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Case Study

A Rare Association of Steven Johnson Syndrome with Lupus

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ABSTRACT

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Stevens-Johnson-Syndrome is a life-threatening dermatological conditions that is characterised by mucositis and epidermal detachment. The underlying etiology of this syndrome is almost invariably secondary to drugs but infrequently other causes such as lupus, infections and vaccinations have been observed. We report as such a rare clinical case presented at ER with gradual onset of dusky erythema at sun exposed areas, mucosal ulcerations with positive nikolsky sign, with history of no drug interaction. Complete blood paramenters showed pancytopenia with lupus +ve nature (laboratory investigations showed positive ANA, anti-ds-DNA). The patient was then diagnosed as systemic lupus erythematosus with initial presentation of steven johnson syndrome. It highlights new conduit in various treatment options and the achievement of a successful outcome. Although this syndrome is invariably associated with history of medications, but in rare instance may present as an initial presentation of lupus, particularly when associated with an initial photodistribution and a prolonged clinical course [1]. Hence, this short communication further illuminates the importance of timely treatment in such acute scenarios.

KEYWORDS:

Stevens-Johnson-Syndrome, Lupus

INTRODUCTION

The presence of steven-johnson-syndrome (SJS) is exceptionally rare in the lupus associated patients. This 21-yer-old female presented with complaint of rashes on her abdomen from three consecutive days. Additionally, the blisters observed on the face with mucosal involvement (Fig 1) questioned for presence of an additional pathological trigger. On further evaluation, the patient was conscious and oriented with no other systemic involvement. Moreover, the patient invariably complained of elliptical joint pain with/out presence of fever. Further lab results suggested hyponatremia with normocytic hypochromic anemia, high leukocyte counts which suggested presence of infection and inflammation as confirmed with up-surge in ESR levels. Following initial evaluations, the patient was admitted to intensive care unit for further clinical and lab evaluation with prompt initiation of the treatment.

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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.

CLINICAL PRESENTATION

Following admission, the patient's blood samples were collected and sent for lupus serology, as suspected in a young girl with polyarthopathy. The results were absolutely enticing with high ANA positivity (3.7 U/l, range <1). Positive ANA suggests rheumatoid, mixed connective tissue disease, scleroderma or systemic lupus erythematosus (SLE). To further confirm the diagnosis ds-DNA was measured and as suspected it came out to be positive (138.7 IU/ml).

On clinical inspection the patient presented with a diffuse blister type lesions on the oral cavity with target lesions on the body surface, specifically abdomen and arms (Fig 2). With time these lesions further changed morphologically and captured larger area of involvement (Fig 1). Moreover, international normalised ratio (INR) was within limits with normal D-dimer test to rule out any coagulopathy if

associated in the presence of lupus. Pulmonology, cardiology and renal parameters were in range.

She was initiated on one proton-pump inhibitor with broad spectrum antibiotic (meropenem) 1 gram twice a day and torsemide 20 mg once a day until the blood results were obtained. Once all results were in hand, we commenced linezolid 600 mg 12 hourly in contrast to meropenem with administration of corticosteroid 20 mg once a day. Furthermore, fucidin ointment was prescribed every 8 hourly. These above-mentioned treatments continued for next 14 days with timely improvement in the patient condition (Fig 3 & 4). Following a successful recovery she was discharged with tapering of steroids, multivitamins, gastric protectors. She was informed and booked in for monthly appointments with the concerning consultants to monitor her progress and well-being.

DISCUSSION

With speculated pathogenesis of SJS it is more efficient to diagnose it on clinical basis. The T-cell mediated reactions with involvement of CD8+ cells which are coincidently show their presence in the blister fluid and may further induce cellular apoptosis. Additionally, the attendance of CD40 cells may play a dominant role in inducing the release of tumor necrosis factor-alpha (TNF-α) which again would further induce cellular apoptosis in the presence of Th1 and Th2 cytokine response [2]. Understandably, these T cell receptors bind to various drug-immune system reactions.

There have been trivial cases reported in regard to the relation between SJS and lupus in the past. Usually it is anti-epileptics responsible for development of erythema multiforme exudativum SJS in lupus patients [3]. In our presentation where the patient has no drug history whatsoever with SCORTEN score of 1, opens a new chapter where lupus associated SJS could a direct presentation. Furthermore, Wetter DA et al. evaluated clinical features of SJS in a 8-year-old girl which showed photophobia, conjunctivitis with other ocular manifestations [4]. In relation to this our patient presented with photophobia with absolutely normal eye examination. Mukkarram et al. presented a similar case in a 30-year-old female which was diagnosed with SLE and later following initiation of the treatment she developed SJS [5]. In correlation to this, our patient did not present with any drug history, hence justifying that there is immune reaction associated triggers playing a parallel role in development of the pathogenesis.

Figures

Figure 1. Figure 1: Clinical picture demonstrating blister type mucosal involvement in a lupus positive 21-year-old-female who presented with systemic illness as evidenced by presence of Steven-Johnson-Syndrome.

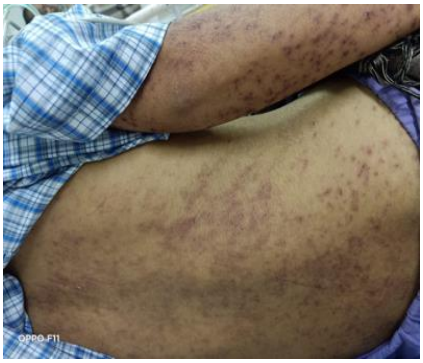

Figure 2: Clinical picture demonstrates widespread early morbilliform eruptions in a lupus positive-SJS patient.

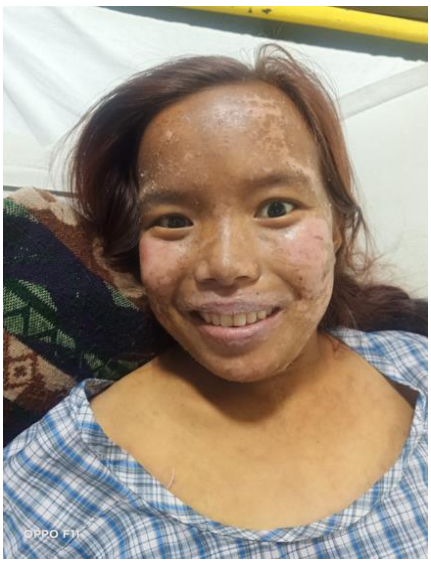

Figure 3: Day 12 of post-treatment picture of a lupus associated SJS patient with 90% recovery rate and total abolition of the blisters.

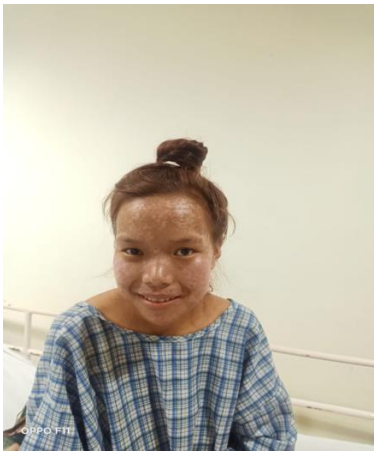

Figure 4: Day 20 of post treatment, where full recovery can be observed with absolute decrease in erythema in a lupus-associated SJS patient.

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