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Case Report

An Indonesian female with Stevens-Johnson syndrome mimicking cutaneous lupus: A case report

Nadya Meilinar Samson, Awalia *

Department of Internal Medicine, Faculty of Medicine, Universitas Airlangga – Dr Soetomo General Academic Hospital, Surabaya, Indonesia

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ABSTRACT

Background: Diagnosing bullous eruptions in systemic lupus erythematosus (SLE) patients is challenging because there are so many differential diagnoses, including drug reactions such as Stevens-Johnson syndrome (SJS)/toxic epidermal necrolysis (TEN) and TEN-like cutaneous lupus erythematosus (LE).
Case presentation: An Indonesian female with a SLE history complained of an erythematous rash over the body 2 weeks ago. A pulse dose of methylprednisolone was given, and after 3 days, of getting bullae appeared all over the body and treatment stopped. The patient is treated in collaboration with dermatology, ophthalmology, and allergy consultants to obtain a diagnosis of SJS supporting skin biopsy. The patient was given methylprednisolone 62.5 mg once daily for 7 days, Gentamicin 80 mg twice daily, and the skin lesion was treated with NaCl 0.9% compression. The patient showed improvement and decreased methylprednisolone dose to 16 mg 3 times a day.
Discussion: Skin biopsy must be obtained to establish the diagnosis between JSJ/TEN and cutaneous LE.
Conclusion: SJS/TEN can occur in SLE patients. The correct diagnosis can reduce the patient's mortality and morbidity.

1. Introduction

Acute cutaneous lupus erythematosus (ACLE) lesions can be described as localized, usually referring to the malar rash, or generalized, involving diffuse macular or maculopapular erythema, often in a photosensitive pattern [1,2]. The most severe form of ACLE is Stevens-Johnson syndrome (SJS)/toxic epidermal necrolysis (TEN)–like vesiculobullous eruption [1,3,4]. SJS and TEN are an uncommon, acute adverse cutaneous drug reactions with high morbidity and mortality [5]. It was reported that 6.3 out of 100,000 people experiencing SJS/TEN [6,7]. The diagnosis of lupus-induced SJS/TEN and classical SJS/TEN is complex and compelling because in both cases, the classical clinical and histopathological features will be present, with epidermolysis reactions and skin detachment due to inflammatory dermatoses with keratinocyte apoptosis and the exact molecular mediators involved [5,8]. We report an Indonesian female with SJS mimicking cutaneous lupus erythematosus (LE).

2. Case presentation

An 18 years old Indonesian female, Javanese ethnic, complained of an erythematous rash all over the body from 2 weeks ago. Initially, the erythematous rash appears on the face forming a butterfly rash, then slowly appears all over the body. The rash is not painful nor itchy. She has never had a rash on her face before and any mouth ulcer. There was joint pain, fever, nausea without vomiting, increased fatigue, hair loss, and decreased appetite for 2 weeks. The fever is sub-febrile, fluctuating, and persists for 2 weeks despite consuming paracetamol. Medical history found prolonged diarrhea (3 weeks), ANA test of positive with low complement, and the patient received pulse dose corticosteroid therapy 3 months ago. 2 months later, the patient underwent upper endoscopy and was diagnosed with pan gastritis erosive. The patient receives sulfasalazine 1000 mg twice daily and ibuprofen 400 mg twice daily. Patient and family were no history of diabetes mellitus, hypertension, kidney disease, liver disease, malignancies, allergy or drug allergy.

Physical examination showed a patient weakness with a heart rate of 107 × /min. The skin efflorescence is a fixed erythematous rash over the

* Corresponding author. Department of Internal Medicine, Faculty of Medicine, Universitas Airlangga – Dr Soetomo General Academic Hospital, Jl. Mayjend Prof. Dr. Moestopo No. 6-8, Airlangga, Gubeng, Surabaya, East Java, 60286, Indonesia.
E-mail address: awalia@fk.unair.ac.id (Awalia).

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cheeks and nasal bridge, spares the nasolabial fold, with multiple erythematous rashes on the upper anterior thoracic region. The patient was also anemic and had a malar rash (Fig. 1). Laboratory investigation showed Hb of 10 g/dL, WBC of 2,290 10^3/μL, and platelet of 10^3/μL. Meanwhile, urine analysis showed 2+ protein and positive erythrocytes. Chest X-ray were within normal limit. The patient received therapy with high calories high protein 1900 kcal/day, NaCl 0.9% of 1500 mL/24 hr, Methylprednisolone injection of 500 mg 1 × /day, and Paracetamol 3 × 500 mg orally.

On the second day, fever, nausea, and fatigue were slightly better but no improvement in the patient's rash. Laboratory examination showed C3 of 39.5 mg/dL, C4 of 16.8 mg/dL, ESR of 66 mm/hour, and procalcitonin of 0.18 ng/mL. On the thrid day, the patient started getting blisters, starting in the face and spreading all over her body, soreness on the lips and body accompanied by pain in swallowing. The skin efflorescence was multiple erythematous macules with well-defined margins, round shape, confluent, and variable size. In the facial, cervical, right, and left brachial, and anterior thoracic regions, multiple blisters filled with clear fluid and loose walls, without any pus or erosions with positive Nikolski's sign (Fig. 2).

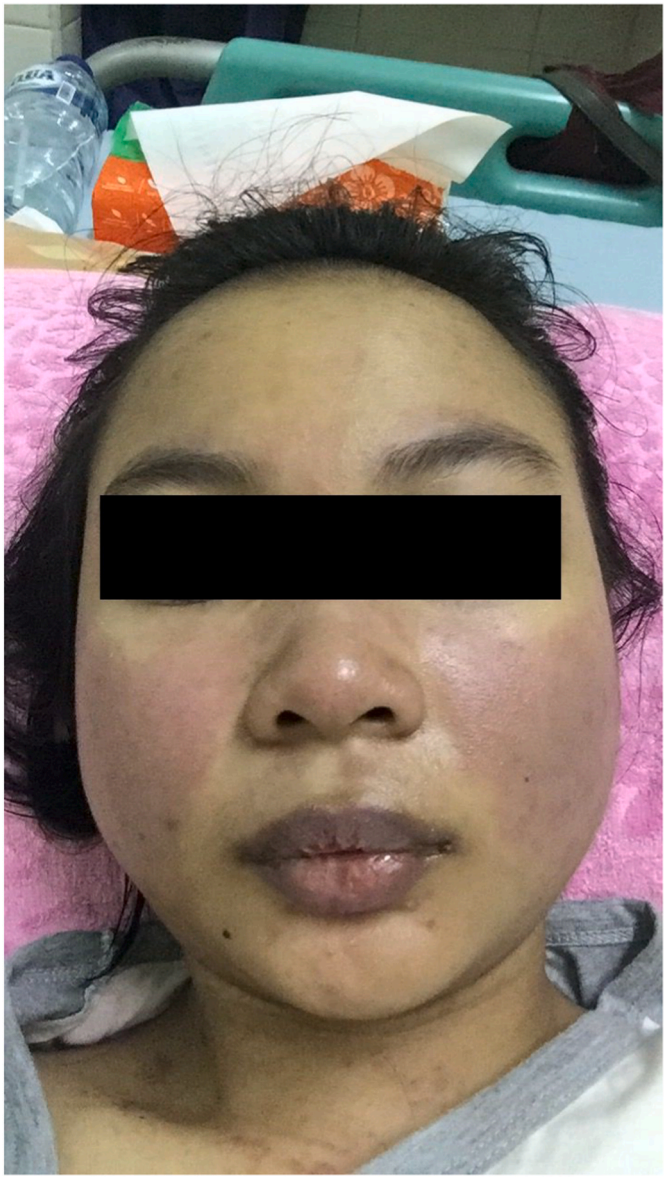

Fig. 1. Patient condition when admitted to the hospital.

The patient then collaboration with dermatology, ophthalmology, and allergy consultant, which diagnosed the patient with SJS (SCORTEN 1) caused by suspect paracetamol, ibuprofen, and/or sulfasalazine. The skin biopsy result was a subepidermal blister with epidermal necrosis, suggested SJS. The patient stopped the suspected drugs and received 1 × 62.5 mg for 7 days. On the 5th day, the blisters became crusted and had a secondary bacterial infection with pus (Fig. 3). The patient became sepsis due to secondary infection and received Gentamicin 80 mg of 2 × /day, and the skin lesion is being wound care with NaCl 0.9%. On the 8th day, Skin recovery and re-epithelialization were established, the temperature decreased, and mucosal complications stabilized. There was residual pigmentation on the site of skin lesions before. On the 9th day, the patient was changed to methylprednisolone 16 mg of 3 × /day and the patient was discharged on day 9 with improving clinical manifestation. Patients showed significant improvement after 6 months of outpatient (Fig. 4).

3. Discussion

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are uncommon, acute adverse cutaneous drug reactions with high morbidity and mortality. SJS can affect anyone with a genetic predisposition: any age, sex, and all races, although it is more common in older people and women. Classification is based on three clinical criteria: individual skin lesions' pattern, distribution, and the maximum extent of the epidermal detachment. Detachment of <10% of the body surface area (BSA) is defined as SJS, 10–30% as SJS/TEN-overlap and >30% as TEN [3,5].

SJS and TEN starts with prodromal symptoms up to 1 week, such as fever, sore throat, coughing, eye burning, myalgia, and arthralgia. Selanjutnya, discrete maculopapular rash, similar to a morbilliform rash. The rash begins on the trunk with subsequent generalization, usually sparing the palmoplantar areas. Macular lesions become purplish, and epidermal detachment occurs, resulting in flaccid blisters that converge and break, resulting in extensive sloughing of necrotic skin. SJS/TEN was the onset of atypical target lesions and/or erythematous or purpuric macules followed by the detachment of epidermis and epithelium, causing extensive denuded skin areas, necrosis, mucosal erosions, usually with the presence of Nikolski's sign. There is an interval time for susceptible drugs triggering hypersensitivity reactions, ranging from 4 to 28 days between the beginning of drug use and the occurrence of signs and symptoms. The highest risk of developing SJS and TEN occurs continuously in the first 2 months of treatment with risk drugs [5,10].

The diagnosis of lupus-induced SJS/TEN and classical SJS/TEN is difficult. Lupus-induced SJS/TEN was a subacute presentation of weeks without systemic involvement and no history of drug ingestion [11]. Classical TEN was acute evolution within 3–4 days or sometimes hours, with a close drug-related causality and negative immunofluorescence [12]. Although the physical signs suggest a diagnosis of SJS/TEN, histopathology of a skin biopsy is necessary to support the clinical assessment and exclude other blistering dermatoses. The histopathological hallmark of these diseases is widespread epidermal necrosis due to death by apoptosis of keratinocytes [3].

Prophylactic antibiotic therapy may be considered for widespread skin involvement and the slightest clinical suspicion of sepsis. Empirical coverage should include one antibiotic with anti-staphylococcal activity (amoxicillin + clavulanic acid, tetracyclines, vancomycin, clindamycin, teicoplanin, linezolid), Gram-negative activity (amikacin, piperacillin + tazobactam, cefoperazone + sulbactam, imipenem), and anaerobic activity such as metronidazole and tinidazole. However, gentamicin was chosen because allergic reactions and drug eruptions caused by aminoglycosides reported in the literature are rare [13].

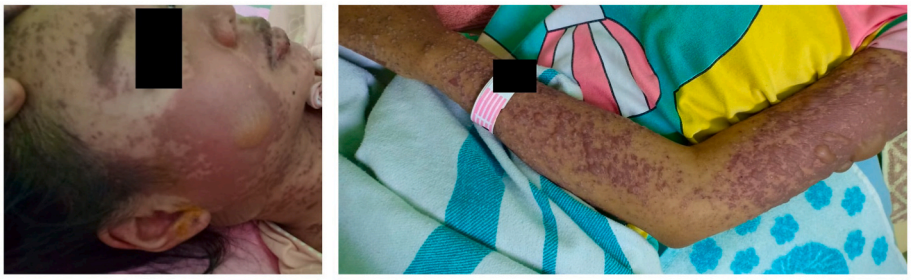

Fig. 2. Bullae appear all over the body on the 3rd day.

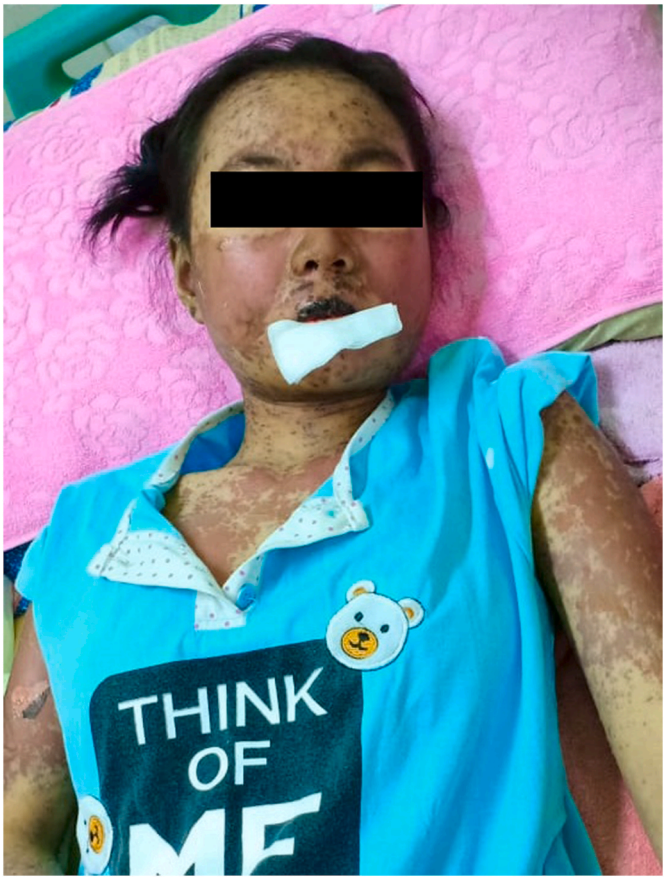

Fig. 3. The patient had secondary infection post bullae bursts on the 5th day.

Fig. 4. After 6 months of routine control, the patient's skin become better and healthy.

4. Conclusion

We reported an 18 years old Indonesian female, with SLE high activity disease with cutaneous lupus manifestation and occurrence of SJS because of suspected drugs of sulfasalazine, paracetamol and ibuprofen. The SJS can occur because of drugs, infections and malignancies. However, it can occur in SLE patients. The skin biopsy revealed a subepidermal blister with epidermal necrosis, which suggested SJS. The patient then stopped the suggested drugs, treated with steroids, wound dressing, and improved clinically. The patient then discharged and advised not to manipulate the lesions, avoid sun exposure, use sun protection lotion, avoid suspected drugs, and routinely control to rheumatology and dermatology clinics.

Ethical approval

Not applicable.

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Author contribution

All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

Registration of research studies

Name of the registry: -.
Unique Identifying number or registration ID: -.
Hyperlink to your specific registration (must be publicly accessible and will be checked): -.

Guarantor

Awalia.

Consent

Written informed consent was obtained from the patient or guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Declaration of competing interest

Nadya Meilinar Samson and Awalia declare that they have no conflict of interest.

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Further reading

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