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Case Report

Generalized papular rash as a rare sentinel sign of pediatric lupus: Hyper-IgE in a 13-year-old girl

Natalia Erica Jahja[1,2], Zahrah Hikmah[1,2]*, Willy Sandhika[3], Anang Endaryanto[1,2]

[1]Dept. of Child Health, Dr Soetomo General Academic Hospital, Surabaya, Indonesia
[2]Dept. of Child Health, Faculty of Medicine - Universitas Airlangga, Surabaya, Indonesia
[3]Dept. of Anatomical Pathology, Faculty of Medicine – Universitas Airlangga, Surabaya, Indonesia

Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disorder with diverse dermatologic manifestations. This case report describes a 13-year-old Indonesian girl presenting with generalized erythematous papules, a rare cutaneous sentinel of pediatric SLE. Despite markedly elevated immunoglobulin E (IgE) levels (>22,000 IU/mL) mimicking hyper-IgE syndrome, histopathological confirmation of subacute cutaneous lupus erythematosus (SCLE) and renal involvement established the diagnosis of SLE. Elevated IgE initially obscured the diagnosis, its association with severe immune dysregulation underscored the need for awareness in pediatric SLE. Treatment with intravenous methylprednisolone, mycophenolate sodium, and hydroxychloroquine led to rapid resolution of skin lesions and renal improvement. This case highlights atypical skin lesions as early signs of systemic autoimmunity, requiring dermatopathological and renal function evaluation.

Keywords: Pediatric lupus, Hyper-IgE, Subacute cutaneous lupus erythematosus, Generalized papular rash.

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1. Introduction

Generalized skin rashes in children are frequently seen at primary care settings and are a main reason for pediatric or dermatologist consultation.[1] Generalized papular rashes in pediatric systemic lupus erythematosus (SLE) are often misdiagnosed as psoriasis or drug reactions, delaying systemic evaluation.[2] *While malar rash and photosensitivity are classic, atypical cutaneous manifestations, like generalized papules, are underrecognized and may* need histopathological confirmation to establish the diagnosis.[1,3] The common etiologies of generalized rashes are viral exanthema, drug allergies, autoimmune disorders (for example, SLE) and rare etiologies, such as systemic diseases (e.g., Still's disease) or genetic disorders (hyperimmunoglobulin E syndrome [HIES]).[1] Serologic test, comprehensive hematology panel, and skin biopsy are recommended if the rash is persistent.[3]

This case report describes an Indonesian child, female, aged 13 years old exhibiting generalized erythematous skin as well as widespread papules as the initial and rarely reported manifestation of SLE. The diagnosis was complicated by markedly elevated immunoglobulin E (IgE) levels, which initially led to differential diagnoses of allergy or HIES.

2. Case Presentation

An Indonesian child, female, aged 13 years old, visited Dr. Soetomo General Academic Hospital with a three-year history of chronic, non-pruritic, recurrent rash, and hair loss (**Figure 1**). She had no history of allergies and was previously treated with antiallergic medications, but her symptoms worsened in 2024. She is the eldest of two siblings, with no family history of similar conditions.

The erythematous papules exhibited a psoriasiform morphology with central hypopigmentation, distributed

*Corresponding author: Zahrah Hikmah
Email: dr.zahrah.hikmah@fk.unair.ac.id

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symmetrically on sun-protected areas (**Figure 2**). Absence of scaling or mucosal involvement further complicated initial differentiation from lichenoid drug eruption. The patient reported no fever, joint inflammation, mucosal involvement, or any systemic symptoms. A positive antinuclear antibodies (ANA) test prompted referral for further evaluation. The histopathological examination confirmed subacute cutaneous lupus erythematosus (SCLE) diagnosis (**Figure 3**).

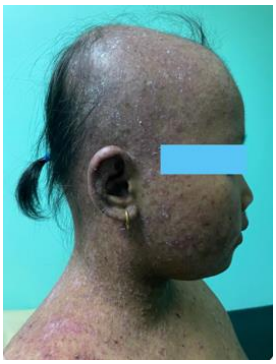

Figure 1: On the face, well-demarcated hyperpigmented plaques, consistent with discoid lupus, were observed, prominently involving both earlobes. Note the absence of malar rash or mucosal lesions.

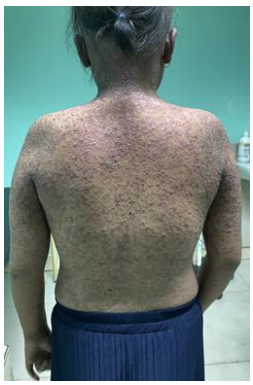

Figure 2: Widespread erythematous papules with psoriasiform scaling on the trunk and extremities.

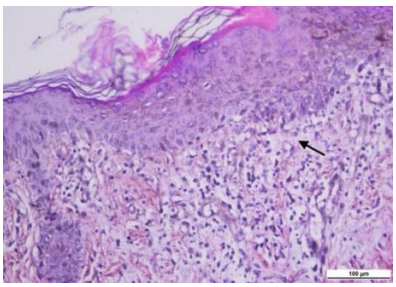

Figure 3: Histopathological findings revealed vacuolar degeneration of the basal epidermal layer and lymphocytic infiltration at the dermo-epidermal junction, consistent with interface dermatitis (black arrow), indicative of SCLE.

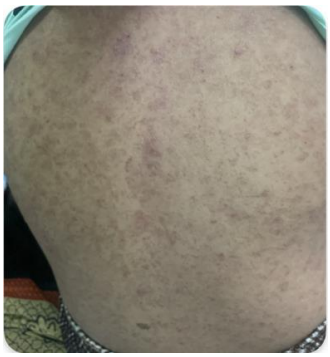

Figure 4: Notable reductions in the severity of lesions were observed after intensive therapy, with remaining erythematous papules starting to diminish on back.

Figure 5: Discoid lupus lesions on both earlobes showed signs of fading. Also, the hair loss improved after intensive therapy.

One of the exceptional primary immunodeficiency disorders, HIES, manifests as a markedly increased level of serum IgE, relapsing staphylococcal skin abscesses, severe pruritic eosinophilic dermatitis, and recurrent sinopulmonary infections.[4] In this case, the patient exhibited elevated IgE levels but lacked the characteristic clinical features of HIES, such as frequent cutaneous or pulmonary infections. Consequently, the patient underwent comprehensive diagnostic evaluations, including immunological tests, renal function tests, and routine urinalysis, with detailed results provided in **Table 1**. According to the 2012 Systemic Lupus International Collaborating Clinics (SLICC) criteria, three clinical conditions as well as two immunologic criteria, were met. Furthermore, the patient also fulfilled the 2019 European League Against Rheumatism (EULAR) criteria**,** achieving a total score of 12. The Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) revealed severe activity with a score of 14 points, as detailed in **Table 2**.

A diagnosis of severe SLE with elevated IgE was confirmed. Treatment included methylprednisolone (MP) pulse therapy (30 mg/kg/day), mycophenolate sodium and hydroxychloroquine. Prednisone (1 mg/kg/day) was administered following the completion of a 3-day MP pulse

regimen. After three weeks of therapy, the patient showed clinical (**Figure 4**, **Figure 5**) and laboratory improvement (**Table 3**). Given the relapsing nature of SLE, with frequent disease flares and limited instances of sustained remission, regular monitoring of disease activity and renal function is essential. The long-term prognosis for this patient remains uncertain.

The patient was asked to sign an informed consent form regarding the case report and images publication.

Table 1: Summary of initial laboratory findings.

Parameter	Result	Reference Value
Hematological		
Hemoglobin	13.1	12–16 g/dL
Leukocyte	6.54 x 10^3/µL	3.37 – 10.0 x 10^3/µL
Thrombocyte	205 x 10^3/µL	150 – 450 x 10^3/µL
Erythrocyte Sedimentation Rate (ESR)	54 mm/h	<20 mm/h
Eosinophils	8.3%	0–6%
Renal Function		
Urea	11 mg/dL	7–20 mg/dL
Creatinine	1.2 mg/dL	0.5–1.1 mg/dL
Glomerular Filtration Rate (GFR)	69 mL/min/1.73 m²	>90 mL/min/1.73 m²
Immunology		
ANA	366 AU/mL	<20 AU/mL
Anti ds-DNA	15.7 IU/mL	< 30: negative
C3	20 mg/dL	90–180 mg/dL
C4	0.3 mg/dL	10–40 mg/dL
IgE	22,915 IU/mL	<100 IU/mL
Urinalysis		
Proteinuria	4+	Negative
Albumin-Creatinine Ratio	Macroalbuminuria (≥150)	<30 mg/g Cr
Leukocyte	20.4/hpf	<5/hpf

ANA = antinuclear antibodies; AU = arbitrary unit; ds-DNA = double-stranded deoxyribonucleic acid; hpf = high power field; IgE = immunoglobulin E; IU = International Unit.

Table 2: Diagnostic and severity criteria of SLE.

Criteria	Details	Score/Points
2012 SLICC	**Clinical conditions**	
	– Chronic cutaneous lupus	3 clinical conditions 2 Immunologic criteria
	– Renal involvement	
	– Non scarring alopecia	
	Immunologic criteria	
	– Positive ANA	
	– Hypocomplementemia	
2019 EULAR	SCLE (4)	12
	Proteinuria >0.5 g/24h (4)	
	Low complement levels (4)	
SLEDAI	Proteinuria (4)	14
	Pyuria (4)	
	Alopecia (2)	
	Rash (2)	
	Low complement levels (2)	

Table 3: Comparison of laboratory parameters pre- and post-treatment.

Parameter	Pre-Intervention	Post-Intervention Week 3
Hematological		
Erythrocyte Sedimentation Rate (ESR)	54 mm/h	12 mm/h
Renal Function		
Urea	11 mg/dL	8.6 mg/dL
Creatinine	1.2 mg/dL	0.7 mg/dL
Uric Acid	9.4 mg/dL	2.5 mg/dL
Glomerular Filtration Rate (GFR)	69 mL/min/1.73 m²	118 mL/min/1.73 m²
Urinalysis		
Proteinuria	4+	1+
Albumin-Creatinine Ratio	Macroalbuminuria (≥150)	80 mg/g Cr
Leukocytes	20.4/hpf	0.5/hpf

Hpf = high power field

3. Discussion

A challenging autoimmune disorder, SLE, has a broad continuum of clinical signs and symptoms. Pediatric SLE, particularly, presents unique challenges, with an estimated annual case reaching 3.3–24 patients/100,000 children. While SLE classic cutaneous features include malar rash and photosensitivity, atypical skin presentations, such as generalized papular eruptions, are rare and often lead to diagnostic delay.[5] It has a multifactorial pathogenesis, involving genetic, hormonal, and environmental influences.[6] This case highlights generalized papules as the sentinel sign of SLE, emphasizing the importance of dermatological vigilance in pediatric patients.

Cutaneous manifestations are often the initial presentation of SLE. SCLE has two morphologic variants: annular and papulosquamous. A study of 58% SCLE patients found 42% had annular lesions, 39% had papulosquamous lesions, and 16% exhibited both. The papulosquamous variant may mimic eczema, psoriasis, or pityriasis. SCLE lesions typically appear in sun-exposed areas, such as the upper thorax (V-distribution), upper back, and extensor surfaces of the arms.[6] In this case, the lesions were distributed diffusely across the trunk, including sun-protected areas, suggesting that the exacerbated condition may be linked to elevated IgE levels.

SLE pathogenesis involves impaired apoptotic cells' elimination and immune tolerance's failure to the antigens of the nuclei. These antigens trigger the autoreactivity of B and T-cells, leading to autoantibody generation, which predominantly of the IgG isotype. This autoantibody formed circulating immune complexes (CICs) when bound to autoantigens and complement proteins, where at various target organs, initiating tissue inflammation and damage.[7] Previous studies have demonstrated that increased IgE production, due to a disrupted immune tolerance, can trigger polyclonal activation of B lymphocytes and abnormal Th2 responses, accompanied by the generation of autoantibodies.[7,8] Autoreactive IgE contributes to disease progression by activating basophils and plasmacytoid dendritic cells (pDC). When this autoantibody binds to double-stranded DNA (dsDNA) and crosslinks with the high-affinity IgE receptor (FcεRI) on basophils, it triggers degranulation, leading to the release of cytokines (including BAFF, IL-6, and IL-4) as well as lipid mediators. These induce B cells differentiating into plasma cells, which further increase the autoreactive IgE and IgG antibodies production.[7,9] Thus, autoreactive IgE exacerbates immune dysregulation in SLE, while non-autoreactive IgE may exert a protective effect by suppressing this activation. A key indicator of IgE-mediated immune dysregulation is the activation of the complement cascade, resulting in hypocomplementemia.[7,9,10]

This patient met the diagnostic criteria for SLE and significantly elevated IgE levels were detected, alongside reduced complement C3 and C4 levels. These findings suggest that elevated IgE levels reflect severe Th2-predominant immune dysregulation in lupus, which presented as an atypical dermatological manifestation in this case. This presentation could be derived from extensive inflammation and IgE-mediated immune dysregulation, leading to keratinocytes necrosis and renal disorders.

About half of SCLE patients fulfil SLE criteria, but systemic symptoms are typically mild. Additionally, SCLE is rare in children.[6,11] This patient demonstrated normotension, severe proteinuria without accompanying edema, and pyuria as the primary renal manifestations, which resolved after intensive therapy. This case underscores the need for routine renal function tests in all children with unexplained cutaneous rashes, even without overt renal symptoms.

The management of pediatric SLE aims to control disease activity, prevent organ damage, and minimize treatment-related toxicity.[5] In this case, the patient responded well to high-dose intravenous MP, mycophenolate sodium,

and hydroxychloroquine, with significant improvement in both cutaneous and renal manifestations.

4. Conclusion

Generalized papular eruptions, though rare, may be a sign of pediatric SLE. Either dermatologists or pediatricians should consider lupus in children with unexplained rashes, particularly with non-pruritic, psoriasiform lesions. Early skin biopsy and renal screening are pivotal to establish the diagnosis and unmask silent systemic involvement.

5. Author Contributions

ZH supervised the manuscript, provided critical revisions, and guided the diagnostic process. AH critically reviewed and corrected the manuscript, ensuring scientific accuracy. NEJ drafted the manuscript, compiled the data, and structured the case presentation and discussion. WS performed histopathological examinations and interpreted microscopic findings. All authors reviewed and approved the final version of the manuscript.

6. Source of Funding

None.

7. Conflict of Interest

All authors affirm no conflict of interest concerning this report.

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