Case study

Stevens-Johnson syndrome secondary to herpetic infection, a case report

ABSTRACT

Stevens-Johnson syndrome (SJS) is a rare and serious bullous toxidermia, it is characterized by an extensive necrosis of the epidermis and mucous membranes, with a high mortality especially in the acute phase. This syndrome is caused in the majority of cases by drugs, but can be of infectious origin especially in children.

Long-term sequelae (cutaneous, ophthalmological, genital, oral, psychological) in survivors are almost constant, hence the interest of surveillance and early detection.

We report the case of an 8-year-old child with Stevens-Johnson syndrome secondary to a herpetic infection with a good evolution under antiviral and supportive symptomatic treatment.

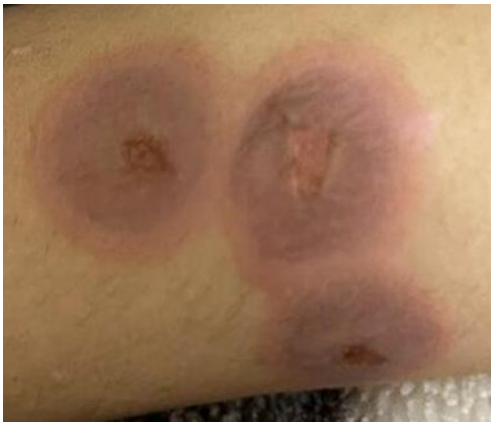
Key words: Stevens-Johnson syndrome, bullous toxidermia, herpes.

1. INTRODUCTION:

Strevens johnson syndrome is an unpredictable, severe, and life-threatening toxidermia characterized by destruction and detachment of skin and mucosal epithelium over less than 10% of the total body surface area.
It is considered a medical emergency and management is primarily focused on support, removal of the causative agent, monitoring, and treatment of superinfection due to the high risk of death associated with these patients.

2. PRESENTATION OF CASE:

Our case is a 08 year old child, from a non-consanguineous marriage, no recurrent infection, no recent medication. He presented 4 days before his admission a flu-like syndrome with bilateral conjunctivitis, evolving in 24 hours to a painful, pruritic and febrile rash, predominantly on the trunk and the roots of the limbs, made of purpuric erythematous macules and pseudococardia with a concentric change of color, then a central bulla [fig. 1].



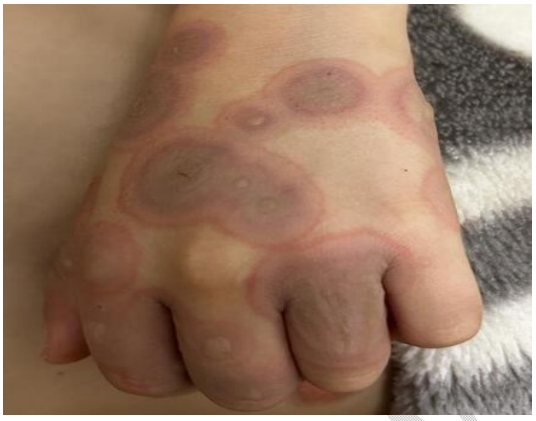


Fig.1. a skin rash made of purpuric erythematous macules and pseudococardia evolving to a concentric color change and then a central bulla.

This is complicated by a diffuse enanthemata with gingivostomatitis made of erosions and hemorrhagic crusts realizing an aspect of sooty cheilitis preventing feeding, associated with a painful bilateral conjunctivitis with sensations of foreign body, lacrimation, photophobia and conjunctival hyperemia [fig. 2].

Fig.2. Diffuse enanthema with gingivostomatitis and bilateral conjunctivitis

The evolution is towards detachments resulting from the coalescence of bullous elements and gives way to a red oozing dermis [fig. 3], with Nikolsky's sign which is positive in particular in the pressure areas.

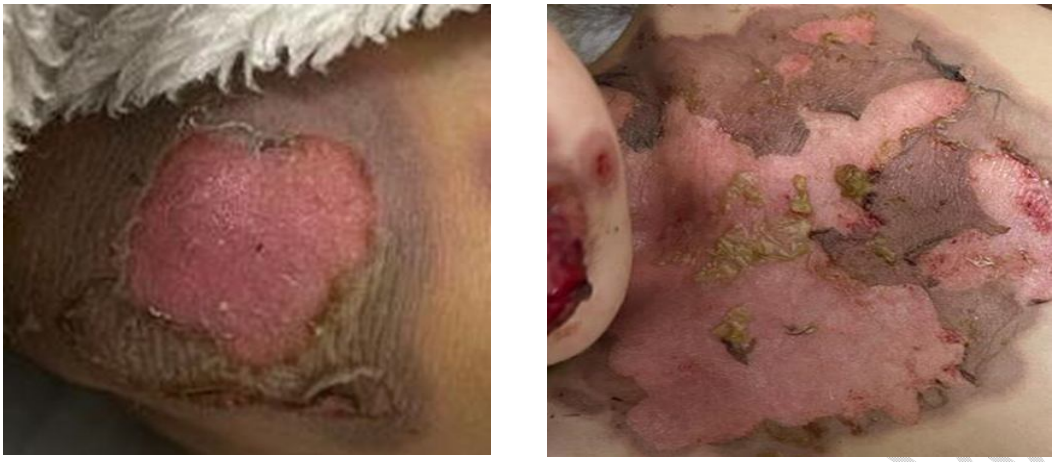


Fig.3. Separations resulting from the coalescence of bubble elements

The patient was hospitalized with a conditioning and a paraclinical check-up, the blood count found a lymphopenia at 600/µl and a neutropenia at 1800/µl, the liver and kidney check-up were without abnormality, an inflammatory syndrome with a ferritinemia at 966 ng/ml and a C-reactive protein at 120 mg/l.
A chest X-ray showed a bronchial syndrome with basal atelectasis of the right upper lobe, a respiratory Multiplex polymerase chain reaction (Multiplex PCR) found a respiratory syncytial virus, the serology of herpes simplex virus 1 and 2 came back positive.
The skin biopsy was performed showing a necrosis of the entire epidermis, which was detached from a slightly altered dermis.
A slit lamp ophthalmologic examination found a herpetic keratoconjunctivitis with a dendritic ulceration on the cornea.
The patient was put on an antiviral (Acyclovir) intravenously for 10 days, an antiviral (ganciclovir) and topical ocular corticosteroid with daily skin care in an aseptic setting.
The clinical and biological evolution was favorable with progressive healing of the lesions [fig. 4], resumption of feeding and normalization of the blood count.

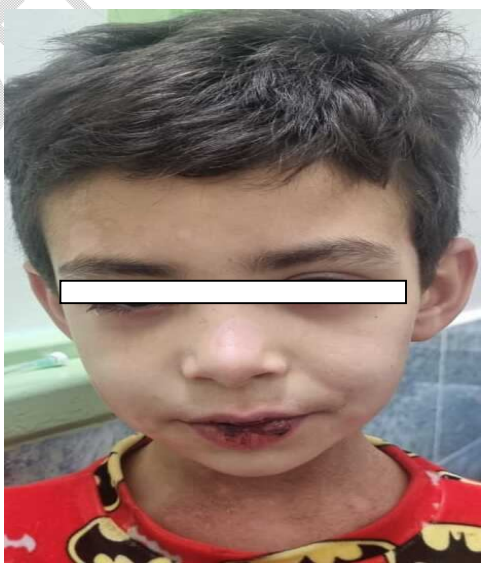


Fig.4. favourable clinical evolution with progressive healing of the lesions

3. DISCUSSION:

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are rare and severe bullous toxidermia characterized by epidermal necrosis, which can evolve extremely rapidly and be life-threatening.

These pathologies affect between 1 and 5 subjects/million, and the incidence and/or severity of these pathologies may be higher in bone marrow transplant recipients, HIV-positive patients infected with Pneumocystis jirovecii, in cases of systemic lupus erythematosus or other chronic rheumatological pathologies.

The most frequent causes are allergic (sulfonamides, penicillin, hydantoin, phenylbutazone and barbiturates) [1, 2], sometimes infectious (in particular by Herpes simplex and then M. pneumoniae), drug allergies are responsible for the most severe forms [1].

SJS begins with a flu-like syndrome with rapid onset of mucosal and then cutaneous signs. The initial rash is painful and febrile, consisting of atypical or pseudo-cocardial macules, predominantly axial, especially on the trunk and the roots of the limbs.

Two dermatological pictures can be seen: a large detachment of sudden installation in "wet linen" on a background of erythema, or detachments resulting from the coalescence of bullous elements in atypical targets. Nikolsky's sign is positive on most of the integument and gives way to a red oozing dermis with an appearance similar to that of a superficial second degree burn.

The extension of epidermolysis must be assessed daily by distinguishing the detached surface and the detachable surface, because this is the essential prognostic factor and a guide for treatment, it is expressed as a percentage of the affected skin surface using the Wallace rule of 9 for children: 17% for the head and neck, 9% for each arm, 18% for the front of the trunk, 18% for the back of the trunk, 14% for each leg, 1% for the perineum
A distinction is generally made between Stevens-Johnson syndrome (SJS) in the case of a detachment < 10% and Lyell syndrome in the case of a detachment > 30%. Forms with a detachment between 10 and 30% are called "transitional" [3].

Complications must be systematically sought because of their severity and the possible cutaneous, ophthalmological, genital, oral and psychological sequelae in survivors, which are almost constant. One third of patients have pulmonary sequelae [4], and patients with M. pneumoniae infection are at greater risk of developing bronchiolitis obliterans [5, 6].

Ocular involvement is present in the acute phase in 2/3 of cases [7], daily ophthalmological follow-up and early and intensive treatment are necessary in order to avoid as much as possible complications such as synechiae, keratitis and corneal erosions. A dry syndrome can appear from the first days and can persist for several years. The main long-term complications are: symblepharon, entropion, trichiasis responsible for keratitis, lacrimation due to obliteration of the lacrimal points, and even opacification of the cornea with a significant decrease in visual acuity [8.9].
Children with Steven-Johnson syndrome should have ophthalmologic, radiologic, and pulmonary follow-up after hospitalization.

To date, no specific treatment has been shown to be superior to supportive care alone in epidermal necrolysis associated with etiological treatment in case of infectious cause [10].
Immunosuppressive treatment to be initiated in the acute phase also remains highly debated.

Systemic corticosteroid therapy, compared to supportive therapy alone, has not been shown to be effective in reducing mortality in the acute phase.

4. CONCLUSION:

Stevens-Johnson syndrome is a rare and serious bullous toxidermia characterized by a necrosis of the epidermis which can be extremely fast devolution engaging the vital prognosis.
The treatment remains symptomatic and etiological in case of infectious cause, the complications and the after-effects can be very severe, they are to be sought and taken care of as early as possible to improve the prognosis.

5. REFERENCES:

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