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A CASE OF PEDIATRIC STEVEN JOHNSON SYNDROME –TOXIC EPIDERMAL NECROLYSIS OVERLAP: TACKLING CHALLENGES THROUGH CONSERVATIVE CARE

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Abstract

This case report outlines the case of a 16-year-old boy presenting with Stevens-Johnson syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) overlap following anti-epileptic drug therapy. Despite initial concerns for sepsis, the patient exhibited significant improvement with multidisciplinary conservative care, including antibiotic adjustment, supportive measures, and gradual tapering of steroids. Notably, liver enzyme elevation resolved with N-Acetyl Cysteine administration. This case highlights the effectiveness of early recognition, prompt intervention, and interdisciplinary collaboration in achieving favourable outcomes in complex cases of SJS-TEN overlap.

INTRODUCTION

Stevens-Johnson syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) overlap constitute a rare yet severe cutaneous hypersensitivity reaction, with potentially fatal consequences if not promptly and comprehensively addressed. The genesis of drug-induced hypersensitivity reactions is frequently associated with exposure to specific medications. Among the most frequently implicated culprits are sulfonamides, anti-epileptic drugs, fluoroquinolones, cephalosporins, and nonsteroidal anti-inflammatory drugs. This case report delineates the unfortunate experience of a 16-year-old boy who, to manage his seizures, received an anti-epileptic medication. Regrettably, this therapeutic intervention triggered the onset of Stevens-Johnson syndrome and toxic epidermal necrolysis overlap, which was successfully managed with good outcome despite several challenges

The case report

A 16-year-old male presented to the emergency room with a eight-day history of skin peeling. He also gave complaint of high grade fever since 10 days which was insidious in onset and was associated with productive cough with yellowish-white scanty sputum.

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Seven days before presentation, he developed multiple grey coloured flat coin shaped lesions as well as round, red, raised lesions over the chest, evolving into fluid-filled lesions with mild itching. Concurrently, he experienced redness, burning, and a yellowish discharge in both eyes, along with fissures, dryness of the lips, and mucosal erosions. His oral symptoms hindered mouth opening and eating, and was associated with dysphagia. The patient had a history of epilepsy for 9 months; initially treated with ayurvedic medication for eight months with poor results. Epilepsy was finally controlled with phenytoin but the patient developed skin lesions after one month of therapy.

At the time of admission, he was febrile (104°F), tachycardic (136 bpm), but with normal blood pressure and spO2. Cutaneous examination revealed exfoliation, fissuring, and xerosis over lips, ears, arms, legs, trunk, back, perianal area, and scrotum. Bullae were observed on the right medial malleolus. Pseudonikolsky sign was positive, and the lesions covered 17% of the Body Surface Area**. (Figures 1 and 2)**

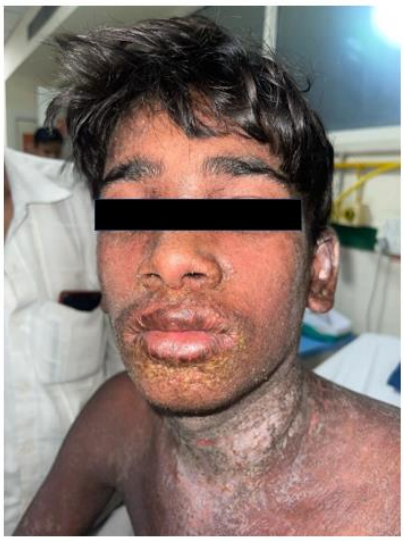

Figure 1: Showing facial and lip involvement at the time of presentation

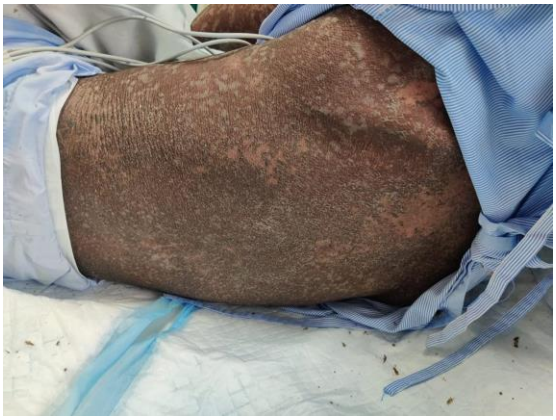

Figure 2: Showing extensive skin denudation over the back

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Investigations were conducted, including hemogram, Liver and Kidney function tests, random blood sugar, inflammatory markers (ESR, CRP, Procalcitonin) blood and urine cultures, USG whole abdomen, skin swab culture, Bleeding Time/Clotting Time, and Chest Xray (PA view). They revealed elevated total leucocyte count, liver enzymes and Procalcitonin. He fulfilled systemic inflammatory response syndrome criteria, prompting admission to the Intensive Care Unit for suspected sepsis.

Multidisciplinary approach was adopted for management of the patient. Extensive Injectable medications, including ceftriaxone (antibiotic cover), dexamethasone (for SJS), levetiracetam (for epilepsy), paracetamol, IV fluids, pantoprazole, topical treatments, and high-protein diet were initiated. Supportive treatment with intravenous fluids, paracetamol and protein rich diet was given.

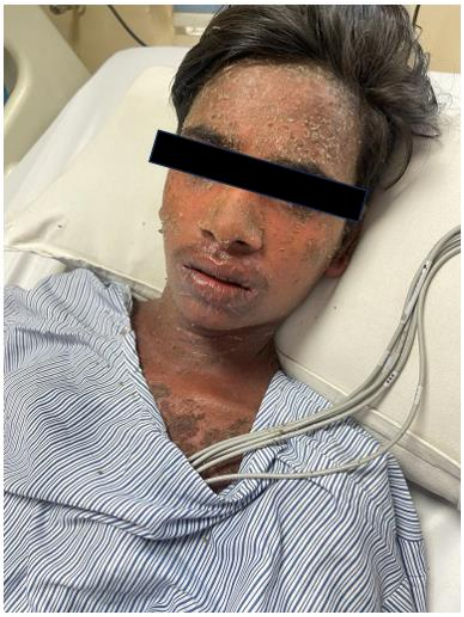

Figure 3: on third day of admission

Figure 4: Significant improvement on day 6 of admission

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New lesions only appeared over the first two days, with marked clinical improvement thereafter **(Figures 3 and 4).** Antibiotics were adjusted based on culture sensitivity of urine culture and skin swab culture report. During the initial days of admission patient experienced anxiety and was very much disturbed. The patient's comfort improved with psychiatric support and anti-anxiety medication.

Elevated SGOT/SGPT enzymes were concerning. Medicine speciality team suspected drug liver injury and N-Acetyl Cysteine was administered in view of that. Patient responded very well to that and liver enzymes showed gradual improvement to normal levels at the time of discharge. As the patient clinically improved and TLC reduced, dexamethasone was gradually tapered and eventually stopped over a period of ten days.

Figure 5: Follow up picture after 1 week of discharge

At the time of discharge, all the lesions had healed with few areas showing crusting. Xerosis and post inflammatory hyperpigmentation were managed with emollients and antihistamines post discharge. On subsequent follow up, no new lesions have appeared and the patient has tremendously improved overall **(Figure 5).**

DISCUSSION

Stevens-Johnson syndrome and toxic epidermal necrolysis (SJS/TEN) represent severe cutaneous adverse reactions impacting mucocutaneous surfaces [1]. SJS is distinguished by the detachment of less than 10% of the body surface area, while TEN involves more than 30% detachment. Overlapping cases, ranging between 10-30% body surface area detachment, fall within the SJS/TEN spectrum. The mortality rate for SJS-TEN spans from 10% to 34% [2].

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In the United States, the estimated annual incidences among adults were 9.2, 1.6, and 1.9 cases per million individuals for SJS, SJS/TEN overlap, and TEN, respectively [2]. Adjusted mortality rates averaged 4.8% for SJS, 19.4% for SJS/TEN, and 14.8% for TEN [2].

Various factors contribute to SJS/TEN, encompassing medications, mycoplasma pneumonia, herpes, hepatitis A, and vaccination [3]. Predominantly, drugs are the primary instigators of this life-threatening condition. Common offenders include sulphonamides, anti-epileptic drugs like phenytoin, carbamazepine, lamotrigine, phenobarbital, and allopurinol, as well as non-steroidal anti-inflammatory drugs such as piroxicam, diclofenac, and nevirapine [4].

The underlying pathophysiology of SJS-TEN involves keratinocyte apoptosis, instigating epidermolysis and subsequent blistering [5]. Cytotoxic T-cell lymphocytes, present in TEN blister fluid, are thought to initiate intracellular enzyme activation, culminating in apoptosis [6]. Among antiepileptics, phenytoin and carbamazepine emerge as the most common offenders [7]. A potential genetic association between the HLA-B 1502 allele and phenytoin-induced Stevens-Johnson syndrome (SJS) in Asian patients, suggesting a possible genetic predisposition to SJS in specific populations. [8].

Clinical evaluation predominantly constitutes the diagnosis of SJS-TEN overlap [9]. Various serum markers have been explored for early TEN detection and disease progression monitoring, such as soluble CD40 ligand, Fas ligand, granulysin, granzyme B, serum high mobility group protein B1, alpha-defensins 1-3 in blister fluid, Bcl-2 expression in dermal infiltrates, thymus and activation-regulated chemokine, and Glutathione-S-transferase-pi expression. IL-15, particularly useful for predicting disease severity and monitoring prognosis, has been identified **[12,13]**. Histological examination of skin cryosections or formalin-fixed sections confirms extensive necrosis across all layers [14]. Direct immunofluorescence staining is crucial to rule out autoimmune blistering disorders, providing insight into immunoglobulin and complement deposition in the epidermis or at the epidermal-dermal junction.

Management of SJS-TEN overlap mandates immediate hospitalization in an intensive care or burn unit **[15].** Discontinuation of the triggering medication is the primary treatment for SJS-TEN Overlap. Systemic steroids are used to halt the progress of disease during active phase. Supportive care, encompassing fluid replacement and nutrition, is paramount due to extensive dehydration resulting from widespread skin loss in SJS-TEN [16]. Cool and wet compresses may alleviate blisters, while certain medications offer symptomatic relief, such as pain medication to reduce discomfort, topical steroids to alleviate inflammation of the eyes and mucous membranes, and systemic antibiotics, administered only in the presence of signs of infection or sepsis, not prophylactically. Studies suggest the efficacy of etanercept and cyclosporine in treatment [17,18].

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Surgical/interventional measures, mainly debridement followed by skin substitute grafting have also been used for wound management. IVIg and Plasmapheresis also play a role in management of severe cutaneous drug reactions [10].

CONCLUSION

SJS-TEN overlap is a grave condition demanding early diagnosis. Clinicians should remain vigilant for clinical symptoms, enabling proactive identification. Early diagnosis, meticulous complication monitoring, and supportive care are pivotal in the management of SJS-TEN overlap. This case is presented due to the patient's complex presentation involving skin and oral mucosa, history of epilepsy, requirement of intensive care and a adoption of a multidisciplinary approach which included team of dermatology, ophthalmology, general medicine, psychiatry and intensive care specialists.

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