Asian Journal of Medicine and Health

Volume 21, Issue 8, Page 142-146, 2023; Article no.AJMAH.98691
ISSN: 2456-8414

Stevens-Johnson Syndrome Secondary to Herpetic Infection: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/AJMAH/2023/v21i8858

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/98691

Case Study

Received: 05/02/2023
Accepted: 09/04/2023
Published: 10/06/2023

ABSTRACT

Stevens-Johnson syndrome (SJS) is a rare but serious drug reaction characterized by extensive necrosis of the skin and mucous membranes. It is considered a medical emergency and requires immediate medical intervention, with a high mortality rate, especially during the acute phase.
Symptoms of Stevens-Johnson syndrome include a rash, blisters, and lesions in the oral mucosa, throat, genitals, and eyes. Before the rash appears, symptoms such as fever, headache, muscle pain, and flu-like signs may occur.
While the causes of Stevens-Johnson syndrome are not fully understood, it is often associated with an allergic reaction to medication. In children, it can also have an infectious origin.
Survivors of Stevens-Johnson syndrome may experience long-term sequelae, including cutaneous, ophthalmological, genital, oral-dental, and psychological issues. Therefore, monitoring and early detection are crucial.
The severity of this pathology makes the reported case particularly compelling. It highlights the importance of medical personnel having knowledge and mastery of this condition in order to improve management and prognosis.
We report the case of an 8-year-old child who presented with a pseudogripal syndrome that rapidly

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progressed in 24 hours to an exanthem characterized by macules in pseudo-rings. The condition was complicated by a severe, diffuse enanthem and resulted in detachment with a positive Nikolsky sign. The clinical and biological evolution was favorable under symptomatic treatment, antiviral medication and intravenous immunoglobulin infusion.

Keywords: Stevens-Johnson syndrome; bullous toxidermia; herpes.

1. INTRODUCTION

“Stevens-Johnson syndrome (SJS) is a rare and potentially life-threatening skin disorder, affecting only 1 to 2 individuals per million annually. The hallmark feature of SJS is the rapid and severe destruction and detachment of skin and mucosal epithelium, which typically involves less than 10% of the total body surface area. While SJS is most often caused by certain medications, infections, particularly those caused by Herpes simplex and Mycoplasma pneumoniae, may also trigger the syndrome” [1,2].

Due to the significant risk of mortality associated with this condition, SJS is considered a medical emergency. The primary focus of management is providing supportive care, removing the causative agent, monitoring for complications, and treating any associated infections.

2. PRESENTATION OF CASE

We present a clinical case with the written consent of the parent. Our case involves an 8-year-old child, male sex, from a non-consanguineous marriage with no history of recurrent infections or recent medication. Four days prior to admission, the child presented with a flu-like syndrome, bilateral conjunctivitis, and painful, pruritic, and febrile rash. The rash was predominantly located on the trunk and the roots of the limbs, consisting of purpuric erythematous macules and pseudococardia with a concentric change of color, and eventually developed a central bulla [Fig. 1].

The child's condition was further complicated by a diffuse enanthemata with gingivostomatitis, characterized by erosions and hemorrhagic crusts that created an appearance of sooty cheilitis, making feeding difficult. The child also experienced painful bilateral conjunctivitis, with sensations of a foreign body, excessive tearing, photophobia, and conjunctival hyperemia [Fig. 2].

The child's condition deteriorated further, as the skin continued to detach due to the merging of blistering lesions. This resulted in a red, oozing dermis [Fig. 3], with positive Nikolsky's sign, especially in areas where pressure was applied.

Upon admission to the hospital, the child's condition continued to worsen, prompting further assessment and evaluation. Blood tests revealed lymphopenia at 600/µl and neutropenia at 1800/µl, while liver and kidney function tests showed no abnormalities. However, there was an inflammatory response, as evidenced by elevated ferritin levels at 966 ng/ml and a C-reactive protein level of 120 mg/l. A chest X-ray revealed bronchial syndrome with basal atelectasis in the right upper lobe, and a respiratory Multiplex polymerase chain reaction (Multiplex PCR) identified a respiratory syncytial virus. Serological testing for herpes simplex virus 1 and 2 came back positive.

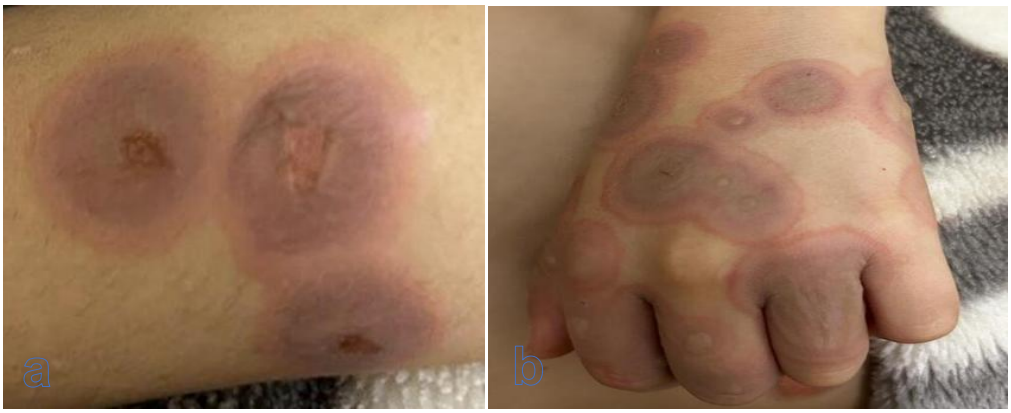

Fig. 1. The skin rash initially appeared as purpuric erythematous macules and pseudococardia, which then evolved to a concentric change of color and finally a central bulla

Fig. 2. Diffuse enanthema with gingivostomatitis and bilateral conjunctivitis

A skin biopsy was performed, which showed necrosis of the entire epidermis that had detached from a slightly altered dermis. A slit-lamp ophthalmologic examination found herpetic keratoconjunctivitis with a dendritic ulceration on the cornea.

To treat the patient, intravenous antiviral medication (Acyclovir) was administered for 10 days with intravenous immunoglobulin infusion, along with topical ocular corticosteroids, an antiviral (ganciclovir) and a lubricant, with daily aseptic skin care. The clinical and biological evolution was favorable, with progressive healing of the lesions [Fig. 4], resumption of feeding, and normalization of the blood count.

3. DISCUSSION

Stevens-Johnson syndrome/toxic epidermal necrolysis (SJS/TEN) is a severe skin reaction that is typically caused by certain medications, with drug allergies being the most common trigger. Sulfonamides, penicillin, hydantoin, phenylbutazone, and barbiturates are the most common drugs associated with severe forms of the syndrome. In some cases, infections, particularly those caused by Herpes simplex and Mycoplasma pneumoniae, may also trigger the syndrome [1,2].

Previously thought to be separate conditions, Stevens-Johnson syndrome and toxic epidermal necrolysis are now considered part of a continuum. Stevens-Johnson syndrome represents the less severe end of the spectrum, while toxic epidermal necrolysis represents the more severe end.

Stevens-Johnson syndrome/toxic epidermal necrolysis (SJS/TEN) is diagnosed primarily based on the patient's clinical presentation. Patients often report systemic symptoms such as fever, malaise, arthralgia, and sore throat. Skin lesions typically begin as erythematous to violaceous and purpuric macules that coalesce to form patches. Targetoid lesions may also be present.

Lesions usually first appear on the trunk and then spread distally to involve the limbs, and flaccid bullae may also be present. The extent of skin involvement determines the diagnosis: SJS involves less than 10% of the body surface area, SJS-TEN overlap involves 10% to 30%, and the most severe form, TEN, involves greater than 30% of the body surface area. Involvement of mucous membranes, including the oral, ocular, and genitourinary mucosa, is nearly universal.

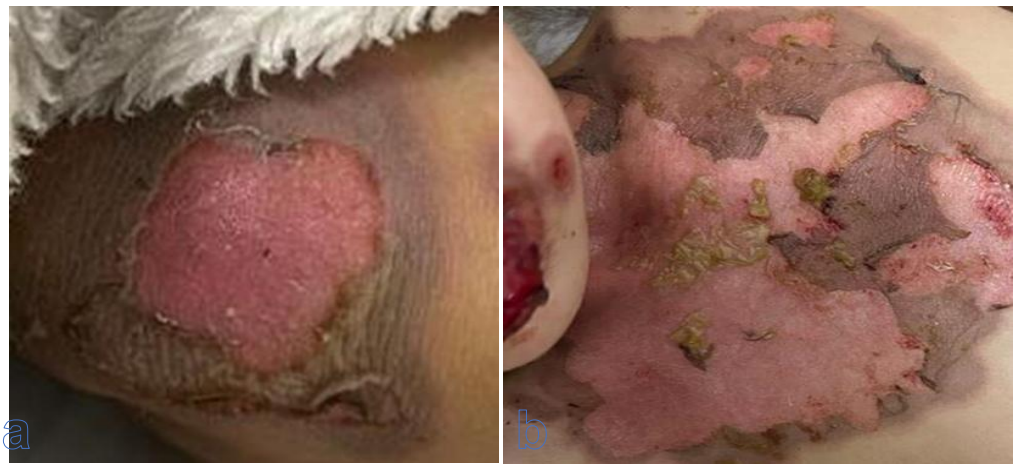

Fig. 3. Detachments resulting from the coalescence of bubble elements

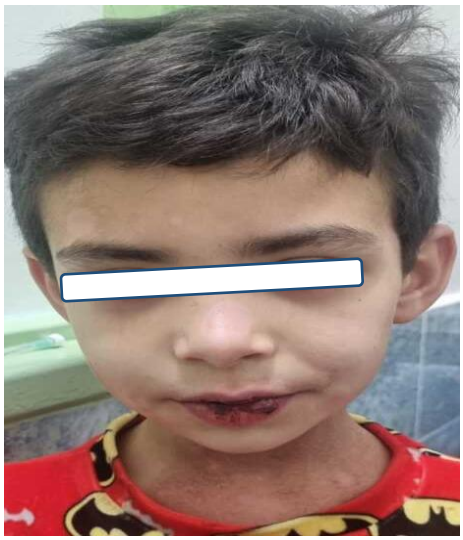

Fig. 4. Favourable clinical evolution with progressive healing of the lesions

"Pseudo-Nikolsky and Asboe Hansen signs are present in most cases. While histopathology is not required for diagnosis, it typically reveals full-thickness epidermal necrosis, subepidermal bullae, and scanty inflammatory infiltrate in the papillary dermis, which are characteristic of SJS-TEN" [3].

"Recently, various serum markers have been studied, which can detect an early case of TEN and signal the progression of early morbilliform rash to a full-blown case of TEN. Some of them include soluble Fas ligand, granzyme B, soluble CD40 ligand, granulysin, serum high mobility group protein B1 (HMGB1), serum lactate dehydrogenase level, alpha-defensins 1–3 in the blister, Bcl-2 expression in the dermal infiltrate, thymus and activation-regulated chemokine, and glutathione-S transferase-pi expression" [4,5,6].

"The disease prognosis can be determined through the use of the TEN (SCORTEN) score, which is based on 7 parameters. These parameters include age ≥40 years, heart rate ≥120/min, presence of cancer/hematologic malignancy, >10% body surface area involvement, raised blood urea nitrogen (>28 mg/dL), serum bicarbonate <20 mmol/L, and serum glucose level > 14 mmol/L, all of which are calculated within the first 24 hours of the patient's admission" [7].

"The management of sequelae is essential, since the disease involves ocular, oral, genitourinary, gastrointestinal, and respiratory mucosae, complications can be plenty, depending on the extent of the disease and the point of therapeutic intervention. Early referral to an ophthalmologist is quintessential for estimation of involvement of ocular mucosa. Visual outcome is better in those receiving ophthalmological treatment (topical steroids and lubricants), preferably within 7 days of onset of disease" [8,9]. Serious ocular complications such as corneal scarring, corneal xerosis, trichiasis, and subconjunctival fibrosis need gas permeable scleral contact lens therapy and amniotic membrane transplantation.

"Cutaneous complications can be managed with nonadherent dressings. Respiratory complications such as bronchitis, bronchiectasis, bronchiolitis obliterans, and bronchiolitis obliterans organizing pneumonia can be treated with aerosols, nebulized saline, bronchodilators, bronchial aspiration, physical therapy, intubation, and mechanical ventilation. While hypopharyngeal stenosis and esophageal strictures are rare complications, removal of oral crusting may be necessary when required. Early enteral feeding is crucial to prevent such sequelae, and severe cases may require laryngectomy" [10].

Literature on the management of SJS-TEN in children is currently limited. However, a recent review [11] revealed that major treatment options included intravenous immunoglobulin (IVIg), systemic corticosteroids such as prednisolone, methylprednisolone, dexamethasone, surgical debridement, and supportive treatment alone.

A small number of children were treated with ulinastatin, plasmapheresis, G-CSF [12], IV pentoxifylline [13], skin substitutes [14], and cyclosporine.

The use of steroids and IVIg was associated with improved outcomes, while those treated with supportive therapy alone were found to have higher morbidity and mortality rates.

4. CONCLUSION

Stevens-Johnson syndrome is a rare and severe skin disorder characterized by necrosis of the epidermis, which can progress rapidly and potentially be life-threatening. Treatment mainly focuses on relieving symptoms associated with steroids and IVIg. Early recognition and prompt management of SJS are essential for preventing severe complications and improving the prognosis. Complications may affect multiple

organ systems, and specialized care from medical and surgical specialists may be necessary to manage these complications. Close follow-up and monitoring are also important to detect any potential long-term effects of the disease.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
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