



## AN INTERESTING CASE OF RECURRENT STEVENS JOHNSON SYNDROME

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### ABSTRACT

Stevens Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are severe cutaneous adverse reactions to drugs characterized by extensive detachment of the epidermis and erosions of the mucous membranes. Here we report a case of Stevens Johnson syndrome with toxic epidermal necrolysis overlap. This case report is to highlight the importance of early diagnosis and necessity for continued avoidance of the inciting drug. In our patient the lesions recurred when the inciting drug was reinstated into the prescription inadvertently. The recurrent lesions were very similar to the initial episode and the patient

responded very well to intravenous immunoglobulins and steroid combination therapy during both episodes.

**Keywords:** Recurrent Stevens Johnson syndrome (SJS), toxic epidermal necrolysis (TEN), Adverse drug reactions, intravenous immunoglobulins, glucocorticoids.

**CASE REPORT**

A 64 year old male patient a known diabetic and hypertensive presented to the hospital with the complaints of blisters all over the body since 2 days. He recently had seizures and was started on phenytoin. Patient was on regular treatment with metformin, diuretics, beta-blockers, atorvastatin and clopidogrel for diabetes mellitus, hypertension and ischemic heart disease respectively.

On examination, patient was hemodynamically stable and systemic examination revealed crusted plaques, erosions and maculopapular rashes on the chest, back, scrotum, upper and lower limbs (Figure 1,2).



**Figure 1- Diffuse erythema and peeling of skin on the anterior chest wall and cheeks.**



**Figure 2- Diffuse erythema and peeling of skin on the posterior chest wall.**

A few bullae and erosions were seen in the gluteal region. Erosions were also seen on the buccal mucosa. His complete blood count, renal function tests, liver function tests, serum electrolytes and blood sugars were normal. MRI brain was normal. Investigations done for malaria, enteric fever, dengue, rickettsiae and leptospirosis were negative. Blood and urine cultures were sterile. A clinical diagnosis of Stevens Johnson syndrome/toxic epidermal necrolysis overlap was made based on the percentage of skin involvement which was 25% in our case.

After ruling out the other causes of rash, since the lesions were severe patient was treated with intravenous antibiotics (cefaperazone 2g Intravenous twice a day), steroids (dexamethasone 12mg Intravenous once a day) and intravenous Immunoglobulins (2 gm/kg bodyweight; total dose-100 gm in five divided doses over a period of five days). Supportive care was given in the form of topical antibiotics and saline soakings. Other co morbidities were managed accordingly. Wound cultures were negative. After a prompt recovery he was discharged and was asked to stop phenytoin. The antiepileptic phenytoin was changed to carbamazepine and levetiracetam.

Patient was readmitted once again after a period of 2 weeks with similar rashes. Even though it was advised to the patient to stop phenytoin it was found that patient had lost the prescription and about a week later patient developed seizures for which he consulted a local physician who restarted phenytoin as the patient did not inform him about the previous episode of reaction to phenytoin. The patient developed similar cutaneous manifestations that he had previously and was referred back to our hospital. Phenytoin was stopped and he was started on carbamazepine and levetiracetam along with intravenous steroids and immunoglobulins. He responded well and recovered completely again.

## DISCUSSION

Stevens Johnson syndrome and toxic epidermal necrolysis are severe mucocutaneous reactions most commonly triggered by medications and characterized by extensive necrosis and detachment of epidermis. When the pathology involves less than 10% of the body surface area it is called as Stevens–Johnson syndrome. If more than 30% of the body surface area is involved it is called as toxic epidermal necrolysis. It is called as Stevens–Johnson syndrome/toxic epidermal necrolysis overlap, when the involvement is between 10-30%.<sup>[1]</sup>

SJS –TEN is essentially drug induced. Graft versus host disease is another well established cause independent of drugs.<sup>[2]</sup> The differential diagnosis includes autoimmune blistering diseases, paraneoplastic pemphigus, pemphigus vulgaris, bullous pemphigoid, acute generalized exanthematous pustulosis (AGEP), disseminated fixed bullous drug eruption and staphylococcal scalded skin syndrome (SSSS).

Common triggers include anticonvulsants, sulphonamides and oxycam non steroidal anti-inflammatory drugs with drug specific incidences ranging from 1 in 10000 to 1 in 100,000 new cases.<sup>[3, 4, 5]</sup> The pathogenic mechanism of the disease remains unknown.<sup>[6]</sup> Symptoms of toxic epidermal necrolysis (TEN) and Stevens Johnson Syndrome (SJS) can be nonspecific and include symptoms such as fever, stinging eyes and discomfort upon swallowing. Typically these symptoms precede cutaneous manifestations by a few days. Early sites of cutaneous involvement are the presternal region of the trunk and the face, but also the palms and soles. Erythema and erosions of the buccal, genital and ocular mucosa occurs in more than 90% of patients and in some cases the respiratory and gastrointestinal tracts are also involved.<sup>[7]</sup> In our patient there were erosions on the trunk, scrotum, upper and lower limbs with involvement of buccal mucosa.

According to a study by Yip et al 50% of patients with TEN develop ocular complications including severe dry eye, acute conjunctivitis, trichiasis, symblepharon, distichiasis, visual loss, entropion, conjunctival membrane, ankyloblepharon, lagophthalmos and corneal ulceration.<sup>[7, 8]</sup> Surprisingly in our patient there was no ocular involvement. SJS-TEN is a disease with high morbidity that is potentially life threatening. Mortality rates are 5% with SJS, 30-40% with TEN and 10-15% with transitional forms.<sup>[2]</sup>

Treatment essentially includes prompt withdrawal of the drug and supportive care. Supportive care includes electrolyte and fluid balance, adequate nutrition, wound care and management of sepsis. Involvement of mucous membrane should be handled appropriately with topical corticosteroids, hygienic mouthwashes, aggressive ocular lubrication and local antiseptics. Early application of corticosteroids presented beneficial effects on SJS/TEN and that combination therapy of corticosteroids and IVIG achieved a better therapeutic effect than the administration of corticosteroids alone.<sup>[9]</sup> In a Chinese study combined therapy with IVIG and corticosteroids reduced mortality significantly.<sup>[10]</sup> A recent Canadian review of SJS in children found that the use of IV Immunoglobulins and corticosteroids had similar outcomes in terms of infectious complications and length of stay, and that both of these treatments were

better when compared with supportive therapy alone.<sup>[11,12]</sup> Our patient received both steroids and immunoglobulins and it can be attributed to recovery without complications in both episodes.

## CONCLUSION

This case of recurrent SJS clearly proves the importance of avoiding the implicated drug in any drug induced adverse reaction. The combination of steroids and immunoglobulins in the treatment of severe SJS and recurrent SJS was found to be beneficial. Education and reassurance should be as much a part of the treatment process as drug therapy to avoid such potentially life threatening situations.

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