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Steven-Johnson Syndrome –A Rare Case Report

Medical Science

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ABSTRACT

Stevens Johnson Syndrome is a severe adverse drug reaction characterized by widespread lesions affecting the mouth, eyes, pharynx, larynx, esophagus, skin and genitals. This condition is usually associated with certain infections mainly herpes, medications and other triggers. It may be present within a wide spectrum of severity. Although, it can effect persons of any age, still a higher preedilction is seen for female gender. In severe cases, systemic corticosteroids are recommended and topical corticosteroids for the oral conditions as well. Recurrence is reported in 25 percent of cases. Here, we are presenting a case of a severe form of Erythema Multiforme also known as Stevens Johnson Syndrome.

INTRODUCTION

The term Erythema Multiforme includes a wide range of clinical expressions, from exclusive mucous membrane or skin erosions to mucocutaneous lesions (EM minor) and, in its more severe forms, there is serious involvement of mucosal membranes of more than one system and skin (EM major, Steven - Johnson syndrome) or a large area of the total body surface including mucousal surfaces (Toxic Epidermal Necrolysis) with constitutional symptoms and, at times, visceral involvement (1). Stevens Johnson Syndrome is a severe adverse drug reaction characterized by widespread lesions affecting the mouth, eyes, pharynx, larynx, esophagus, skin and genitals. It almost invariably involves the oral mucosa. The spectrum of severe cutaneous adverse drug reactions includes SJS or TEN, hypersensitivity syndrome (HSS), anaphylaxis and angioedema, serum sickness, and cutaneous vasculitis (2).

CASE SUMMARY

A 38 year old female patient reported in the Department of Periodontology and Oral Implantology, Sri Guru Ram Das Institute of Dental Sciences and Research, Sri Amritsar, with a chief complaint of pain and swollen gums in upper anterior region of the jaw since one month. She complains of bleeding from gums and pus discharge from the area. Patient complains of soreness of mouth and burning sensation on eating foods.

The patient also gave a history of recurrent vesicles and ulcerations in the lower limbs below the knees and feet, which she noticed first when she was 18 years of age. Later, few years after her marriage, she again got similar vesicular eruptions and rashes all over her body and oral mucosa; along with watering of eyes, following which she got admitted to hospital and was given medication for the same to which she responded well and was discharged later. The patient gives history of allergy to dapsone. There is no significant family history.

On clinical examination, there was erythematous, bulbous gingival enlargement seen in the upper anterior region of the jaw, which extended in the region from 23 to 13. It was tender on palpation and bleeding on probing was seen. Left central incisor (21) was mobile along with sinus tract formation with pus discharge. Multiple grossly carious teeth were present. Patient got removable partial denture in the upper anterior region few months back, which was now removed due to inflamed gums.

On cutaneous examination, lower limbs showed healing lesions which were irregularly shaped with ill defined borders. No fresh lesions were observed. A continuous watery discharge was seen from the eyes.

The medical investigations revealed that patient was anaemic with haemoglobin 9.5mg/dl, total leukocyte count of 10.800 / $mm^{3.}$ 0.9 mg/dl serum creatinine and 22.8 mg/dl blood urea. Urine examination showed few red blood cells in it.

DISCUSSION

Stevens-Johnson syndrome was first described in 1922 as an extraordinary, generalized epidermal eruption. It is accompanied by fever, inflammation of the buccal mucosa, and severe purulent conjunctivitis. Many etiologic factors have been identified in the pathogenesis of Stevens- Johnson syndrome, usually categorized as iatrogenic, infectious, or idiopathic. Iatrogenic causes are usually drug related, including antibiotics like sulphonamides, penicillins, tetracycline, erythromycin, cephalosporins, and variety of anti inflammatory drugs like aspirin and phenylbutazone.

Incidence ranges from 1.2 to 6 cases per million per year (3). In 70% of SJS cases, drugs are found to be causative agents and more than 100 such agents have been reported. Ruggiero et al., reported SJS in two children with brain tumour while receiving cranial irradiation and anticonvulsant therapy with Phenobarbital (5). Similarly, Adeloye et al. reported two cases of Stevens-Johnson syndrome in patients with penetrating head wounds who were treated with phenobarbitone (4).

Skin lesions that first appear as erythematous macules soon become edematous and papular, forming the characteristic skin lesions of Stevens- Johnson syndrome: target lesions. Target lesions are concentric rings resembling the iris of the eye. They may extend over the entire body and form bullae or vesicles within 24 to 96 hours after onset of the rash. Nikolsky's sign (separation of the epidermis from the

dermis with pressure) is usually present. When confluent, the lesions may resemble a second-degree burn. Oftentimes, sloughing of the

epidermis results. This characteristic resemblance of a burn injury often results in the application of silver sulfadiazine, which has its own complications. (6).

Erythema multiforme (EM) could easily be mixed with SJS since both present with rash and oral mucosal erosion. The typical target lesions in EM have three concentric zones: central dusky disk, middle pale ring, outermost erythematous halo and they are not found in SJS and TEN (7).

Conjunctivitis is the most frequent ocular complication, causes the eyelids to become swollen, crusted, and ulcerated, with ensuing pain and photophobia. Oral lesions, which commonly begin as vesicles, rupture, forming a gray-white membrane. The lips are often painful and crusted with blood. Many patients suffer severe dysphagia, preventing them from ingesting adequate amounts of fluids and nutrition. Oral mucous membranes are usually severely eroded for 10 to 14 days; however, prolonged ulceration of the gastrointestinal tract months after recovery has been reported (10).

SJS has to be clinically differentiated from viral stomatitides, pemphigus, EM, TEN and the sub-epithelial immune blistering disorders like pemphigoid. There are no specific diagnostic tests for SJS (8). Main differential diagnosis was toxic epidermal necrolysis (TEN), wherein the manifestations would be much more severe and over 30% of the skin surface area would be involved. Eruptions secondary to viral diseases could be ruled out due to the absence of typical prodromal symptoms. (9)

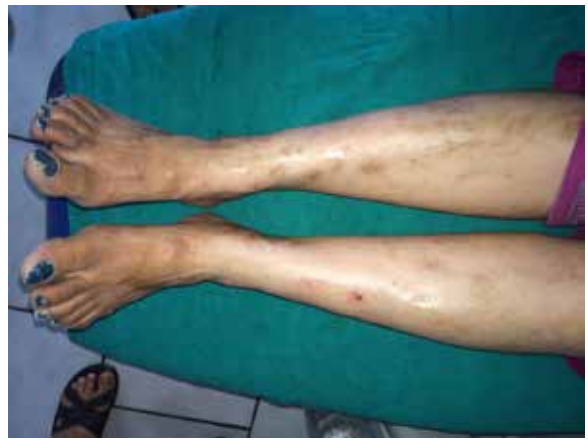
Early diagnosis with the prompt recognition and withdrawal of all potential causative drugs is essential for a favorable outcome.

Several agents have been tried for the management of this disorder. Systemic corticosteroids are used in the early stage of SJS and TEN. However, it should not be recommended when extensive skin loss has already occurred (11,12). Topical antiseptics like 0.5% silver nitrate or 0.05% chlorhexidine are usually used for skin lesions to prevent secondary infections. Antiseptic or antibiotic eye drops and eye ointments should be liberally used on ocular lesions (9). In severe cases, the patient must be transferred to burn units and measures such as environmental temperature control, careful and aseptic handling, and sterile field creation must be taken. Complications such as thromboembolism and disseminated intravascular coagulation and damage to vital organs such as the kidney deteriorate the prognosis

CONCLUSION

Stevens-Johnson syndrome is a potentially fatal disorder with a strong alliance to some medications. Dental practitioners must therefore be careful in prescribing the drugs to their patients. It is important to distinguish oral SJS for their early diagnosis, prompt management, and proper follow up.

Picture showing hypertrophied gingiva, bleeding from gums and pus discharge



Picture of lower limbs showed healing lesions which were irregularly shaped with ill defined borders, no fresh lesions were observed

Picture showing healed lesions after 1 week of oral prophylaxis but poor plaque control

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