics include a lymphocytic infiltrate at dermal-epidermal junction and around dermal blood vessels, dermal edema, epidermal keratinocyte necrosis and subepidermal bullae formation.

EM has high density of cell infiltrate rich in T-lymphocyte. by contrast SJS/TEN is characterised by a cell poor infiltrate of macrophages and dendrocyte with strong TNF-alpha immunoreactivity.  

Treatment of Erythema Multiforme depends on the severity of the clinical features. Mild forms usually heal in 2–6 weeks; local wound care, topical analgesics or anesthetics for pain control and a liquid diet are often indicated in these situations. For more severe cases, intensive management with intravenous fluid therapy may be necessary. Oral antihistamines and topical steroids may also be necessary to provide symptom relief. Systemic corticosteroids have been used successfully in some patients, but evidence to support their use for erythema multiforme is limited. HAEM is often effectively managed with acyclovir (200 mg, 5 times a day for 5 days), but only if the therapeutic scheme is started in the first few days. If erythema multiforme keeps recurring, a continuous low dose of oral acyclovir is necessary. Oral acyclovir has been shown to be effective at preventing recurrent HAEM, and the protocols may include 200–800 mg/day for 26 weeks. If acyclovir treatment fails, valacyclovir can also be prescribed (500 mg twice a day).

Complications include Hypopigmentation or hyperpigmentation, Scarring, Dehydration and electrolyte disturbances, Secondary bacterial infections/sepsis, Ocular complications, including corneal ulceration, anterior uveitis, panophthalmitis, corneal opacities, symblepharon formation, and blindness, Esophageal, bronchial, urethral, vaginal, and anal strictures (rare), Possible GI hemorrhage, renal failure, and respiratory failure in severe cases. Lip adhesion is also an unusual complication of erythema multiforme.

Most cases of EM are self-limited, with lesions evolving over 1-2 weeks and subsequently resolving within 2-3 weeks.

CONCLUSION

Erythema multiforme minor, EMM, SJS, and TEN represent a spectrum of immunologically mediated disorders that are often precipitated by infection or drug therapy. The exact pathogenic mechanisms of each disorder remain unclear. Patients can sometimes have resolution of the disease with various immunosuppressive, antimicrobial, and supportive strategies. Severe disease, however, can still lead to significant long-term morbidity and mortality. As there remains no specific diagnostic test, early clinical recognition of disease remains essential to promptly initiate appropriate treatment.

References