Management of paediatric dermatological emergencies

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Abstract

Most skin diseases can be safely managed in outpatients; however, in a significant proportion of cases, immediate and aggressive intervention is required. Recognition of these more serious and potentially life-threatening dermatological presentations is imperative in order to provide correct emergency treatment and limit morbidity and mortality. This article looks at conditions which may present in childhood that might require urgent treatment. These include infection, drug reactions, erythroderma, congenital ichthyoses (especially collodian baby and Harlequin ichthyosis), Stevens–Johnson syndrome, toxic epidermal necrolysis, infantile haemangiomas and epidermolysis bullosa.

Keywords adverse drug reaction; dermatology; emergency; erythroderma; infection

Paediatric skin disorders account for a significant number of cases presenting to Emergency departments or Paediatric assessment units with around half being due to infectious causes. Skin problems may also present in the newborn period and these may necessitate prompt treatment. Whilst many children will have minor conditions, or at least diseases that can be safely treated as an outpatient a minority will present with skin manifestations of diseases that require urgent treatment.

Erythroderma

This is a relatively rare presentation in children, accounting for less than 1% of paediatric dermatology clinic presentations, but is potentially life-threatening. Erythroderma or generalised exfoliative dermatitis is defined as an inflammatory disorder with erythema affecting more than 90% of the body surface area. It is often associated with generalised lymphadenopathy blistering or scaling, Complications include hypothermia, hypernatraemic dehydration, hypoalbuminaemia, septicaemia and high-output cardiac failure.

In infants, hereditary ichthyoses may also present with erythroderma. These are a large group of heterogeneous disorders that are characterised by rough dry scaly skin. They include non-bullous ichthyosiform erythroderma, bullous ichthyosiform erythroderma, Netherton's syndrome and Conradi–Hünermann syndrome. Children with these conditions have a genetically deficient skin barrier function, which predisposes them to inflammatory skin responses.

Immunological conditions presenting with erythroderma include Omenn syndrome due to severe combined immunodeficiency (SCID). This disorder is characterised by failure to thrive, erythroderma, recurrent infections and lymphadenopathy. Similar presentations occur in graft versus host disease and hypogammaglobulinaemia.

Regardless of its cause, the treatment of erythroderma should be prompt and vigorous. Maintaining adequate oral or parenteral fluid intake and monitoring serum electrolytes is central to supportive treatment. Attention should also be paid to wound care, barrier nursing and thermoregulation. Topical preparations of greasy emollients should be used liberally to hydrate the skin and prevent fissuring. The blisters and erosions can be managed with 0.01% potassium permanganate soaks and systemic antibiotics if necessary.

Collodian baby

This condition is slightly more common in preterm infants. A taut, yellowish film is stretched over the skin. Eyelids and lips may be tethered and everted (ectropion and eclabion), nasal passages obstructed and the pinnae flattened. Constriction may lead to digital ischaemia and fingers can appear sausage-shaped. Typically the membrane peels away and reforms, drying out with fissures and shedding over 1–4 weeks. Common underlying conditions are erythrodermic or lamellar ichthyosis. Restrictive dermopathy is an important differential diagnosis. This presents as stiff baby syndrome with generalised taut thick tethered and unyielding skin at birth which does not shed.

Harlequin ichthyosis

Harlequin ichthyosis is the most severe form of congenital ichthyosis, characterized by a thickening of the keratin layer in fetal human skin. Thick, diamond-shaped plate-like scales are present at birth; these restrict the movement of the infant and can lead to contractures. In most cases mutations in the *ABCA12* gene are found. The *ABCA12* gene provides instructions for making a protein that plays a role in the transport of lipids in the epidermis. Without sufficient quantities of this protein the skin is thick and inflexible. Movement is restricted and splits develop in the thick scale giving deep red fissures. Respiratory insufficiency may be present due to restricted chest movement and also because of prematurity.

Harlequin ichthyosis can usually be distinguished from collodian baby but cases with intermediate features have been seen. For both conditions complications include hypothermia, dehydration, hypernatraemia and sepsis due to the erosions (Table 1).

Acute erythrodermic psoriasis (Figure 1)

This is rare and presents as widespread pustules on red and tender skin. There may be a history of chronic plaque psoriasis or triggers include infections and drugs including sudden withdrawal of corticosteroids. Systemic symptoms may be present including fever, headache, muscle weakness, diarrhoea and vomiting. After a few days the pustules coalesce to form lakes of sterile pus which dry and peel followed by successive crops.

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Treatment of collodian babies and Harlequin ichthyosis

- Humidified incubators initially transferring to a cot as soon as possible dressing the skin with cotton tubular bandages and greasy emollient.
- Monitor blood counts.
- Measure urea electrolytes and creatinine.
- Apply emollient to cleanse skin and ointment such as white soft paraffin or emulsifying ointment with feeds and nappy changes.
- Ensure emollient is kept sterile.
- Beware of opportunistic infection including MRSA, Candida and Pseudomonas and swab skin every 2–3 days.
- NG feeding may be needed for poor sucking.
- Involve an ophthalmologist in care.
- Avoid prophylactic antibiotics and placing of IV or umbilical lines unless essential.
- Oral retinoids for Harlequin ichthyosis commenced as soon as possible.

Table 1

Skin swabs should be taken to exclude infection and liberal emollients applied. A skin biopsy is needed to confirm diagnosis. Strict fluid balance and monitoring of urea, electrolytes, creatinine and calcium is needed. A dermatologist should be involved in care as usually systemic immunosuppressant therapy e.g. methotrexate, ciclosporin or acitretin is required. An important differential diagnosis is acute generalized exanthematous pustulosis (AGEP). AGEP is an acute febrile drug reaction which results in numerous small sterile pustules on erythematous skin. Unlike Stevens–Johnson syndrome there is no mucosal involvement. Treatment of AGEP is supportive care and withdrawal of the causative drug.

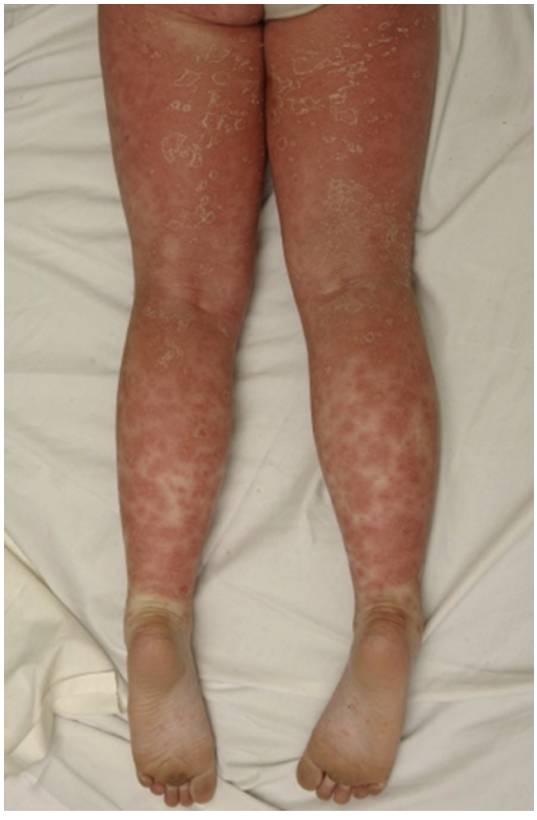

Figure 1 Acute erythrodermic psoriasis.

Staphylococcal scalded skin syndrome (SSSS) (Figure 2)

SSSS is an uncommon disease of early childhood. It typically affects children less than 5 years old. It is important to differentiate SSSS (synonym Ritter's disease, pemphigus neonatorum) from Toxic Epidermal necrolysis (TEN) and this is usually a clinical diagnosis. The characteristic feature of skin shedding upon light pressure (Nikolsky's sign) is usually present but skin biopsy can be helpful in cases where there is diagnostic doubt. In SSSS, there is superficial separation within the epidermis, just beneath the stratum corneum whereas in TEN there is dermo-epidermal separation and full thickness epidermal necrosis. The superficial nature of the skin shedding in SSSS ensures that healing occurs without scarring. However, infants are vulnerable to fluid loss and infection and without prompt treatment it may be fatal.

Toxic shock syndrome (TSS)

TSS similar to staphylococcal scalded skin syndrome is caused by exotoxins produced by *Staphylococcus aureus* or *Streptococcus pyogenes* (Group A Streptococcus). It is an acute, multi-system illness that leads to tissue damage, disseminated intravascular coagulation and organ dysfunction. Even with optimal care it still has a high mortality (5–15%) and is a diagnostic and therapeutic challenge.

TSS can present at all ages with fever, hypotension, and shock. Shortly after birth it is most commonly due to transmission of toxigenic *S. aureus* from either an intrauterine infection or one acquired at the time of delivery. The extensive skin rash initially resembles scarlet fever and can develop into erythroderma. Rapid multi-organ failure can occur within 8–12 hours after onset of symptoms. There may be hyperaemia of the mucous membranes and desquamation of the skin on the hands and feet 10–21 days after disease onset.

Biochemical disturbances often include elevated creatinine, serum transaminases and bilirubin. *S. aureus* is cultured in less

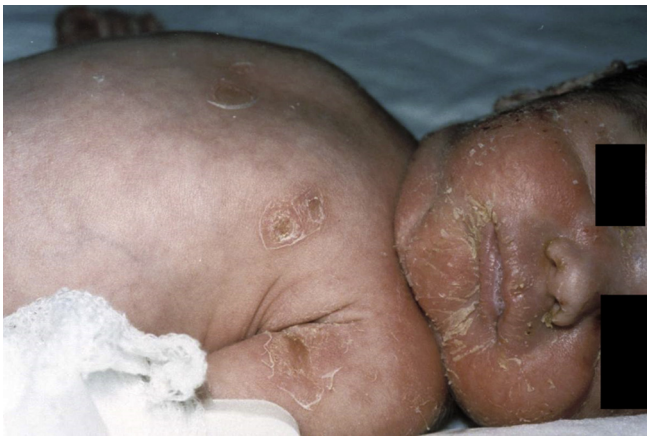

Figure 2 Staphylococcal scalded skin syndrome.