DOI: 10.1111/pde.13820

WILEY Pediatric Dermatology

Idiopathic toxic epidermal necrolysis in an adolescent

Abstract

A 10-year-old girl, suspected 2 days prior to have streptococcal pharyngitis, presented with diffuse erythema, tense bullae, Nikolsky-positive desquamation, as well as ulcerations of her oral and genital mucosa. She denied recent travel, sick contacts, or preceding and concurrent use of medications, including over-the-counter and herbal supplements. A comprehensive viral polymerase chain reaction (PCR) panel, *Mycoplasma pneumoniae* PCR and IgM, streptococcal molecular antigen test, urine culture, blood culture, and rheumatologic serologies were negative. Based on the patient's clinical presentation and biopsy results, she was diagnosed with idiopathic toxic epidermal necrolysis.

1 | INTRODUCTION

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are severe and potentially fatal mucocutaneous diseases.[1] In the United States, the estimated incidence of TEN among children is 0.4/1 000 000 per year.[2,3] Medications and infectious agents are the most common reported causes. We report a challenging diagnosis of TEN with uncertain etiology in a pediatric patient.

2 | CASE

A 10-year-old girl initially presented with acute onset of throat pain, fever, rash, and erythematous tonsils with white exudate. She denied the use of any medications, including over-the-counter and herbal supplements. There was no history of medication allergies, recent travel, sick contacts, or preceding illness. Oropharyngeal rapid group A streptococcus antigen test was negative, and she was discharged on an empiric course of amoxicillin; however, the prescription was never filled.

Two days later, she returned febrile and tachycardic. She had diffuse erythema, tense bullae, and Nikolsky-positive desquamation covering 80% of her body surface area, as well as severe oral and vaginal mucositis (Figure 1). C-reactive protein was elevated (22.64 mg/dL, normal <3 mg/dL), and a chest radiograph was consistent with interstitial pneumonia. The remainder of her laboratory workup was normal. Punch biopsies demonstrated subtotal epidermal necrosis with sparse lymphocytic infiltrate on hematoxylin and eosin stain and negative direct immunofluorescence microscopy.

It was initially suspected that her case was infection-mediated. She was started empirically on azithromycin for possible *Mycoplasma pneumoniae*–induced rash with mucositis. However, a comprehensive viral respiratory polymerase chain reaction (PCR)* panel was negative, *Chlamydia pneumoniae* and *M. pneumoniae* blood PCR and IgM antibodies were negative on two separate occasions. A second oropharyngeal group A streptococcal molecular antigen test was negative. Two blood cultures and a urine culture demonstrated no bacterial growth. Rowell syndrome (erythema multiforme-like lesions associated with lupus erythematosus) was ruled out by negative screening tests for a autoimmune disease.

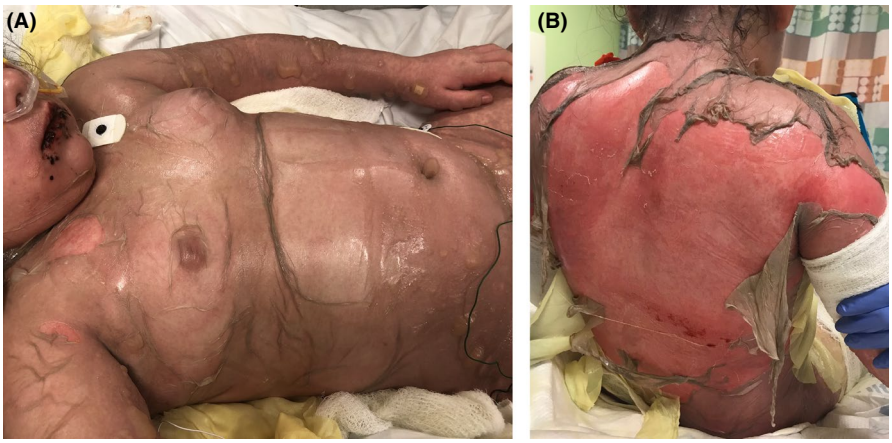

FIGURE 1 A 10-y-old girl with toxic epidermal necrolysis. A, Hemorrhagic crusting of upper and lower vermillion lips with multiple large, tense clear/yellow fluid-filled bullae on arms and abdomen and Nikolsky-positive desquamation on abdomen and chest. B, Diffuse desquamation revealing bright pink denuded dermis on the back

With no clear trigger, she was diagnosed with idiopathic TEN. On day 10 of hospitalization, she was transferred to another hospital for escalation of care with 90% body surface area involvement and about 50% total desquamation. Happily, she fully recovered.

3 | DISCUSSION

The incidence of idiopathic SJS and SJS/TEN overlap in the pediatric population is estimated to be 5%-16.6%.[4,5] Searching for both common and rare causes of SJS/TEN is critically important before identifying a case as idiopathic (Table 1). Often overlooked entities include food ingredients, insecticides, acetaminophen, and herbal remedies.[5] Additionally, infectious and autoimmune diseases must be ruled out.

In our case, despite the thorough history and workup, an etiology was never found. This suggests that the pathophysiology of SJS/TEN is highly complex and still poorly understood. A patient's predisposition to SJS/TEN is likely multifactorial and an interplay between genetics, the immune system, and environment. An underlying disease process or medication exposure needed to trigger the eruption may not be universal. Idiopathic SJS/TEN is therefore a particular challenge, as the clinical picture alone must guide the clinician toward the most appropriate management, one which optimizes the child's outcome and reduces mortality.

TABLE 1 Uncommon reported causes of pediatric SJS/TEN

Vaccines	Hantavirus vaccine
	Measles vaccine
	Measles, mumps, rubella (MMR) vaccine
	Meningococcal B vaccine
	Varicella vaccine
	Yellow fever vaccine
Insecticides	Lambda-cyhalothrin
	Thiamethoxam
Over-the-counter medications	Acetaminophen (eg, paracetamol)
	Dimenhydrinate
	Multi-ingredient cold medication
Prescription medications	Benzodiazepines
	Bronchodilator (eg, albuterol)
	Chemotherapy agents (eg, methotrexate, dactinomycin, vincristine)
	D-penicillamine
	Oral contraceptives
	Warfarin
Herbal/Vitamin supplements	Ascorbic acid
	Ayurvedic drugs
	Homeopathic medicines (eg, Chinese herbs)
Procedures	Allogenic hematopoietic stem cell transplantation
Infectious and autoimmune diseases	*Chlamydia pneumoniae*
	Graft versus host
	Linear IgA bullous dermatosis
	Systemic lupus erythematosus
	Toxic shock syndrome

ENDNOTE

* Comprehensive viral PCR panel: influenza A, influenza B, adenovirus, coronavirus, parainfluenza virus, respiratory syncytial virus A and B, chlamydia pneumonia, mycoplasma pneumonia, human metapneumovirus, human rhinovirus, enterovirus

Keywords
idiopathic, pediatrics, toxic epidermal necrolysis

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