Pediatric Dermatology Emergencies

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PRACTICE **POINTS**

- Staphylococcal scalded skin syndrome, impetigo, eczema herpeticum, Langerhans cell histiocytosis, infantile hemangiomas, and IgA vasculitis all present potential emergencies in pediatric patients in dermatologic settings.
- Early and accurate identification and management of these entities is critical to avoid short-term and long-term negative sequalae.

Many pediatric skin conditions can be safely monitored with minimal intervention, but certain skin conditions are emergent and require immediate attention and proper assessment of the neonate, infant, or child. We review the following pediatric dermatology emergencies so that clinicians can detect and accurately diagnose these conditions to avoid delayed treatment and considerable morbidity and mortality if missed: staphylococcal scalded skin syndrome (SSSS), impetigo, eczema herpeticum (EH), Langerhans cell histiocytosis (LCH), infantile hemangioma (IH), and IgA vasculitis.

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Many pediatric skin conditions can be safely monitored with minimal intervention, but certain skin conditions are emergent and require immediate attention and proper assessment of the neonate, infant, or child. The skin may provide the first presentation of a potentially fatal disease with serious sequelae. Cutaneous findings may indicate the need for further evaluation. Therefore, it is important to differentiate skin conditions with benign etiologies from those that require immediate diagnosis and treatment, as early intervention of some of these conditions can be lifesaving. Herein, we discuss pertinent pediatric dermatology emergencies that dermatologists should keep in mind so that these diagnoses are never missed.

Staphylococcal Scalded Skin Syndrome

Presentation—Staphylococcal scalded skin syndrome (SSSS), or Ritter disease, is a potentially fatal pediatric emergency, especially in newborns.[1] The mortality rate for SSSS in the United States is 3.6% to 11% in children.[2] It typically presents with a prodrome of tenderness, fever, and confluent erythematous patches on the folds of the skin such as the groin, axillae, nose, and ears, with eventual spread to the legs and trunk.[1,2] Within 24 to 48 hours of symptom onset, blistering and fluid accumulation will appear diffusely. Bullae are flaccid, and tangential and gentle pressure on involved unblistered skin may lead to shearing of the epithelium, which is a positive Nikolsky sign.[1,2]

Causes—Staphylococcal scalded skin syndrome is caused by exfoliative toxins A and B, toxigenic strains of *Staphylococcus aureus*. Exfoliative toxins A and B are serine proteases that target and cleave desmoglein 1, which binds keratinocytes in the stratum granulosum.[1,3] Exfoliative toxins disrupt the adhesion of keratinocytes, resulting in bullae formation and subsequently diffuse sheetlike desquamation.[1,4,5] Although up to 30% of the human population are asymptomatically and permanently colonized with nasal *S aureus*,[6] the exfoliative toxins are produced by only 5% of species.[1]

In neonates, the immune and renal systems are underdeveloped; therefore, patients are susceptible to

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SSSS due to lack of neutralizing antibodies and decreased renal toxin excretion.[4] Potential complications of SSSS are deeper soft-tissue infection, septicemia (blood-borne infection), and fluid and electrolyte imbalance.[1,4]

Diagnosis and Treatment—The condition is diagnosed clinically based on the findings of tender erythroderma, bullae, and desquamation with a scalded appearance, especially in friction zones; periorificial crusting; positive Nikolsky sign; and lack of mucosal involvement (Figure 1).[1] Histopathology can aid in complicated clinical scenarios as well as culture from affected areas, including the upper respiratory tract, diaper region, and umbilicus.[1,4] Hospitalization is required for SSSS for intravenous antibiotics, fluids, and electrolyte repletion.

Differential Diagnosis—There are multiple diagnoses to consider in the setting of flaccid bullae in the pediatric population. Stevens-Johnson syndrome or toxic epidermal necrolysis also can present with fever and superficial desquamation or bullae; however, exposure to medications and mucosal involvement often are absent in SSSS (Figure 2).[2] Pemphigus, particularly paraneoplastic pemphigus, also often includes mucosal involvement and scalding thermal burns that are often geometric or focal. Epidermolysis bullosa and toxic shock syndrome also should be considered.[1]

Impetigo

Presentation—Impetigo is the most common bacterial skin infection in children caused by *S aureus* or *Streptococcus pyogenes*.[7-9] It begins as erythematous papules transitioning to thin-walled vesicles that rapidly rupture and result in honey-crusted papules.[7,9,10] Individuals of any age can be affected by nonbullous impetigo, but it is the most common skin infection in children aged 2 to 5 years.[7]

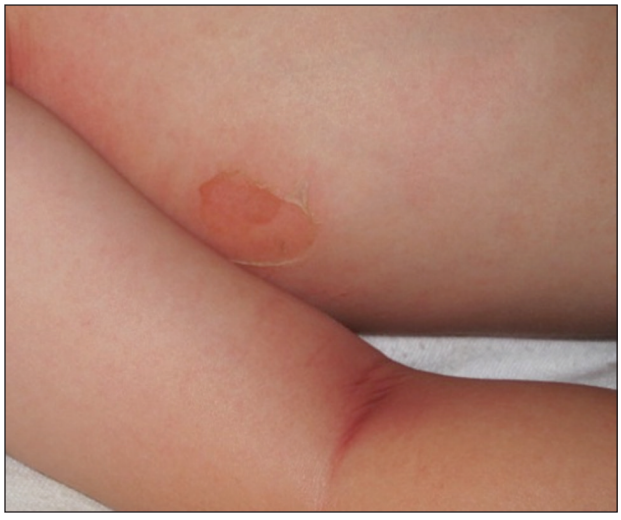

FIGURE 1. Staphylococcal scalded skin syndrome. Erythema of the axilla and antecubital fossa and an erosion on the right flank. The skin was tender to the touch.

Bullous impetigo primarily is seen in children, especially infants, and rarely can occur in teenagers or adults.[7] It most commonly is caused by the exfoliative toxins of *S aureus*. Bullous impetigo presents as small vesicles that may converge into larger flaccid bullae or pustules.[7-10] Once the bullae rupture, an erythematous base with a collarette of scale remains without the formation of a honey-colored crust.[8] Bullous impetigo usually affects moist intertriginous areas such as the axillae, neck, and diaper area[8,10] (Figure 3). Complications may result in cellulitis, septicemia, osteomyelitis, poststreptococcal glomerulonephritis associated with *S pyogenes*, and *S aureus*–induced SSSS.[7-9]

Diagnosis—Nonbullous and bullous impetigo are largely clinical diagnoses that can be confirmed by culture of a vesicle or pustular fluid.[10] Treatment of impetigo includes topical or systemic antibiotics.[7,10] Patients should be advised to keep lesions covered and avoid contact with others until all lesions resolve, as lesions are contagious.[9]

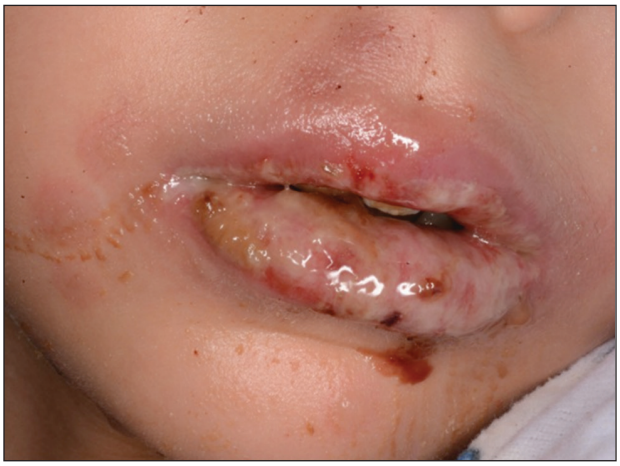

FIGURE 2. Stevens-Johnson syndrome secondary to trimethoprim-sulfamethoxazole exposure. Ulceration of the upper and lower lips highlight mucosal involvement.

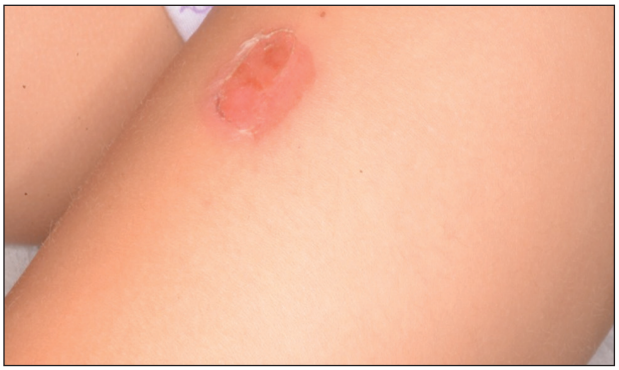

FIGURE 3. Bullous impetigo. A burst bulla on the anterior aspect of the left thigh.

Eczema Herpeticum

Presentation—Eczema herpeticum (EH), also known as Kaposi varicelliform eruption, is a disseminated herpes simplex virus infection of impaired skin, most commonly in patients with atopic dermatitis (AD).[11] Eczema herpeticum presents as a widespread eruption of erythematous monomorphic vesicles that progress to punched-out erosions with hemorrhagic crusting (Figure 4). Patients may have associated fever or lymphadenopathy.[12,13]

Causes—The number of children hospitalized annually for EH in the United States is approximately 4 to 7 cases per million children. Less than 3% of pediatric AD patients are affected, with a particularly increased risk in patients with severe and earlier-onset AD.[12-15] Patients with AD have skin barrier defects, and decreased IFN-γ expression and cathelicidins predispose patients with AD to developing EH.[12,16,17]

Diagnosis—Viral polymerase chain reaction for herpes simplex virus types 1 and 2 is the standard for confirmatory diagnosis. Herpes simplex virus cultures from cutaneous scrapings, direct fluorescent antibody testing, or Tzanck test revealing multinucleated giant cells also may help establish the diagnosis.[11,12,17]

Management—Individuals with severe AD and other dermatologic conditions with cutaneous barrier compromise are at risk for developing EH, which is a medical emergency requiring hospitalization and prompt treatment with antiviral therapy such as acyclovir, often intravenously, as death can result if left untreated.[11,17] Topical or systemic antibiotic therapy should be initiated if there is suspicion for secondary bacterial superinfection. Patients should be evaluated for multiorgan involvement such as keratoconjunctivitis, meningitis, encephalitis, and systemic viremia due to increased mortality, especially in infants.[12,15,16]

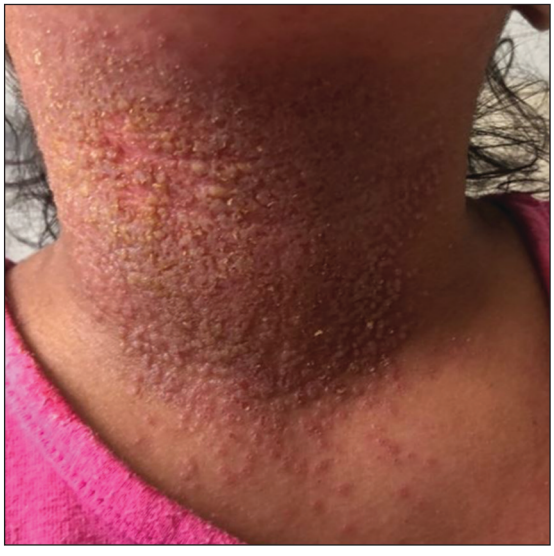

FIGURE 4. Eczema herpeticum. Diffuse and confluent punched-out and crusted erosions on the neck.

Langerhans Cell Histiocytosis

Presentation—Langerhans cell histiocytosis (LCH) has a variable clinical presentation and can involve a single or multiple organ systems, including the bones and skin. Cutaneous LCH can present as violaceous papules, nodules, or ulcerations and crusted erosions (Figure 5). The lymph nodes, liver, spleen, oral mucosa, and respiratory and central nervous systems also may be involved.

Langerhans cell histiocytosis affects individuals of any age group but more often is seen in pediatric patients. The incidence of LCH is approximately 4.6 cases per million children.[18] The pathogenesis is secondary to pathologic Langerhans cells, characterized as a clonal myeloid malignancy and dysregulation of the immune system.[18,19]

Diagnosis—A thorough physical examination is essential in patients with suspected LCH. Additionally, diagnosis of LCH is heavily based on histopathology of tissue from the involved organ system(s) with features of positive S-100 protein, CD1a, and CD207, and identification of Birbeck granules.[20] Imaging and laboratory studies also are indicated and can include a skeletal survey (to assess osteolytic and organ involvement), a complete hematologic panel, coagulation studies, and liver function tests.[18,21]

Management—Management of LCH varies based on the organ system(s) involved along with the extent of the disease. Dermatology referral may be indicated in patients presenting with nonresolving cutaneous lesions as well as in severe cases. Single-organ and multisystem disease may require one treatment modality or a combination of chemotherapy, surgery, radiation, and/or immunotherapy.[21]

Infantile Hemangioma

Presentation—Infantile hemangioma (IH) is the most common benign tumor of infancy and usually is apparent a few weeks after birth. Lesions appear as bright red papules, nodules, or plaques. Deep or subcutaneous lesions present as raised, flesh-colored nodules with a blue hue and bruiselike appearance with or without a central patch

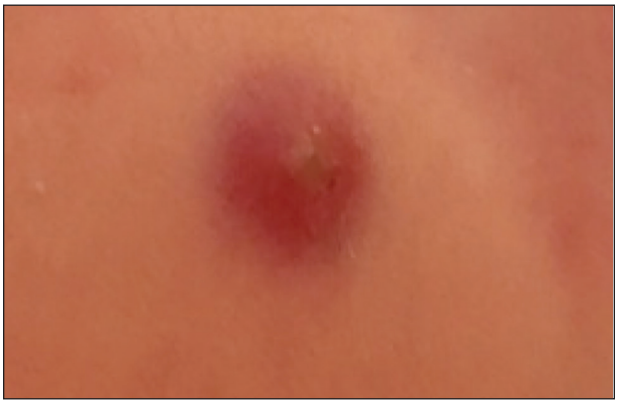

FIGURE 5. Langerhans cell histiocytosis. Congenital red to slightly violaceous nodule with an overlying pustule on the right cheek.

of telangiectasia[22-24] (Figure 6). Although all IHs eventually resolve, residual skin changes such as scarring, atrophy, and fibrosis can persist.[24]

The incidence of IH has been reported to occur in up to 4% to 5% of infants in the United States.[23,25] Infantile hemangiomas also have been found to be more common among white, preterm, and multiple-gestation infants.[25] The proposed pathogenesis of IHs includes angiogenic and vasogenic factors that cause rapid proliferation of blood vessels, likely driven by tissue hypoxia.[23,26,27]

Diagnosis—Infantile hemangioma is diagnosed clinically; however, immunohistochemical staining showing positivity for glucose transporter 1 also is helpful.[26,27] Imaging modalities such as ultrasonography and magnetic resonance imaging also can be utilized to visualize the extent of lesions if necessary.[25]

Management—Around 15% to 25% of IHs are considered complicated and require intervention.[25,27] Infantile hemangiomas can interfere with function depending on location or have potentially fatal complications. Based on the location and extent of involvement, these findings can include ulceration; hemorrhage; impairment of feeding, hearing, and/or vision; facial deformities; airway obstruction; hypothyroidism; and congestive heart failure.[25,28] Early treatment with topical or oral beta-blockers is imperative for potentially life-threatening IHs, which can be seen due to large size or dangerous location.[28,29] Because the rapid proliferative phase of IHs is thought to begin around 6 weeks of life, treatment should be initiated as early as possible. Initiation of beta-blocker therapy in the first few months of life can prevent functional impairment, ulceration, and permanent cosmetic changes. Additionally, surgery or pulsed dye laser treatment have been found to be effective for skin changes found after involution of IH.[25,29]

Differential Diagnosis—The differential diagnosis for IH includes vascular malformations, which are present at birth and do not undergo rapid proliferation; sarcoma; and kaposiform hemangioendothelioma, which causes the Kasabach-Merritt phenomenon secondary to platelet trapping. Careful attention to the history of the skin lesion provides good support for diagnosis of IH in most cases.

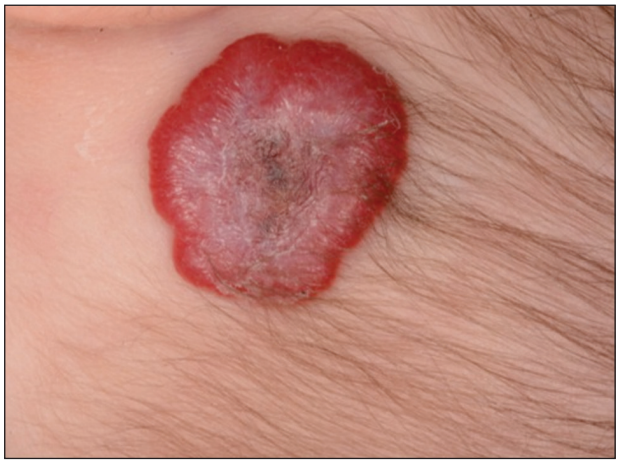

FIGURE 6. Ulcerated superficial infantile hemangioma in an 8-week-old neonate. Crusting and erosion were noted at the center of the red plaque with white discoloration surrounding the crust, an indicator of prior ulceration.

IgA Vasculitis

Presentation—IgA vasculitis, or Henoch-Schönlein purpura, classically presents as a tetrad of palpable purpura, acute-onset arthritis or arthralgia, abdominal pain, and renal disease with proteinuria or hematuria.[30] Skin involvement is seen in almost all cases and is essential for diagnosis of IgA vasculitis. The initial dermatosis may be pruritic and present as an erythematous macular or urticarial wheal that evolves into petechiae, along with palpable purpura that is most frequently located on the legs or buttocks (Figure 7).[30-34]

IgA vasculitis is an immune-mediated small vessel vasculitis with deposition of IgA in the small vessels. The underlying cause remains unknown, though infection, dietary allergens, drugs, vaccinations, and chemical triggers have been recognized in literature.[32,35,36] IgA vasculitis is largely a pediatric diagnosis, with 90% of affected individuals younger than 10 years worldwide.[37] In the pediatric population, the incidence has been reported to be 3 to 26.7 cases per 100,000 children.[32]

Diagnosis—Diagnosis is based on the clinical presentation and histopathology.[30] On direct immunofluorescence, IgA deposition is seen in the vessel walls.[35] Laboratory testing is not diagnostic, but urinalysis is mandatory to identify involvement of renal vasculature. Imaging studies

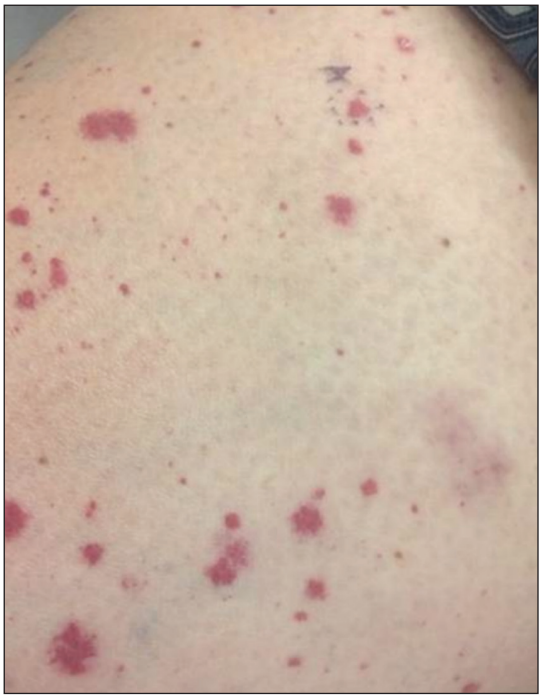

FIGURE 7. IgA vasculitis. Palpable petechiae and purpura on the leg.

may be used in patients with abdominal symptoms, as an ultrasound can be used to visualize bowel structure and abnormalities such as intussusception.[33]

Management—The majority of cases of IgA vasculitis recover spontaneously, with patients requiring hospital admission based on severity of symptoms.[30] The primary approach to management involves providing supportive care including hydration, adequate rest, and symptomatic pain relief of the joints and abdomen with oral analgesics. Systemic corticosteroids or steroid-sparing agents such as dapsone or colchicine can be used to treat cutaneous manifestations in addition to severe pain symptoms.[30,31] Patients with IgA vasculitis must be monitored for proteinuria or hematuria to assess the extent of renal involvement. Although much more common in adults, long-term renal impairment can result from childhood cases of IgA vasculitis.[34]

Final Thoughts

Pediatric dermatology emergencies can be difficult to detect and accurately diagnose. Many of these diseases are potential emergencies that that may result in delayed treatment and considerable morbidity and mortality if missed. Clinicians should be aware that timely recognition and diagnosis, along with possible referral to pediatric dermatology, are essential to avoid complications.

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