Case Report ISSN (O):2395-2822; ISSN (P):2395-2814

Endodontic Management of a Patient with Steven Johnson Syndrome – A Case Report.

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Received: October 2018
Accepted: October 2018

ABSTRACT

Many recent drug therapies have evolved for the management of postoperative pain and infection. Although adverse reactions are rare it still remains a great threat to the patient. Stevens-Johnson syndrome(SJS) is one such severe hypersensitive reaction which is mostly drug induced and characterized by febrile illness, polymorphic lesions of skin and mucous membrane, acute blisters or erosions.. It can be precipitated by infection, vaccination, systemic diseases, physical agents, foods and drugs. In SJS Involvement of oral and/or mucous membranes may be so severe that the patient may be unable to eat or drink. Mucosal involvement may include erythema, edema, sloughing, blistering, ulceration, and necrosis .This paper will focus on endodontic management in a patient with the past history of Steven Johnson syndrome and the necessary precautions to be taken during the treatment and management of such patients.

Keywords: Stevens Johnson syndrome, sulpha drugs, adverse drug reaction.

INTRODUCTION

Steven Johnson syndrome was first recognised by Albert Mason Stevens and Frank Chambliss Johnson, the American pediatricians who first published about this syndrome in 1922.[1] It is a severe form of Erythema Multiforme and a disorder of the immune system. It begins with the appearance of fever, malaise, photophobia and also eruptions on oral mucosa, genital mucosa and skin.[2] Steven Johnson syndrome is a rare condition with incidence of around 2.6 to 6.1permillion people per year.[3] SJS and TEN are the two forms of skin diseases that are life threatening. The characteristic difference between these two is based on the type of lesion present and the amount of body surface area involved.[4] Criteria for diagnosis is epithelial detachment which is found to be less than 10% of body surface area (BSA) in SJS and more than 30% of BSA detachment is TEN.Steven Johnson syndrome can be precipitated by infection, vaccination, systemic diseases, physical agents, foods and drugs.[5] More than 100 drugs are Associate with SJS but drugs that are commonly associated with it are anibacterials, sulfonamides, anticonvulsants, NSAIDS and oxide inhibitors.[5] The field of severe drug reactions includes steven johnson syndrome or TEN, hypersensitivity syndrome (HSS), anaphylaxis and angioedema, serum sickness and cutaneous vasculitis.[6]

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The incidence of Steven Johnson syndrome is found to be increased in immunocompromoised and HIV infected population.[7] Sepsis from widespread skin infection, respiratory tract infection renal failure and cardiac complications can lead to death. Reexposure to the same precipitating factor can aggravate SJS with more severe clinical symptoms.[7]

As illustrated, the immune response may be triggered by the binding of an antigenic drug (e.g. carbamazepine [CBZ]), to a specific HLA allele (e.g. HLAB*1502) on a keratinocyte, which are the main antigen presenting cells (APC) in SJS/TEN. Then, specific T cell receptors (TCR) of theCD8+ cytotoxic T lymphocytes (CTLs) recognize the drug-HLAcomplex. Upon the activation, CTLs or NKT cells produce cytokinesand chemokines, as well as the cytotoxic proteins, particularly, secretorygranulysin and lead to extensive keratinocyte apoptosis.[8]

Histopathology

SJS shows widespread necrosis of the epidermis and little vascular inflammation of dermis.[7] Cell death occurs which causes epidermis to separate from dermis. Histopathologically most of the disease is located in the epidermis as this is the site

where drug and its metabolite are bound with less inflammation in the dermis as found in Epidermal necrolysis. The main cytokine involved is tumor necrosis factor (TNF) – α.[9]

Genetics and Signals in SJS_TEN

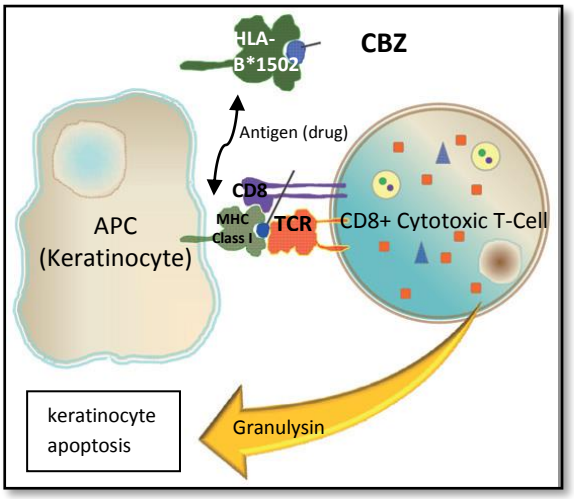
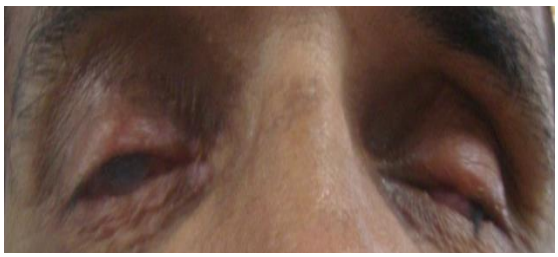


Figure 1: A model of keratinocyte apoptosis induced by the immunesynapse of drug-HLA-TCR interaction in SJS/TEN

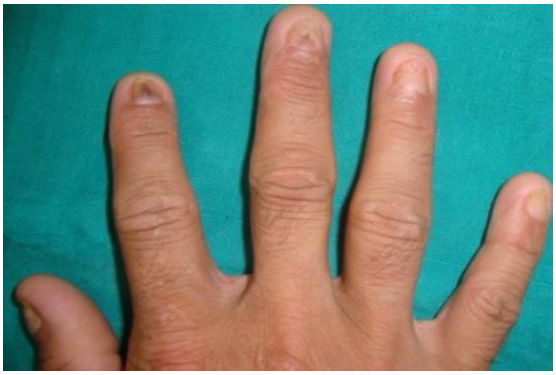
CASE REPORT

The present case report is about a 40 year old male patient who reported with a chief complaint of pain and sensitivity in the upper front region of the jaw and poor esthetics. Dental examination revealed poor oral hygiene, pain and tenderness with 22 and deep carious lesion with 15, 21,34,35.His past medical history revealed SJS that was precipitated due to the drug sulphonamide (septran), diagnosed at the age of 12 yrs with no family history of the same.

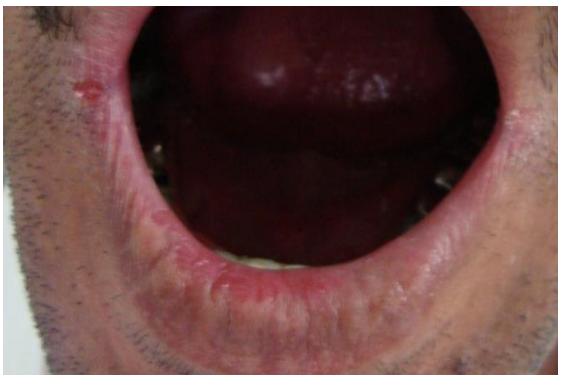
On general examination, the vital signs were normal and the other findings of the patient revealed poor eye sight, clubbed fingers of both hands and feet, xerostomia and angular chelitis. Clinical diagnosis was grossly carious 22 and irreversible pulpitis with 15,21,34,35. The patient was advised extraction with 22 and non surgical endodontic treatment with the other teeth.



Poor Eye sight



Clubbed fingers



Angular Chelitis

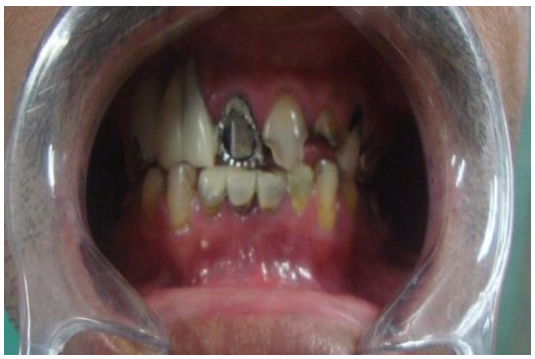
Patients with SJS are found to be very sensitive and more prone to secondary infections, so the patient was pre medicated. As the patient had an history of allergy to Sulpha drugs, a consent was obtained from the patients family physician and also a written consent was signed from the patient and the treatment was planned. He was prescribed antibiotics and analgesics i.e (Amoxycyllin and Ketorol-DT). Glycaemic controlcan be a problem in such patients as a result of stress and previous treatment that they had undergone with systemic steroids. And as hyperglycaemia is one of the risk factors for poor outcome, close control of blood glucose should be maintained. So patient was advised to undergo routine blood investigations to rule out any discrepancies. Blood pressure was also monitored and was within the normal limits.

Treatment

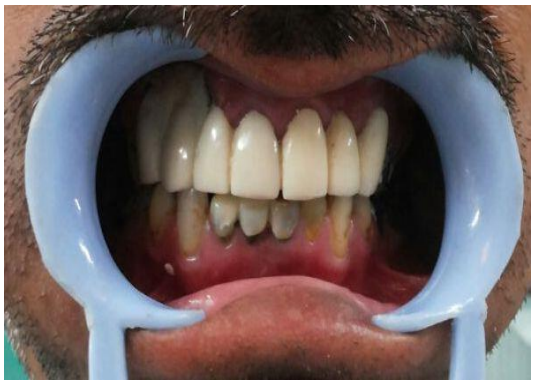
Present case was not an acute phase of SJS but revealed a past history of it. So the focus lies on the considerations taken during the treatment under total aseptic conditions to avoid its recurrence. Care was taken to complete the treatment in minimum visits so as to avoid the chances of any nosocomial infection. Strict sterilization was maintained during the treatment. In between the treatment the vital signs were monitored for hypotension , tachycardia etc. Patient was addressed to get the tetanus prophylaxis done. Pre operative radiographs were obtained and after detailed clinical and radiographic examination the treatment was carried

out.LA with Adrenaline was administered under his physicians consent. Root canal treatment was done with 15,21,34,35. During the treatment the teeth were isolated with rubber dam and access preparation was done. Working length was determined and cleaning and shaping was done. In between the instrumentation the canals were irrigated with17% EDTA and 5.25% Sodium hypochlorite solution. 2% chlorhexidine was used as a final irrigant. Finally the canals were obturated using AH plus sealer and Guttapercha.

Patient was recalled after one week and was found asymptomatic. Patient was advised all ceramic bridge but he refused owing to the financial problem so, porcelain fused to metal crowns were given in the anterior region. Extraction was done with 22.



Pre –operative



Post - operative

DISCUSSION

There are no standard guidelines found for the treatment of SJS. Discontinuation of the offending drug is a priorityand mostly the patients are treated symptomatically.[7] Patients with Steven Johnson Syndrome needs to undergo dressings, fluid replacement and antibiotic therapy so there management can be best done in burn wards.[7] Many a times it is difficult to choose an antibiotic for such patient as no reliable test exists to establish a link between a particular drug and SJS for any individual case.[3] Skin lesions in SJS shows positive pseudo- Nikolskys sign (epidermal peeling sign) which is elicited by applying thumb or lateral pressure on the skin over bony prominences.[10] Such patients have considerable fluid loss and often present with dehydration and renal impairment. Careful monitoring of fluid balance with strict input and output charts is essential.[11] The mortality rate of SJS is around 5% and the death risk can be estimated using SCORTEN scale.[12]

In the present case it was found that at the age of 12 years, patient suffered from upper respiratory infection and fever for which he was prescribed the drug Septran by general physician. Slowly he started developing oral thrush and rash on the body, but it was misdiagnosed as some type of viral infection. It progressed further resulting in loss of vision and was then diagnosed as Steven Johnson syndrome; the reason being patients allergy to Sulpha drugs.

Xerostomia is one of the common finding in patients with SJS.[13] It causes constant sore throat, burning sensation, difficulty in speaking and swallowing, hoarseness and/or dry nasal passages. It can even lead to gum disease and tooth loss. If left untreated, xerostomia decreases the oral pH and significantly increases the development of plaque and dental caries.[14] In this case this was one of the reason for patients recurrent oral infections and caries. So the patient was advised to use fluoridated tooth paste and also advised high-fluoride varnishonce every 1 to 3 months to reduce the susceptibility of enamel to acid challenges. Products containing sodium lauryl sulfate should be avoided as they may contribute to the formation of aphthous ulcers or canker sores.

Patient was blind by one eye and very poor eye sight of the other one. He was going to undergo Osteo-odonto-keratoprosthesisa eye surgery. It is a technique used to replace damaged cornea where corneal transplantation is a failure. This surgery requires patients own tooth root and the alveolar bone to support an apical cylinder. It is mainly indicated in patients with blindness due to severe end stage Steven johnson syndrome, chemical burns, pemphigoid etc.[15] As many of the tooth were already endodontically treated and very few vital teeth were remaining, so the patient was encouraged to conduct a routine mouth examination checking for red, white or dark patches, ulcers or tooth decay. Patient was treated with special attention to airway and hemodynamic stability and pain control.

Avoid use of silver sulfadiazine one of the commonly used antiseptic, instead 0.5% silver nitrate or 0.05% chlorhexidine can be used to clean wounds.[16] Although Steven–Johnson Syndrome can be caused by viral infections and malignancies, main cause is medications and the leading cause appears to be the use of antibiotics particularly Sulpha drugs.[17] NSAIDS are most commonly prescribed in routine dental practice. Adverse

reactions to these drugs are rare but still cases are reported of SJS even due to the use of Diclofenac Sodium.[7] In this case patient was prescribed with Ketorol- DT which is also a commonly used analgesic in dental pain. It is an NSAID heterocyclic acetic acid derivate and allergic reactions (anaphylactoid reactions, asthma, bronchospasm, Stevens–Johnson syndrome, toxic epidermal necrolysis) have been reported with its use.[18] So proper history and care is needed while prescribing drugs to the patients with SJS.

CONCLUSION

Steven Johnson Syndrome needs to be handled with strict sterilization and with proper care and special attention while prescribing the drugs. If an acute case reports to clinic the patient must be hospitalized to avoid any other complications.

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How to cite this article: Dhote S, Gunwal M, Shori D, Shenoi P. Endodontic Management of a Patient with Steven Johnson Syndrome – A Case Report. Ann. Int. Med. Den. Res. 2018; 4(6): DE20-DE23.

Source of Support: Nil, **Conflict of Interest:** None declared