

# Systemic lupus erythematosus presenting as stevens johnson syndrome in a thirty years old female: a case report

## Abstract

**Background:** SJS occurs almost exclusively secondary to drugs but very rarely, SLE can be an inciting factor for SJS without presence of an offending drug. The association is extremely rare, however few cases have been reported.

**Case presentation:** We present a case report of a thirty year old female who presented to the rheumatology clinic with complaints of rash on her face, swelling of the lips and oral ulcers with severe hemorrhagic cheilitis. She also complained of persistent high grade fever with development of blisters on her face and oropharyngeal mucosa. She had been suffering from pain in multiple joints which did not respond to NSAIDs. A diagnosis of SLE presenting as SJS was established. Patient was given good supportive care, steroids, DMARDs and analgesics, to which she responded well and recovered.

**Conclusion:** Stevens Johnson Syndrome is an uncommon, severe dermatological condition usually caused secondary to drugs. Secondary cause of the disease may rarely be co-existence of an autoimmune condition like SLE, as present in our patient.

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**Mohammad Shariq Mukarram**  
Mohammad Shariq Mukarram**Correspondence:** Mohammad Shariq Mukarram, Jinnah Medical College Hospital, Karachi, Pakistan, Tel +923343572852, Email shariq.msm@gmail.com**Received:** July 24, 2017 | **Published:** February 28, 2019

**Abbreviations:** SJS, stevens johnson syndrome; SLE, systemic lupus erythematosus; NSAIDs, non-steroidal anti-inflammatory drugs; DMARDs, disease modifying anti-rheumatic drugs; PIP, proximal interphalangeal; MCP, Meta carpo phalangeal; CBC, complete blood count; UCE, urea creatinine electrolytes; CRP, C, reactive protein

## Introduction

Stevens Johnson Syndrome (SJS) is a severe dermatological condition commonly caused secondary to drugs or infections, with epidermal detachment involving less than 10 percent of the body's surface area.<sup>1</sup> Only a few cases have been previously reported in association with Systemic Lupus Erythematosus.<sup>2,3</sup> Systemic Lupus Erythematosus (SLE), is a multisystem, autoimmune disorder in which skin is the second most common organ involved.<sup>4</sup> There is more prevalence of SLE in females with the ratio being 10:1 when compared to males, especially females of fertile age group.<sup>5</sup> We report a similar case of SLE presenting with SJS in a 30 years old female.

## Case report

A 30 year old female of Asian descent with no known comorbid presented to the rheumatology clinic with complaints of an erythematous rash on her face for the past 4 months. The rash was associated with swelling of her lips. She also complained of recurrent oral ulcers, frequent hair fall, persistent high grade, fever and multiple well developed blisters and bullae on her mouth and limbs. Fever was associated with flu like symptoms and pain in multiple joints. The pain was gradual in onset, progressive in nature, moderate to severe in intensity involving the small joints of hand, wrist, knee and elbow joint bilaterally. She has been taking routine analgesics for this pain which proved to be of no therapeutic benefit. Patient had also been experiencing photosensitivity for past couple of months. No family history of any connective tissue disorder or autoimmune disease was present. No history of weight loss or night sweats was given by the patient. No involvement of genitalia and perianal region was present. On further evaluation, no history of drug use was evident. Patient was advised hospital admission.

Upon admission, on examination patient was febrile and found to be well oriented with time, place and person. Bilateral conjunctivitis was noted along with severe hemorrhagic cheilitis. Signs of synovitis were seen bilaterally in PIP joints, MCP joints, wrist, elbow, shoulder and knee joints. Mild restriction of movement was present. Presence of an erythematous rash over the cheek could clearly be appreciated along with blisters on her mouth.

Her initial investigation revealed a normal CBC, UCE and urinalysis. High CRP of 85mg/L was present. Cultures were done to rule out infection, which turned out to be negative. Autoimmune workup was done. RA factor was negative however, anti-CCP antibodies turned out to be positive (>200U/mL). ANA was homogeneously positive along with anti-Ds-DNA. Complement levels were within the normal limits. Hence, a diagnosis of SLE was formed.

Skin biopsy done revealed epidermal cell necrosis with perivascular lymphocytic infiltrate, suggestive of Stevens Johnson Syndrome. Appropriate supportive treatment was started for her SJS which encompassed gastric protective agents, conjunctival treatment, intravenous fluids, hydrogen peroxide soaks and neomycin ointment for topical application. The patient responded well to the treatment and started recovering. For SLE she was initially commenced with corticosteroid (prednisone) and Hydroxychloroquine. Steroids were gradually tapered off and Azathioprine was started. Patient was discharged and advised regular follow up visits.

## Discussion

The incidence of SJS is 2-3 cases per million populations annually in USA and Europe commonly affecting females as compared to males with a female to male ratio being 2:1. On the other hand, SLE occurs in 1 out of 2000 individuals and commonly affects women of childbearing age.<sup>6,7</sup> Half a million population in Europe and approximately one-fourth population of USA suffered from SLE.<sup>3</sup> Lee HY et al. have reported a few cases of SJS previously in the age group mentioned above in association with SLE.<sup>3</sup> In our case, patient was in fertile age period. Etiology of SJS in most patients remains unclear

and is mostly a result of adverse drug reaction but other uncommon causes have also been identified such as graft versus host reactions, infections, vaccinations and SLE.<sup>3</sup> In SJS, dermatological lesions are typically characterized by erythema, epidermal detachment, ulceration of skin areas and hemorrhagic erosions.<sup>8</sup> Although, drugs such as antibiotics and anticonvulsants are the most common cause of SJS,<sup>9</sup> no drug relation was established in our case; rather patient had complaints of multiple joint pains along with high grade fever and active synovitis. Later, she developed erythematous rash on her face followed by lip swelling and formation of blisters and bullae on face and ulcers in oropharyngeal cavity. There was no involvement of genitalia and perianal region in our patient, a feature suggestive of SLE associated SJS, also evident and reported by Lee HY et al. in their case report.<sup>3</sup>

Wetter DA et al. studied clinical features of SJS during an 8-Year Period at Mayo Clinic and reported that signs and symptoms related to eyes are commonly present in SJS patients such as photophobia, conjunctivitis, exudates and other ocular manifestations (including cicatrizing changes in the eye).<sup>1</sup> Our patient presented with conjunctivitis and photophobia however, after thorough eye examination, no other eye signs or symptoms were noted. She was prescribed topical eye drops and ointment for local application and with time, her eye symptoms improved and it was observed that overall our patient had responded well to therapy.

The percentage of deaths in relation with SJS accounts for 1-5%, as previously reported.<sup>10</sup> The best treatment in SJS is given to the patient by making prompt diagnosis, identifying the cause that led to the disease, providing good supportive care and ensuring that the patient receives treatment related to the causative agent.<sup>11</sup> Use of corticosteroid in SJS is debatable as previously investigators suggested that the use of corticosteroids has led to poor prognosis and the patient is more prone to infections.<sup>12</sup> But nowadays, it is suggested that the administration of dexamethasone is beneficial and effective for the patients with acute SJS,<sup>13</sup> but it does not preclude patients from receiving high dose IVIG, especially when SJS becomes severe.<sup>1,3,4</sup>

## Conclusion

SJS is a rare, life threatening cutaneous condition. To prevent the occurrence of complications, early diagnosis and prompt treatment is required. Fortunately, our patient did not develop any complication due to in time presentation and management.

## Consent

Written informed consent was obtained from the patient for publication of this case report.

## Acknowledgments

None.

## Conflicts of interest

The authors declare no conflicts of interest.

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