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Sequelae of a Treated Stevens Johnson Syndrome- A Case Report

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Abstract:- Stevens Johnson syndrome (SJS), a life threatening mucocutaneous disease can present with many variations and rapidly worsen in a short period of time. Various etiologies are associated with these conditions, with adverse drug reaction being the most common. While the reaction itself is self-limited, little is known about the frequency and risk factors associated with long term SJS sequelae. Presenting a case of a 40 years old male patient who presented to our dental OPD with the chief complaint of burning sensation in the mouth with complete loss of eyesight, all these resultant due to treatment of Stevens Johnson Syndrome. Therapeutic management was done after proper investigations. This case emphasizes the importance of recognition of long term sequelae of Stevens Johnson syndrome as this is a rare but serious condition that must be diagnosed and treated appropriately. Clinicians should be aware of the potential for long term complications among Stevens Johnson survivors.

Keywords:- *Stevens Johnson Syndrome, Drug Reaction, Burning Sensation, Long Term Sequelae.*

I. INTRODUCTION

SJS (Stevens Johnson Syndrome) lies on a spectrum of immune-mediated mucocutaneous diseases which can present with many clinical variations and rapidly worsen in a short period of time. Various etiologies are associated with these conditions, with adverse drug reaction being the most common. It presents as a severe exfoliative reaction affecting mainly the skin and mucous membranes with significant morbidity and mortality. Since SJS not only limits to the skin and oral mucosa, a host of consequences can occur and as a result survivors are often left with debilitating multi-organ complications. While the reaction itself is self-limited, little is known about the frequency and risk factors associated with long term SJS sequelae. Stevens-Johnson syndrome is estimated to affect two to seven per million people each year. Although it is more common in older people and women, it can affect individual with genetic predisposition irrespective of age, sex and races. [1] More than 50% of SJS survivors suffer from long-term sequelae of the disease. SJS does pose a significant risk of mortality as high as 5% [2]

II. CASE PRESENTATION

A 40 years old male patient presented to our dental OPD with the chief complaint of burning sensation in the mouth which aggravated on taking spicy food. Patient also had complaint of difficulty in swallowing solid food. He had a history of chemical injury (Mercury and other compounds) spilled all over the body including the eyes 23 backs in the year 1999, where he experienced blurry vision at that time. Since then he started losing his eyesight gradually. Patient visited AIIMS, New Delhi in 2017 for the same where the case was diagnosed as both eye SJS sequelae with severe dry eye with right conjuctivalisation of cornea with symblepharon upper lid and lower lid with entropion upper lid with trichiasis and left keratinization of cornea with recurrent symblepharon. Patient was advised for Right upper lid entropion correction/ symplepharon release with mucosal membrane graft. Surgery was planned as follows: 1st stage : Lid margin reconstruction, 2nd stage: Occular surface rehabilitation and 3rd stage: Keratoprosthesis/ Penetrating keratoplasty. Mucosal membrane graft was taken from Mandibular labial mucosa of lower lip. However, no intervention was done in left eye. Despite all these surgical procedures, he completely lost his eye sight..

The patient had no deleterious habit and his personal history was normal. General condition of patient was fair with pallor present. All vitals were normal. Koilonychia was present wrt hands and legs. Entire oral mucosa appeared

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shiny. Area of ulceration, bleeding and erosion was present wrt labial and buccal mucosa. Tongue appeared depapillated along with pain on swallowing. Entire oral mucosa was found to be tender on palpation. Scrappable non tender curdy white patch present wrt dorsal surface of tongue. Saliva was found to be thick and frothy.

With the intraoral findings, we came up with the differential diagnosis of Diabetic stomatitis and Anaemic stomatitis along with Oral candidiasis on dorsal surface of tongue. CBC and Fasting blood sugar was advised. Patient was found to be slightly Anaemic and uncontrolled Diabetic. Patient was advised for physician consultation for high blood sugar level. Initial management of patient was undertaken with topical antifungal, antiseptic and mouthwash as salivary substitute along with Lycopene, neurobion forte and probiotics. Patient was reviewed after 7 days. Blood investigation repeated after 7 days showed improvement in blood sugar level after intake of anti Diabetic drugs Sitagliptan and Metformin Hydrochloride (550 mg) as prescribed by physician. Oral mucosa showed discernible changes towards betterment with improvement in oral symptoms. Patient was advised for continuation of same medication and periodic follow up.

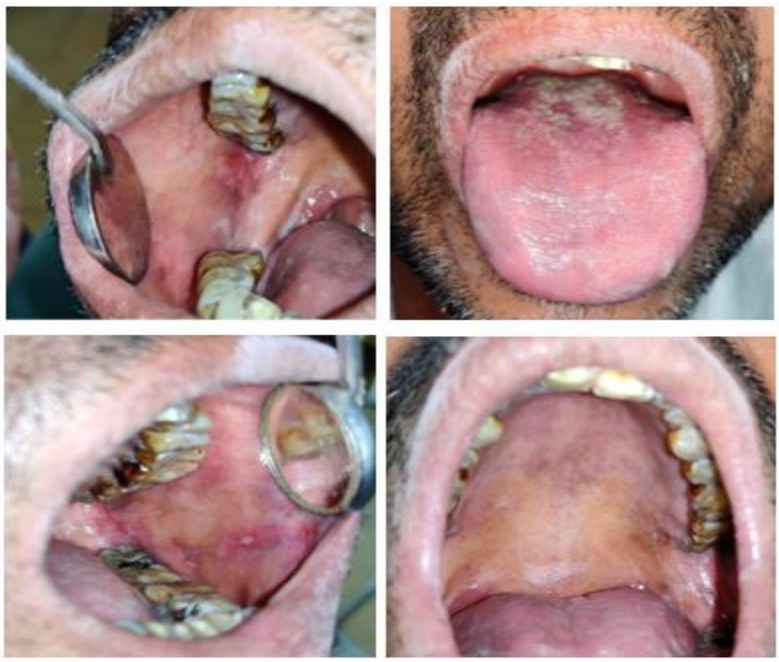

Fig 1 Pre treatment intraoral pictures

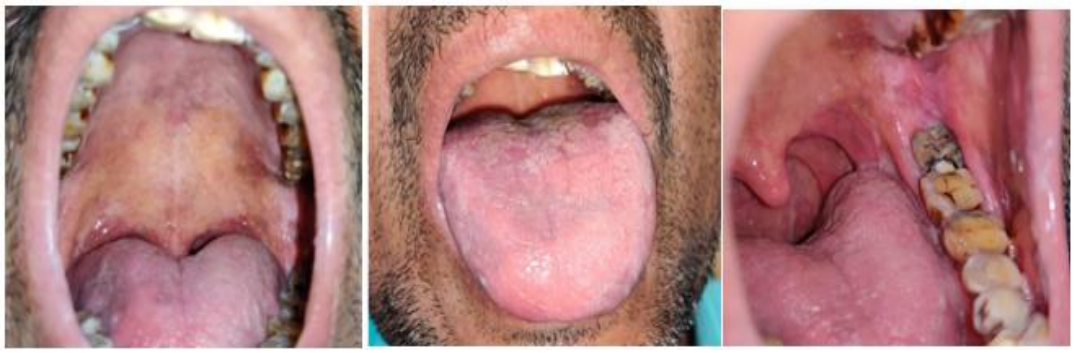

Fig 2 Post treatment intraoral pictures

III. DISCUSSION

This patient, who had SJS episode 23 years back, had visited doctors on and off for various reasons but developed oral stomatitis and glossitis because of variable reasons. At first, he had multiple surgical interventions, second he was on Steroid therapy and most importantly at present he had metabolic and Anaemic consequences. When presented to us, he is already Diabetic, which resulted in Xerostomia, burning sensation and opportunistic infections in mouth. So the patient is required to correct his Xerostomia, Anemia and systemic Diabetes Mellitus. This paper emphasizes the importance of thorough case history and proper examination for successful management of vivid hisory. It is important to recognize long term sequelae of Stevens Johnson syndrome as this is a rare but severe life threatening condition that requires prompt diagnosis and appropriate treatment.

In the acute stage, oral mucosal involvement is seen in up to 100% of SJS patients, which occurs in the form of mucositis and ulceration. 40% of survivors is reported to have dry mouth. In addition to this, saliva of SJS survivors tends to

be more acidic and reduced in quantity with abnormal viscosity which may lead to dental caries, gingival inflammation, and periodontitis. Other oral manifestations include depapillation of the tongue.[3] Manifestations of SJS outside of the skin, eyes, and oral mucosa are often not addressed clinically since they are not well recognized**.** Despite the highest involvement of the eyes and skin, multiple organ systems, such as pulmonary, gastrointestinal/hepatic, oral, otorhinolaryngologic, gynecologic, genitourinary, and renal systems can also be affected.[4] A multidisciplinary team must be integrated to assess organ damage and mitigate the chronic sequelae.

IV. CONCLUSION

SJS is still an ongoing challenge for clinician as it presents with symptoms that could mimic a plethora of conditions and much more challenging for dental practitioners due to the rarity of presentations and dental orientation. A thorough medical history and proper physical examination along with oral health assessment is crucial to facilitate early diagnosis and prompt treatment for prevention of serious complications.

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