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Incomplete Stevens-Johnson Syndrome Related to *Mycoplasma Pneumoniae* Infection: A Rare Case Report

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ABSTRACT: Stevens-Johnson Syndrome (SJS) is a rare and severe mucocutaneous disease characterized by widespread epidermal necrosis and mucosal involvement affecting children and adults. It is an immune-mediated disease usually secondary to drug reaction, but in few cases it is an extrapulmonary complication of *Mycoplasma pneumoniae* infection. We report a case illustrating that the incomplete Steven-Johnson syndrome associated to *Mycoplasma pneumoniae* infection may present with skin and oral mucosal involvement alone without conjunctival or genital involvement.

Keywords: Stevens-Johnson Syndrome (SJS), Mucocutaneous, *Mycoplasma pneumoniae.*

CASE REPORT

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INTRODUCTION

Stevens-Johnson syndrome (SJS) is a common dermatologic disease that is characterized by a prodromal illness followed by severe cutaneous symptoms and mucosal involvement. It is considered an adverse reactive process usually secondary to a drug reaction although *Mycoplasma Pneumonia* and *herpes virus* infection are also well documented causes [1].

There is a consensus in the literature that all the patients harbored SJS developed an immunological hypersensitivity reaction after antigenic challenge [2].

The diagnosis of SJS is based mainly on clinical signs as cutaneous target lesions and mucosal involvement confirmed by histological analysis.

There are no standardized guidelines for treatment of SJS. Recognition and prompt discontinuation of the offending agent is necessary to control the disease [2].

CASE REPORT

A 10-year old boy, with no significant past medical history, was hospitalized at the pediatric department.

His mother stated that about 10 days prior, the patient developed headache, cough and mild nasal congestion. He was given amoxicillin with clavulanic acid, paracetamol and mefenamic acid anti-inflammatory drugs without any improvement after 3 days of therapy. 2 days after these symptoms, he developed a progressive concentric skin blisters and ulcerative lesions that began on his feet and hands and became generalized. This was associated to oral ulcers and lip blisters quickly ruptured and covered by crusts.

During this period, the patient was seen by general practitioner and given prescriptions for acyclovir and an antifungal treatment, but his condition was worsened.

On admission, a cough with expectoration of purulent sputum and a fever of over 39˚C were noted.

Physical examination showed vesicular lesions over face, a rash over his extremitie, and concentric lesions over the trunk and abdomen (Figure 1).

Oral mucosa and lips showed multiple erosions and ulcers covered with yellow excudates. The patient reported that oral lesions were painful, and caused him to have difficulty swallowing.

No lesions elsewhere were noted and, in particular, examination of the genitalia was normal. There was no evidence of petechiae or conjunctival involvement.

Based on the history and skin findings, the diagnosis of SJS was evoqued, whether associated to a drug reaction or an infectious one. Peripheral blood test results were non contributory. Cultures for herpes simplex virus (HSV), serologies for Epstein-Barr virus (EBV) and cytomegalovirus (CMV) were negative. *Mycoplasma pneumoniae* immunoglobulin G (IgG) and IgM titers were elevated showing a Mycoplasma pneumoniae infection.

A chest X-ray radiograph was performed and showed bilateral interstitial alveolar opacities with ascension of the diaphragmatic dome (Figure 2). Based on this picture and the profuse greenish sputum, the patient was tested for tuberculosis which was negative.

Lymphocyte Transformation Test was negative excluding the possibility of drug reaction.

A skin biopsy of an erythematous macule with a necrotic center on the abdomen was performed in the dermatology department showed epidermal necrosis and a mild lymphocytic infiltration in the upper dermis suggestive of Stevens-Johnson syndrome (SJS).

The diagnosis of SJS secondary to *Mycoplasma pneumoniae* infection has been retained.

He was started on oral clarithromycin (500 mg *2/ day) for one week initially to cover for *Mycoplasma.* Topical ointment for skin lesions was prescribed by the dermatologist.

The patient was refered to our department (dental medicine department, Fattouma Bourguiba Teaching Hospital, Monastir Tunisia) for the management of the extensive and painful oral lesions.

The exam showed synechic ulceration and bulla on both upper and lower lips (Figure 3). After abundant hydration with saline, the patient was able to open his mouth.

The intra oral examination revealed an extensive ulceration including buccal mucosa, tongue, and palate. The gums presented no evidence of lesion. Some areas were covered with whitish deposits (Figure 4).

The patient was treated with topical corticosteroid (prednisolone 20 mg 3 times a day) as a mouth wash. His mother was advised to clean his son's mouth carefully with a compress soaked in nystatin, in order to treat the candidal fungal superinfection.

After 3 days of therapy his clinical state improved and he started to be able to consume adequate fluids and food.

All symptoms improved gradually (Figure 5). After 2 weeks of treatment a good regression of skin and mucosal changes was observed. Hence, the patient was discharged home with follow-up with Pediatric, Dental Medicine and Dermatology departements.

A follow-up assessment one month after discharge was normal (Figure 6).

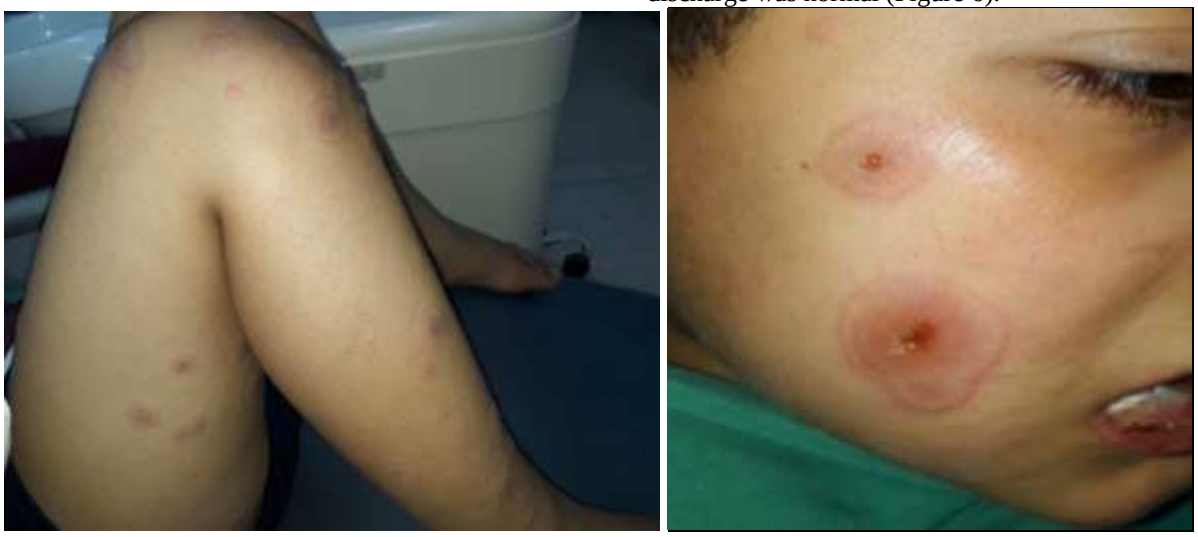

Figure 1: Target lesions

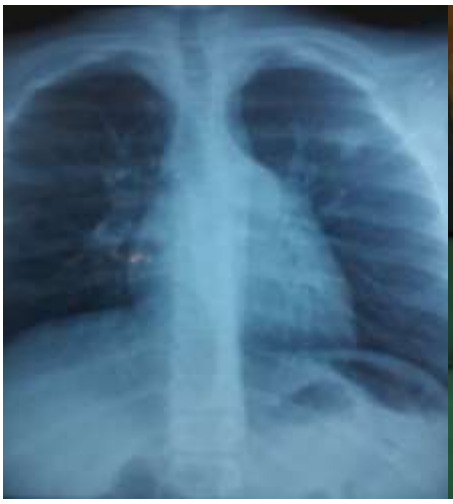

Figure 2: Chest X-ray radiograph

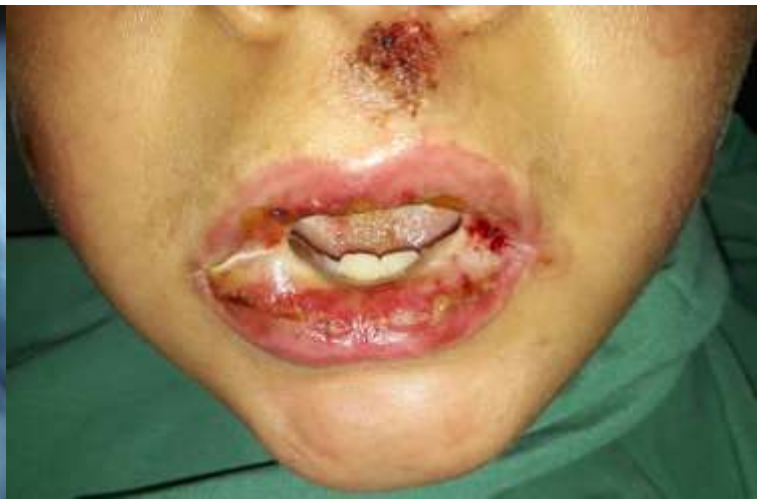

Figure 3: Extensive ulcer and bullae of the lips

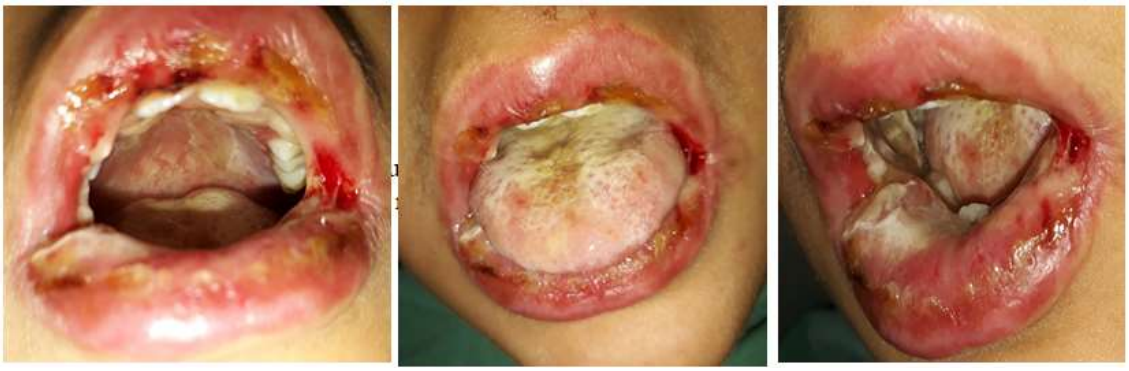

Figure 4: Intra oral view: ulcerative lesions, whitish deposits on the tongue and yellow excudates covered the buccal mucosa

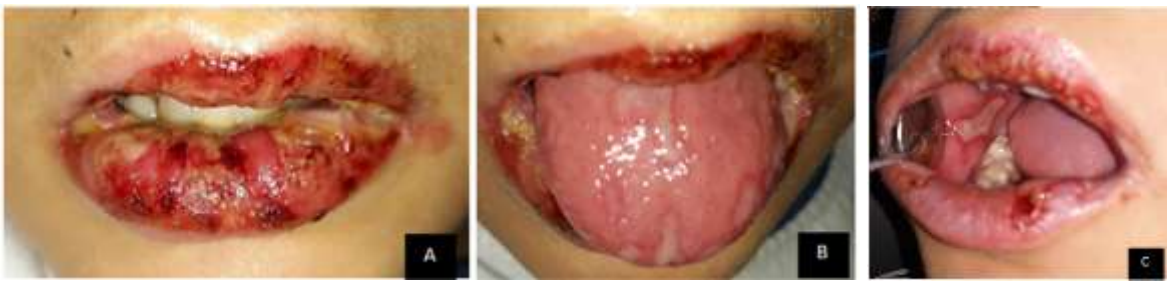

Figure 5: Follow up: a) after 3 days of therapy. b) after a week. c) after 10 days

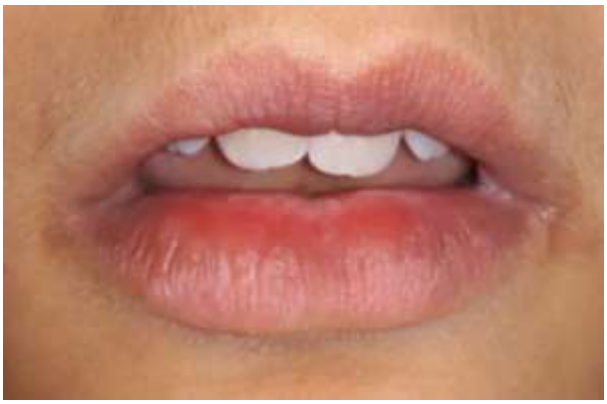

Figure 6: Follow up after one month

DISCUSSION

Stevens-Johnson Syndrome (SJS) is a severe mucocutaneous disease that was first described by Stevens and Johnson in 1922.

According to the International Classification of Diseases, Ninth Revision (ICD-9) code for SJS, the clinical criteria include involvement of skin and at least 2 mucous membranes [3].

The oral and esophageal mucosa, eyes, and genital areas may be affected with different degree associated to moderate skin involvement presenting with targetoid or morbilliform eruptions and minimal skin shedding [4].

Our case illustrates that incomplete presentation of this syndrome can occur with only skin involvement and oral mucosa lesions alone, without conjunctival or genital involvement.

In children, infections are the most commonly identified cause of SJS, with *Mycoplasma pneumoniae* (Mp) implicated most frequently [5].

In 25% of patients with Mp infection, extrapulmonary complications may appear, including severe mucocutaneous blistering complications [6].

Nowsheen S *et al*., [4] concluded that in the pediatric patients group, SJS was associated to infection in more than a half of patients and the main infection noted was Mycoplasma in 91%.

Other associated infections include HSV, Mycobacterium Tuberculosis, Epstein-Barr virus (EBV), group A streptococci, hepatitis B virus, and enteroviruses [5]. Therefore our patient was tested for HSV, EBV, CMV and Tuberculosis infections, with negative results.

Our case about a 10-year-old boy was in agreement with literature. As the patient population seen with this disease (SJS associated Mp infection) was mostly male children and young adults [7].

An average age at diagnosis was 10 years in the SJS pediatric patients, and 11.5 years according to Nowsheen *et al*., [4] and Olson *et al*., [8] studies, respectively.

These two studies showed that male were more affected. Nowsheen *et al*., [4] and Olson *et al*., [8] reported that boys were 62% and 63%, respectively.

The clinical features of disease in our case was similar to previous published case series of SJS associated Mp infection. Prodromal respiratory illness and fever were common. SJS associated Mp was characterized by extensive mucosal lesions and less severe distribution of skin involvement [8]. Oral synechiae was also reported [9].

Our patient did not present ophthalmic lesions. Nowsheen *et al*., [4] reported that all SJS associated Mp patients presented mucosal lesions but only 76.2% of young patients had ophthalmic involvement.

The diagnosis of SJS is based mainly on clinical signs confirmed by histological analysis showing full-thickness epidermal necrosis due to extensive keratinocyte apoptosis [1].

Diagnosis of Mp infection can be through either culture isolation or serological detection of IgM/IgG antibodies. ELISA for IgM antibodies is the cheapest and most widely available test [5]. Recently, PCR testing for Mp has begun to replace or accompany serology, allowing greater diagnostic accuracy [8].

Establishing the right diagnosis is extremely important because it allows proper management and prognostication.

SJS should be distinguished from erythema multiforme (EM) and toxic epidermal necrolysis (TEN). The current consensus is that SJS and TEN are likely variants of the same disease, typically associated with drug reaction, and less frequently with infections such as Mp distinct from EM, which is strongly linked to herpes simplex virus infection [9].

Histopathological findings, such as perivasculitis and necrosis of the epidermis, are similar in SJS and TEN, but differ from EM. The main differences between SJS and TEN are in percentage of skin detachment and outcomes. EM is characterized by a milder cutaneous illness with symmetrically distributed, round, red papules that may progress to target lesions, but do not engage the mucosa and exhibit systemic symptoms [7].

Due to the less severe cutaneous manifestations and organ involvement in the SJS associated Mp, several authors proposed renaming this entity to Mp-associated SJS-like disease with mild or no skin lesions, Mp-associated mucositis, atypical SJS, incomplete SJS, or Fuch syndrome [8]. Canavan *et al*., propose the designation of ‘‘Mycoplasma induced rash and mucositis’’ as a distinct clinical entity based on the following: 1) Distinct morphology with prominent mucositis and – when cutaneous involvement is present - a characteristic sparse vesiculobullous and/or targetoid eruption, 2) Milder disease course with infrequent long-term sequelae and exceedingly rare mortality, 3) Pathophysiology that is distinct from other erythema multiformeespectrum diseases including direct cutaneous infection, 4) Potentially distinct management approaches [9].

The treatment of SJS is mostly supportive. The role of intravenous immunoglobulin (IvIg) and steroids is still controversial. IvIg has demonstrated the capacity to block the apoptotic ligand (Fas ligand) from binding to the Fas receptor, thus preventing keratinocyte apoptosis and subsequent epidermal detachment [5]. Steroids have not been proven to have any beneficial role and some recent studies indicate that they may even worsen the outcome [5].

In case of Mp infection, antibiotics against this species should be administered. The macrolide or tetracycline antibiotics are effective in this case and improve respiratory symptoms, but the organisms may still be cultured from the respiratory tract even after treatment, sometimes for months. The penicillin or cephalosporin families have no effect on Mp because of the absence of a cell wall.

CONCLUSION

General or specialist practitioner and dentist should be aware of the following;

- The search for the etiological factor is essential in SJS cases
- Mycoplasma is a frequent trigger of SJS in pediatric patients
- Multidisciplinary team care
- Long term follow up is necessary.

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