Case report

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A Case Report of Steven Johnson Syndrome

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ABSTRACT

Introduction: Mucosal involvement and extensive epidermal detachment are hallmarks of the uncommon but severe mucocutaneous reaction known as Stevens-Johnson Syndrome (SJS). SJS is triggered primarily by medications or infections. It presents a significant clinical challenge due to its rapid onset and potential for life-threatening complications. T lymphocyte-mediated drug hypersensitivity reactions are a key component of the pathophysiology, which is a complex interaction of immunological systems, genetic predisposition, and environmental variables. A painful rash that appears suddenly and causes extensive mucosal lesions are the typical clinical signs, which precede prodromal symptoms. The mitigation of systemic consequences through early recognition, quick withdrawal of the offending agent, and supportive care are essential components of management. Increased rates of morbidity and mortality are still linked to SJS, even with treatment advances.

Case presentation: A case of a 65-year-old female patient with a known case of Type 2 Diabetes mellites presented to the hospital with an abrupt onset of fever, rashes all over the body, photophobia, conjunctival itching, burning, eyelids, swollen lips and dysphagia. The reaction was evoked after the intake of the Tablet Phenytoin. She was treated in the ICU with general measures along with corticosteroids and antibiotics. Here, we describe a very rare case where phenytoin is implicated in causing SJS.

Conclusions: Stevens-Johnson syndrome (SJS) is an immune-mediated disease characterized by a prodromal illness followed by severe mucocutaneous symptoms. Early detection, timely intervention, and multidisciplinary management can be done to reduce the effects of this potentially fatal syndrome.

Keywords: *Case, Report, Syndrome*

INTRODUCTION

Mucocutaneous lesions and systemic involvement are hallmarks of the rare but severe dermatological disorder known as Steven-Johnson Syndrome (SJS). Since its first description by Stevens and Johnson in 1922, SJS has presented several clinical practice challenges because of its diverse aetiology, fluctuating appearances, and propensity for rapid worsening.[1] This disease spectrum with high morbidity and mortality; previous studies have reported mortality rates for SJS to be around 19.4 % to 29%.[2]

Stevens-Johnson syndrome/toxic epidermal necrolysis is a rare and unpredictable reaction to medication that involves drug-specific CD8+ cytotoxic lymphocytes, granule-mediated exocytosis and tumor necrosis factor-alfa (TNF–alpha)/ death receptor pathway.[3] Genetic factors include human leukocyte antigen (*HLA*) allotypes that lead to an increased risk of SJS when exposed to anticonvulsants and allopurinol.[4] Multiple drugs, infections, and underlying medical problems have all been identified as possible triggers for the multifactorial etiology of SJS. Notably, a large body of research has been done in the literature about the correlation between certain medications, specifically sulfonamide antibiotics, anticonvulsants, and nonsteroidal anti-inflammatory medicines (NSAIDs), and the onset of SJS.[5]

The illness begins with nonspecific symptoms such as fever and malaise, and upper respiratory tract symptoms such as a cough, rhinitis, sore eyes, and myalgia. Over the next three to four days, a blistering rash and erosions appear on the face, trunk, limbs, and mucosal surfaces. Mucosal ulceration and erosions can involve lips, mouth, pharynx,

oesophagus and gastrointestinal tract, eyes, genitals, and upper respiratory tract. About half of patients have involvement of three mucosal sites.[6]

There are no specific blood tests yet to diagnose SJS/TEN. The diagnosis is based upon identification of characteristic signs and symptoms, a detailed history from the patient and/or relatives, a thorough physical examination and the results of blood tests and a skin biopsy. An additional skin biopsy for immunofluorescence may also be performed to exclude other conditions such as autoimmune blistering disorders which may have similar manifestations to SJS.[7]

Early recognition and prompt cessation of the offending agent are critical in managing SJS to halt disease progression and reduce morbidity and mortality. Supportive care plays a pivotal role, encompassing meticulous wound care, fluid and electrolyte management, nutritional support, and vigilant monitoring for complications such as infection and organ dysfunction.[8] Despite aggressive interventions, SJS can lead to severe complications, including sepsis, respiratory compromise, and long-term sequelae such as scarring and ocular complications, emphasizing the importance of a multidisciplinary approach to management.[9]

The most common long-term complications of Stevens-Johnson syndrome/toxic epidermal necrolysis are ocular (including blindness), cutaneous (pigmentary changes and scarring), and renal. Mucosal involvement with blisters and erosions can lead to strictures and scarring.[10] We report the case of Stevens-Johnson syndrome brought on by phenytoin.

This study aims to highlight the significance of early detection, timely intervention, and multidisciplinary management in reducing the effects of this potentially fatal syndrome by presenting a recent case of SJS. Moreover, its objective is to augment the current corpus of literature about SJS, consequently augmenting our comprehension of its aetiology, clinical trajectory, and remedial approaches. Similarly, the study highlights phenytoin as the triggering factor for SJS which is one of the rare and serious adverse reactions because phenytoin drug-induced hypersensitivity SJS is uncommon.

CASE REPORT

The case is a 65-year-old female patient who visited to the emergency department with an abrupt onset of fever, painful rashes along with blisters all over the body, and dysphagia on the day of admission. Her daughter-in-law mistakenly gave her a Tablet Phenytoin instead of a Tablet Metformin because of ignorance after which she suddenly, developed bumps starting from the trunk and spreading all over the body. Her eyelids and lip were swollen. Later, the patient had blisters and crusts. Thus, she was taken to one of Hospital of Raxual, India. Further, she was referred from there to one of the hospital of Parsa on dated 2080/12/23 after which she was brought- to the emergency unit and then shifted to ICU of the hospital.

The patient has a known history of DM for one year and was under Tablet Metformin and Glimepiride.The patient had no prior surgeries, no previous hospitalization, and no known allergies. The patient denied using alcohol, tobacco, or any illicit substances. She denied any history of anemia, easy bruising or bleeding, yellowing of skin, rashes, and a prior history of blood transfusions.

On examination, her blood pressure was 120/70 mmHg, heart rate was 77 beats per minute, respiratory rate was 20 breaths per minute, the temperature was 36.9°C (98.4°F), weight was 51 kg, height was 165 cm and body mass index was 20.56 kg/m^2. She was alert and oriented to time, place, and person and looked sad, tired, and anxious.

The examination revealed blisters and crusts all over the skin and mucous membranes of the mouth and lips. Further signs of early mucosal involvement were present like photophobia, conjunctival itching, burning, eyelids, and lip were swollen and dysphagia. Another finding was presence of myalgia with target lesion i.e. blistering rash and erosions.

Subsequently, skin lesions were on the face/thorax and are symmetric, ill-defined, coalescing, erythematous macules with purpuric centers. The skin was tender to touch, with pain out of proportion to findings (pain felt by the patient is much stronger than what the visible symptoms suggest indicating severe inflammation or damage to the skin).These macules progress to vesicles/bullae, which were of sloughing off with slight pressure. The scalp, palms, and soles of feet are usually spared but maybe the early site for lesions.

There was no evidence of pneumonia on chest x-ray examination. Results of hematologic and biochemical evaluation were as follows; white cells:13, 500/ in mm^3, packed cell volume of 33.4%, Haemoglobin: 11 gm/dl, lymphocyte: 20%, MCHC:31.2%, Random blood glucose: 270 mg/dl, urea: 21 mg/dl, creatinine: 0.82 mg/dl. Biopsy and culture were not done.

It was suspected Stevens-Johnson syndrome because of the clinical findings as there are no formal diagnostic criteria for SJS/TEN. Clinical features started in a patient

as a febrile illness with malaise, headache, cough, and/or rhinorrhea 1-3 days before skin lesions appear. Signs of early mucosal involvement include photophobia, conjunctival itching, burning, and dysphagia. Other features include myalgia and/or arthralgia with target lesions.

The patient had a similar nature of clinical features suggestive of Stevens-Johnson syndrome. This evidence has pointed out SJS, therefore the diagnosis was drawn based on findings. The patient was admitted to the ICU from Emergency where she received Inj. Meropenem 1gm IV TDS, Inj. Dexona 8mg OD, Cap. Tortiplex -Z PO BD, Inj. Regular Insulin as per sliding scale, Gel. Oroheal TDS (30 minutes after a meal) and Gel Zytee TDS (30 minutes before a meal). The patient's clinical symptoms improved after 9 days of ICU stay with conservative management. She was discharged with on therapeutic regimen of tab. Glycomet 500 mg BD, Tab. Linamet 5gm OD, Tab. Glycosome 40mg PO OD for 5 days followed by 35 mg PO OD for 3 days further 30,25,20 and 15 mg PO OD for subsequent 3/3/3/3 days and tab. cefixime 200mg PO BD, Tab. Peptard 20mg PO OD and Gel Oroheal BD. She was instructed to follow up as an outpatient for further workup after 2 weeks.

Data was collected using multiple methods such as individual interviews with the patient and patient attendant, analysis of patient records and documents, and observations of physical findings. Ethical considerations were ensured by obtaining informed consent, ensuring anonymity and confidentiality, and avoiding harm to the patient.

DISCUSSION

Steven Johnson Syndrome (SJS) and toxic epidermal necrosis are acute, self-limited diseases, rare but life-threatening adverse drug reactions. Anti-epileptic SJS is a severe cutaneous adverse reaction and are the major cause of the condition. In this case, the cause of SJS was use of anti-convulsant (Phenytoin).[11]

SJS is a life-threatening clinical condition that is usually manifested with fever, red rashes, blisters formation, oral and ocular lesion, genital and anal lesions. In present case the initial sign and symptoms were fever, rashes all over the body and dysphagia. Later the clinical features were associated with blisters and crust all over the skin and mucous membrane of mouth and lips, photophobia, conjunctival burning and itching.[12]

Stevens-Johnson Syndrome was suspected for the patient of the current study based on the clinical findings. Prior literature reported that SJS can be diagnosed on the basis of clinical features of the patient and routine blood investigations, i.e. Complete Blood Count, C -Reactive Protein, Renal Function Test and Chest x-ray.[12]

Hematologic and biochemical evaluation of the patient of the current report revealed that elevated white blood cells, count with normal packed cell volume, decrease haemoglobin, increase in random blood glucose level with normal renal function test. There was no evidence of pneumonia on chest x-ray examination. Prior to the previous case report revealed elevation in cardiac biomarkers. Echocardiogram revealed global hypokinesia with elevated Prothrombin time and activated partial thromboplastin time and decrease in platelet count. Results of liver function tests were normal.[13] (myocarditis) Likewise, contradict to the current case report, a case report also revealed that there was normal glucose level in patient with phenytoin induced SJS. [14] (phenytoin induced).

Regarding the management of SJS, the patient was treated based on the signs and symptoms and supportive care were provided in the Intensive Care Unit of Narayani Hospital. Corticosteroids and antibiotics therapy was administered i.e. Inj. Dexona 8 mg IV OD and Inj. Meropenem 1 gm IV TDS were administered along with mouth gel Oroheal in the affected areas TDS. Strict aseptic precautions were used and vital signs were monitored hourly with strict intake and output monitoring. The patient was discharged on the 9th day of admission with a therapeutic regimen of Tab. Glycomet, Tab. Linamet, Tab. Glycosono, Tab. Cefixime and Gel Oroheal and instructed for follow up after 2 weeks. The current management was aligned with the prior evidence.[12]

This evidence indicated the hourly monitoring of vital signs, maintenance of fluid volume, insertion of catheterization for output, NG tube for intake, Transfusion of Blood as per need, application of anti-septic solutions on affected areas, prescription of corticosteroids and anti-biotics as therapeutic management and strict aseptic precautions along with monitoring of drugs adverse reaction.[12] This case study highlights phenytoin as the triggering factor for SJS which is one of the rare and serious adverse reaction because phenytoin drug induced hypersensitivity SJS is uncommon.

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