A case of Stevens-Johnson syndrome
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Abstract
Background: Stevens-Johnson syndrome (SJS) is an acute self-limited disease. The incidence of this disease is low, but there is a significant impact on child as well as parents because of its extensive involvement of the body. This paper reports a case of SJS in an eight year old male child along with the clinical features and treatment options.

Case Presentation: A male child of 8 years was referred with the chief complaint of ulcers in the mouth since 3 days. Past medical history revealed severe drug reaction one month back, for which the patient attended a pediatric hospital elsewhere, where it was diagnosed as Steven-Johnson Syndrome and the treatment was given for the same. After one month of treatment, the child was referred to our department because of the persisting oral ulcers. On extra oral examination, skin lesions were healed whereas intra-oral examination revealed multiple painful ulcers on upper and lower labial mucosa, with accompanying painful cervical lymphadenopathy. The treatment consisted of topical application of hexigel and multivitamin supplements were prescribed. On follow up after 15 days, examination showed healed ulcers. The patient fully recovered within a month.

Conclusion: Symptomatic management of the oral lesions is necessary in order to enable the patient to have oral feeds to maintain nutritional balance. (El Med J 2:3; 2014)

Keywords: Drug Reaction, Stevens-Johnson Syndrome

Introduction
Stevens-Johnson syndrome (SJS) is named after Steven and Johnson who coined the term in 1922. SJS is a severe hypersensitive reaction that can be precipitated by infection, vaccination, systemic diseases, physical agents, foods and drugs [1]. The drugs that cause SJS commonly are antibiotics (sulfonamides), anticonvulsants (phenytoin, phenobarbital and carbamazepine), NSAIDs (oxicam derivatives) and oxide inhibitors (allopurinol) [2]. This syndrome may present as a nonspecific febrile illness (malaise, headache, cough, rhinorrhea) with oral and perioral involvement, polymorphic lesions of skin and mucous membrane characterized by acute blisters and erosions. It is a rare condition with an incidence of 0.05 to 2 persons per million populations per year [3].

Case Report
A male child of 8 years was referred to the Department of Pedodontics and Preventive Dentistry, Narayana Dental College and Hospital, Nellore with the chief complaint of ulcers in the mouth since 3 days. Past medical history revealed severe drug reaction one month back, for which the patient attended a pediatric hospital elsewhere, where it was diagnosed as SJS and the treatment was given for the same. The parents had photographs of the child taken before the treatment (Figures 1 and 2). This was the first dental visit of the patient.

The child weighed 15 kg and had average build. The child looked weak. Family history was not contributory. On extraoral examination, skin lesions were healed whereas intraoral examination revealed multiple painful ulcers on upper and lower labial mucosa (Figures 3 and 4). The ulcers were oval in shape with inflamed borders. Upper lip showed healing lesion with crust formation. Excess salivary secretion was evident and there was painful cervical lymphadenopathy. His parents explained that the child had fever one month back and had gone to local pharmacist for the medicine. However, the parents were not able to not tell the details of the medicine taken. After the child took the medicines, he developed severe itching and vesicle formation followed by ulceration overall the body, without remission from fever. His symptoms became worse including generalized weakness and toxicity. They went to a pediatric hospital where the child was diagnosed as having SJS and was treated for the condition. After one month of treatment, the child was referred to our department because of the persisting oral ulcers. The treatment consisted of topical application of hexigel and multivitamin supplements were prescribed. On follow up after 15 days, examination showed healed ulcers. The patient fully recovered within a month.

Discussion
SJS had for years been considered an extreme variant of erythema multiforme (EM), with toxic epidermal necrolysis (TEN) being a different entity. A group of experts proposed a new classification in...
which SJS was separated from the EM spectrum and added to TEN, thereby creating a new spectrum of severe drug-related diseases [4]. The criteria for diagnosis of SJS are epithelial detachment less than 10% of body surface area (BSA) and widespread erythematous or purpuric macules of flat atypical targets. SJS and TEN are severe cutaneous disorders characterized by acute skin blisters and mucous membrane erosions. In TEN, necrosis of the epidermis and other epithelia are seen. The distinguishing factor between the two is the extent of skin involvement with it being <10% in SJS and >30% for TEN.

More than 100 drugs have been associated with the development of SJS/TEN in single case reports or retrospective studies [5]. SJS 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. One of the undesirable side-effects of highly active anti-retroviral therapy (HAART) in HIV management is SJS [6]. Mycoplasma pneumonia infection has also been reported in association with SJS [7]. An association between intake of herbal drugs and onset of EM or SJS has been reported as an extremely rare occurrence [8]. A case of SJS secondary to use of diclofenac for control of post-extraction pain has been described by Shetty et al because it is uncommon [5].

In the oral cavity, SJS causes widespread ulcerative lesions. A prodrome occurs in about 30% of cases and may begin within 1 to 3 weeks of starting a new drug and lasts 1 to 2 weeks before the onset of mucocutaneous manifestations, presenting with flu-like symptoms, sore throat, headache, arthralgias, myalgias, fever, bullous and other rashes, pneumonia, nephritis or myocarditis. Ocular changes such as dry eyes and symblepharon that resemble those of mucous membrane pemphigoid may be noted. Balanitis, urethritis and vulval ulcers may occur. SJS has to be clinically differentiated from viral stomatitis, pemphigus, EM, TEN and the sub-epithelial immune blistering disorders like pemphigoid. SJS, its severe form TEN, and mucous membrane pemphigoid (MMP) are the major autoimmune causes of conjunctival scarring. The conjunctivitis varies from a papillary reaction with watery discharge to a membranous conjunctivitis with sloughing of the conjunctival epithelium. Predilection of the disease can be done with the following clinical picture of lesions on the skin mucosa junction (pluriorificialis), on hands (dorsal surfaces) and plantar surfaces of the feet.

Early diagnosis with the prompt recognition and withdrawal of all potential causative drugs is essential for a favorable outcome. Intravenous fluid replacement must be initiated immediately upon admission using saline solution. Early initiation of massive oral nutrition by nasogastric tube to minimize protein loss promotes healing and decreases the risk of stress-induced ulcers. Corticosteroids have for years been the mainstay therapy for SJS in most cases, as in our case. They suppress the intensity of reaction, control the extension of the necrotic process, decrease the involved area, reduce fever and discomfort, and prevent damage to internal organs when given at an early stage and at a sufficiently high dosage. Topical antiseptics like 0.5% silver nitrate or 0.05% chlorhexidine are usually used for skin lesions to prevent secondary infections. Complications such as thromboembolism and disseminated intravascular coagulation and damage to vital organs such as the kidney deteriorate the prognosis.

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Conclusion
Symptomatic management of the oral lesions is necessary in order to enable the patient to have oral feeds to maintain nutritional balance.

Competing interests: The authors declare that no competing interests exist.
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