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Erythema Multiforme: A Rare Debilitating Condition

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ABSTRACT

Erythema multiforme (EM) is an acute mucocutaneous disease which has been associated with herpes simplex virus (HSV) infection and drugs. This report presents a case of EM minor with a prompt diagnosis and appropriate treatment. A 19-year-old male, a military recruit, reported with a chief complaint of multiple, diffuse, painful oral ulcerations for more than 3 days. An intraoral examination showed multiple large ulcers on the bilateral lateral borders and ventral surface of the tongue, bilateral buccal mucosae, and gingivae. "Target" lesion was found on the skin surface of his body or extremities. The patient was treated with corticosteroids. A follow-up oral examination revealed that all oral ulcers had completely cleared up after the 7-day corticosteroid treatment. One year after treatment, the patient remained disease-free. We concluded that EM is a mucocutaneous disease that requires a prompt and precise diagnosis. Once the disease is confirmed by the clinical presentation, it usually dramatically responds to treatment with a medium or high dose of corticosteroids. Key words: erythema multiforme, target lesion, corticosteroids.

Introduction

Erythema multiforme (EM) is an acute inflammatory disorder, usually self-limiting and often recurrent. The term EM includes a wide range of clinical presentations: a form with oral involvement only (oral EM), a mucocutaneous forms of various severity, with one or more mucosal localizations (EM minor, EM major, Stevens-Johnson syndrome [SJS]), and forms affecting large areas of the body surface (toxic epidermal necrolysis [TEN]).

It is believed to be a sequela of a cytotoxic immunologic attack on keratinocytes expressing non-self-antigens, which are primarily caused by microbes (viruses) and drugs [1,2,3].

EM usually affects apparently healthy young adults, and several reports have suggested that males are affected more frequently than females. The peak age of occurrence of EM is between 20 and 40 years, although 20% of cases occur in children. The disease is often recurrent and is precipitated by a preceding herpes simplex virus (HSV) infection in up to 70% of cases [4].

We present a case of EM minor in a 19 year old male, military recruit, who reported with severe lesions in the oral and maxillofacial region and was diagnosed and managed successfully.

Case Presentation

A 19 year old male, a military recruit, presented, with chief complaint of gradually increasing painful oral ulcerations with spontaneous bleeding. A detailed case history revealed that the symptoms appeared 3–4 days back. To start with, there was redness itching sensation and dull pain in the oral cavity and over lips, followed by ulcers and bullae. Bullae ruptured to form encrustations over lips. No history of febrile episode was present. There was no history of 2drug intake before the onset of these lesions. No other mucosal surface involvement history was present. Clinical examination revealed edema and cracking with dark brown encrustations present on the lips (Fig 1). Bleeding ulcers were present on labial mucosa, tongue and gingivae (Fig 2,3). Few hyperemic papules and macules were also present. Pharyngeal and laryngeal examination was normal. No neck nodes were palpable. A diagnostically significant finding was the presence of two target lesions on the palmar surface of left hand (Fig 4). Other systemic examination was normal.

Routine hematological investigations were within normal range. Fasting ESR was 28 mm in first hour by Westergreen method. Liver function tests revealed slightly raised transaminases. Investigations for hepatitis B and C, and HIV were negative. Clinically, diagnosis of erythema multiforme was made. The patient was treated with a medium dose of corticosteroids for 1 week, and the oral ulcerations healed (Fig 5,6,7).

Discussion

The causes of EM and its disease spectrum are numerous. Herpes simplex virus (HSV), mycoplasma pneumoniae, and drug reactions are the most common. HSV accounts for more than one half the cases of EM [5]. Other less common causes of EM include vaccinia, bacteria, fungi, drugs, irradiation from radiographic studies, malignancy (carcinomas and lymphomas), and certain collagen-vascular diseases (lupus erythematosus, dermatomyositis, and periarteritis nodosa) [6,7].

The exact pathogenesis of EM minor / EM major is unknown. It has been suggested that an immunologically mediated (i.e., lymphocytic) reaction to an infectious agent or drug leads to 3skin and mucosal lesions concentrated at the dermal-epithelial junction. In Herpes associated EM it is most likely that HSV-DNA fragments in the skin or mucosa precipitate the disease [8].

CD34+ cells transport fragments of HSV to the epithelium, and T cells accumulate in response to HSV antigens and damage cells. In contrast, drug-associated EM seems to involve CD8+ T-cell attack and expression of tumor necrosis factor alpha (TNFα) in lesional skin in the absence of HSV-DNA [9]. EM minor is considered the mildest form of EM and is characterized by

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skin lesions, which are usually symmetrically distributed on the extensor surfaces of the arms and legs. Rashes are various but typically are iris' or target' lesions or bullae on extremities. Intraoral lesions occur predominantly on the non- keratinised mucosae and are most pronounced in the anterior parts of the mouth. The lips are also commonly affected and are swollen and cracked, bleeding and crusted. Typically oral lesions progress through diffuse widespread macules to blisters and ulceration although only ulceration may be seen at presentation. In these cases, diagnosis may be difficult.[1,10]

EM major is characterised by involvement of multiple mucous membranes. Generally EM major is a more severe form of the disease than EM minor and, in addition to the oral cavity, the genital, ocular, laryngeal and oesophageal mucosae may be affected. The skin lesions may be atypical and characterised by bullae and affect a greater area [10]

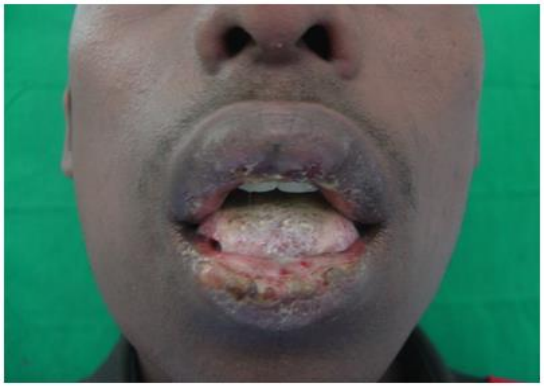

Fig 1. Clinical photo showing cracking and encrustations on the lips, tongue ulcerations

Fig 2. Clinical photo showing bleeding ulcers on the lower labial mucosa, gingiva

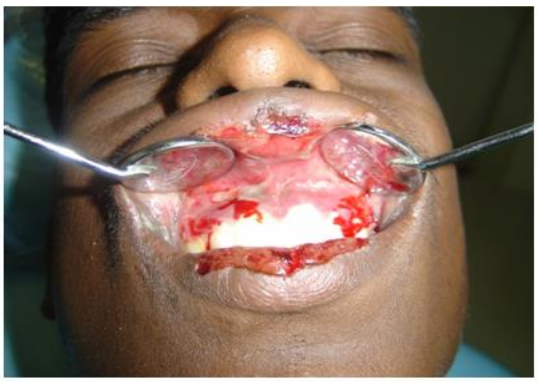

Fig 3. Clinical photo showing bleeding ulcers on the upper labial mucosa, gingiva

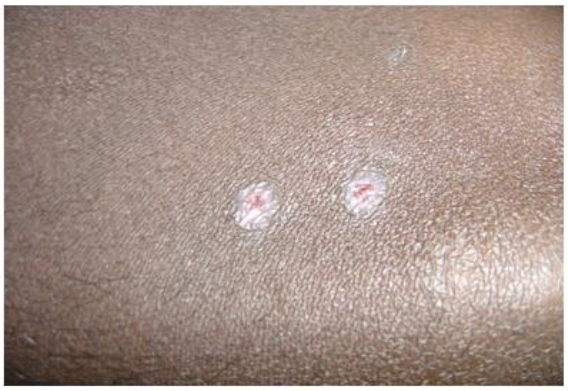

Fig 4. Target or iris lesions on skin of hand

Fig 5. Clinical photo demonstrating how the oral ulcerations had respponded after the 5 days corticosteroid treatment

Fig 6. Clinical photo demonstrating how the oral ulcerations had healed after the 1-week corticosteroid treatment

Fig 7. A visibly healthy individual after 3 months

SJS causes widespread lesions affecting the mouth, eyes, pharynx, larynx, oesophagus, skin and genitals. It almost invariably involves the oral mucosa. Ocular changes, which resemble those of mucous membrane pemphigoid – dry eyes and symblepharon may result. Balanitis, urethritis and vulval ulcers may occur and it, may be followed by sicca syndrome, or even Sjogren's syndrome [11].

Because there are serious sequelae involved in the most severe manifestations of this disease spectrum, it is important to recognize the clinical signs and symptoms of various forms of the disease so that a prompt diagnosis can be made and a proper treatment regimen can be started.

A diagnosis of EM can be difficult to readily establish, and there can be a need to differentiate from viral stomatitides, pemphigus, TEN and the sub-epithelial immune blistering disorders (pemphigoid and others). There are no specific diagnostic tests for EM and the diagnosis is mainly clinical supported if necessary by biopsy [1].

Although the use of corticosteroids for treating EM is controversial [12], it remains the main treatment modality for EM minor [13], such as in this case. The patient was treated with a medium dose of corticosteroids for 1 week, and the oral ulcerations healed without subsequent recurrence for 1 year. The patient's dramatic response to steroid treatment and the absence of recurrence for 1 year also favor a diagnosis of EM, and help rule out the possibility of pemphigus vulgaris, cicatricial pemphigoid, and oral aphthous ulcerations which are either refractory to treatment or more recurrent in nature. In addition, steroid treatment is contraindicated for HSV infection; if the patient's disease had been an HSV infection, he would not have experienced the dramatic response to the steroid treatment.

In conclusion, EM is not life threatening except in its most severe form. By recognizing the early EM oral lesions along with skin lesions if any, the dentist has a responsibility in the early diagnosis of the disease, which is of utmost prognostic importance. It usually dramatically responds to treatment with a medium or high dose of corticosteroids.

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