CJIM | Case Report

Successful abortive treatment with cyclosporine in a pediatric patient with Stevens-Johnson syndrome

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Statement of Significance:

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are rare, life-threatening dermatologic drug reactions with no standardized therapies. Recent evidence suggests cyclosporine may reduce mortality in SJS/TEN patients, but only a few case reports have demonstrated its use in pediatric populations. Our report describes a case of early pediatric SJS successfully treated with cyclosporine. We propose that cyclosporine be considered as an abortive therapy in early pediatric SJS and may halt significant disease progression.

DOI: 10.47265/cjim.v3i1.4140

Introduction

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are a spectrum of dermatologic emergencies characterized by life-threatening diffuse skin necrosis and sloughing. There are currently no standardized therapeutics for SJS/TEN, but evidence collected primarily in adults suggests that cyclosporine may improve mortality.[1,2] We report a case of SJS in a 17-year-old girl in whom early cyclosporine treatment resulted in rapid cessation of disease progression.

Case Report

A previously healthy 17-year-old girl presented to the emergency department with erythematous papules and plaques involving her extremities and trunk. Two weeks prior, she began taking trimethoprim-sulfamethoxazole (TMP-SMX) for medication-resistant acne. She was not on any additional new or prior medications. Two days prior, she noted lip pain and swelling and developed a facial rash, which subsequently spread to her trunk and extremities. She endorsed oral ulcers, odynophagia, dysphonia, conjunctival erythema, ocular pruritis, and dysuria. She immediately self-discontinued TMP-SMX after initial appearance of the rash.

On admission, she was afebrile, but tachycardic (HR 120s). Complete blood count and complete metabolic panel were significant only for a mildly decreased absolute lymphocyte count of 1.3 x 10^9(normal range 1.5-5.0 x 10^9) and mildly elevated aspartate aminotransferase of 39 U/L (normal range 5-30 U/L). She did not have eosinophilia or leukocytosis. Her exam was notable for erythematous papules and plaques involving the face, trunk, and bilateral upper and lower extremities including the palms without

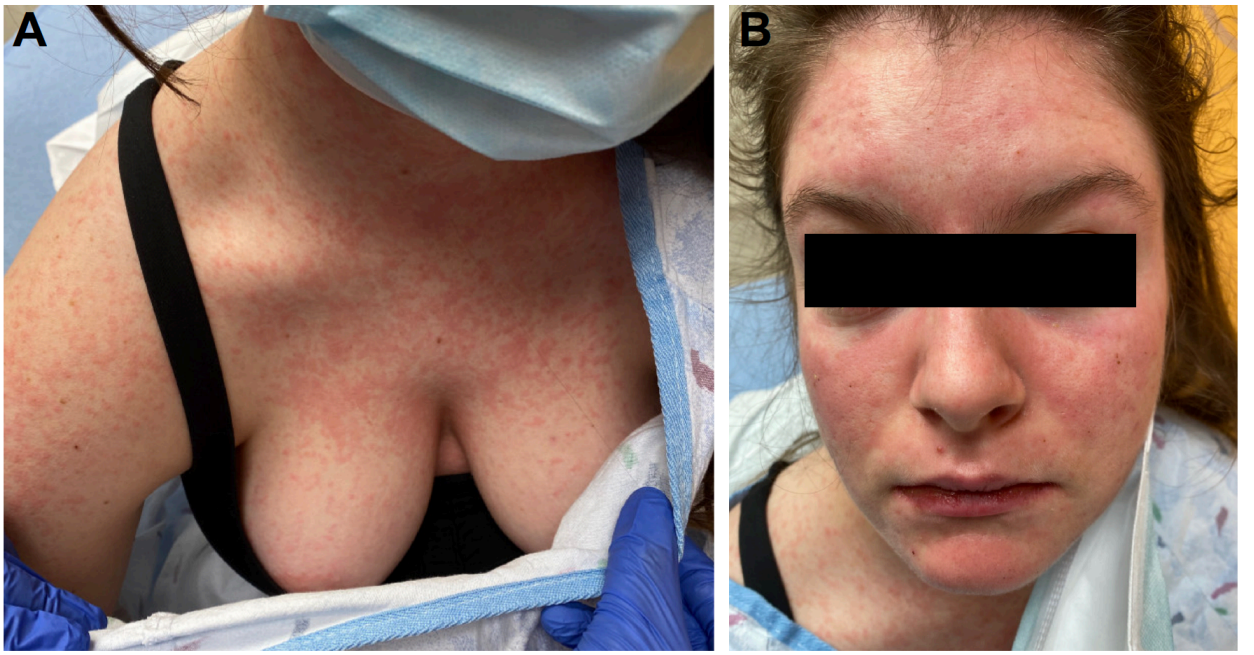

Figure 1. Clinical appearance of papules and plaques on trunk (a) and face (b) at admission prior to administration of cyclosporine. Erosions and crusting are present on the lips.

evidence of sloughing (Fig. 1). Hemorrhagic crusting and erosions were present on the lips and confluent erosions were present on the hard palate and frenulum of the tongue. Genital exam was notable for erosions of the clitoris and posterior fourchette. Ophthalmologic exam was significant for bilateral conjunctival injection and ~15% bilateral conjunctival fluorescein staining without bulbar staining. She was diagnosed clinically with SJS given cutaneous exam findings, mucositis of three sites, and timing of the eruption following the culprit drug and a skin biopsy was deferred. Her SCORTEN[3] on admission was 1 (heart rate >120) and she started oral cyclosporine 5 mg/kg/day divided twice daily. Her lesions improved within twenty-four hours (Fig. 2) and she was discharged two days after presentation on one week of oral cyclosporine at 5 mg/kg/day followed by one week at 2.5mg/kg/day. One week after discharge, her lesions had largely resolved without any discernable oral or ocular involvement.

Discussion

Here we describe a pediatric case of early SJS treated with oral cyclosporine. While previous studies have suggested that cyclosporine may reduce mortality in SJS/TEN in adults,[1,2] few have examined cyclosporine in pediatric SJS patients. Additionally, most pediatric case reports examined children with treated with cyclosporine in combination with other therapeutics, most often corticosteroids and/or IVIG.

Our patient presented two days after initial rash onset and cyclosporine was initiated on day of presentation. In contrast to prior pediatric case reports [4,5] involving patients with severe disease (skin necrosis >10% body surface area), our patient started cyclosporine prior to epidermal necrosis and did not develop subsequent necrosis. She was discharged after two days compared to the average of 9.4 days in pediatric SJS.[6] Her rapid

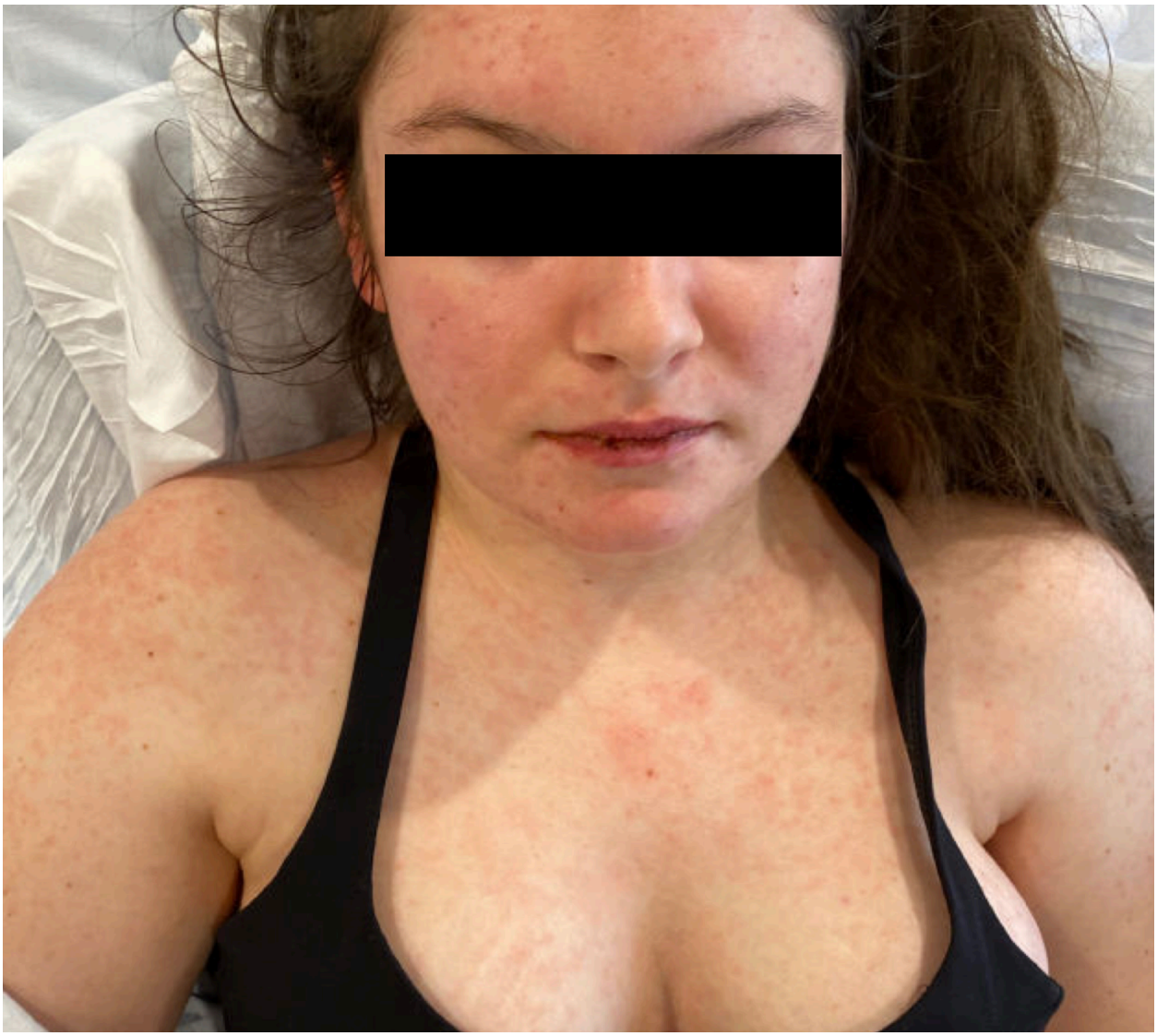

Figure 2. Clinical appearance of lesions on trunk and face one day after presentation.

cutaneous improvement suggests that cyclosporine may function as a well-tolerated abortive therapy in early SJS. The underlying pathophysiology of SJS/TEN includes massive keratinocyte necrosis induced by cytotoxic T-cells, NK cells, TNF-a, and granulysin.[7] Inhibition of T-cell activation by cyclosporine thus prevents accumulation of cytokines and downstream mediators of SJS/TEN. Our experience expands on previous reports and demonstrates early treatment with cyclosporine prior to skin necrosis may prevent T-cell mediated disease progression and hasten recovery. Larger studies will be necessary to confirm our findings demonstrating effective use of cyclosporine in early pediatric SJS.

ARTICLE INFORMATION

Accepted for Publication: July 23 2023.
Published Online: September 22 2023.
DOI: 10.47265/cjim.v3i1.4140

Cite this article: Dinh Timothy A, Bode Helen P, Nieman Elizabeth L, Zwemer Eric K, Ziemer Carolyn M. Successful abortive treatment with cyclosporine in a pediatric patient with Stevens-Johnson syndrome. Carolina Journal of Interdisciplinary Medicine (CJIM) 2023;3(1):56-59.

Conflict of Interest Disclosures: The authors have no conflicts of interests to declare.

Funding/Support: None

Ethics Approval and Consent to Participate: Informed consent was obtained from the parent/patient to publish photographs submitted with this manuscript.

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