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Original Research Article

Pediatric dermatological emergencies in tertiary health care

Vrutika H. Shah*, Vidya D. Kharkar

Department of Dermatology-Venereology-Leprosy. Seth GS Medical College and K. E. M. Hospital, Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Vrutika H Shah,
E-mail: vrutikashah25@gmail.com

ABSTRACT

Background: In recent years, the demand for dermatological consultations in the emergency department has increased. However, most of the publications of dermatological emergencies deal with the general population, without taking into account the changes in frequencies found in young children (ages 0-14 years). Dermatological emergencies in pediatric age group are not uncommon. The aim of our study was to analyse the spectrum of dermatological emergencies in the pediatric age group.
Methods: A hospital-based observational study was conducted at the Department of Dermatology of tertiary care centre, between September 2019 and February 2020. Thirty pediatric patients (age ≤16 years) who had visited Dermatology Outpatient Department (OPD) and emergency department were included in this study.
Results: Total thirty patients were included in this study. The male to female ratio was 2:1. The average age of the study was 7.6 years. Maximum cases were in school-aged children (6 to 12 years), which were 12 (40%). The average duration of illness was 36.04 days. The most common condition was inherited conditions (23%) followed by primary cutaneous infections (20%). The cases were divided as 7 (23%) inherited conditions, 6 (20%) cases of infections, 5 (17%) cases each of vasculitis and cutaneous adverse drug reaction and 7 (23%) miscellaneous cases.
Conclusions: Pediatric dermatological emergencies require an interdisciplinary approach for timely intervention. Standardised protocol should be followed in their management to prevent complications like acute skin failure. Early referral of these patients with primary skin conditions to the dermatologists might result in better clinical outcome and decreased mortality.

Keywords: Cutaneous emergencies, Pediatric, Skin manifestations

INTRODUCTION

Dermatological emergencies are not uncommon in pediatric age group. These conditions could be a primary skin disorder, a coexisting skin disorder or a primary systemic disorder with cutaneous manifestations.[1] Skin disorders have been estimated to represent 4-6% of all pediatric emergency care unit visits, with only 30% of these being true emergencies.[2] There are very few reported studies on pediatric dermatological emergencies (PDE) in inpatient care as well as outpatient care or overall. As there is paucity of studies in this regard, we conducted this study to delineate the pattern of pediatric dermatological emergencies (PDE) in tertiary health care.

METHODS

It was a hospital-based observational study conducted at the Department of Dermatology of tertiary care centre, between September 2019 and February 2020. Pediatric patients (age ≤16 years) who had visited Dermatology

Outpatient Department (OPD) and emergency department were included in this study. A written informed consent was taken from their parents to use their data in this study. All patients above 16 years of age and those who did not consent were excluded from the study. Detailed demographic data, history and clinical examination were recorded. The diagnosis was made based on clinical features supported by necessary investigations wherever required. Skin biopsy was performed wherever necessary. Final diagnosis was made on the basis of history, clinical examination, and additional investigations. Statistical analysis was done using Statistical package for social sciences (SPSS) version 20 (SPSS, Inc. Chicago, IL, U.S.A.). Normally distributed continuous variables were expressed as the mean. Categorical data were reported as numbers and percentages. Ethical committee approval was not taken as it was an observational study.

RESULTS

Thirty patients which presented as pediatric dermatological emergency in dermatology department were enrolled in this study. The male to female ratio was 2:1. The age of patients ranged from 1 month to 16 years. The average age of the study was 7.6 years.

Table 1: Age-wise and sex-wise distribution of cases.

Age group (years)	Male	Female	Total
0-1	0	3	3
1-2	4	0	4
3-5	3	1	4
6-12	8	4	12
12-16	5	2	7
Total	20	10	30

Table 1 demonstrates age-wise and sex-wise distribution of cases. Maximum cases were in school-aged children (6 to 12 years), which were 12 (40%). The duration of illness ranged from 1 day to 450 days. Figure 1 depicts duration of illness in bar diagram. The average duration of illness was 36.04 days. The dermatologic diagnoses were grouped in following categories for analysis: hereditary conditions, primary cutaneous infections, drug reactions, vasculitis, and others. The most common condition was inherited conditions (23%) followed by primary cutaneous infections (20%). The thirty diagnoses were divided as seven (23%) inherited conditions, six (20%) cases of infections, five (17%) cases each of vasculitis and cutaneous adverse drug reaction (CADR) and seven (23%) miscellaneous cases.

Seven inherited conditions included 2 of lamellar ichthyosis (LI) (Figure 2), 1 of X-linked recessive ichthyosis (XLRI) (Figure 2), 1 of epidermolytic hyperkeratosis (EHK) (Figure 2), 1 of Epidermolysis Bullosa simplex (EBS) (Figure 3) and 2 of epidermolysis bullosa dystrophica (EBD) (Figure 3).

Seven miscellaneous conditions encompassed 1 case each of bullous mastocytosis (BM) (Figure 5), urticaria pigmentosa (Figure 5), infantile hemangioma [(Figure 5), eruptive lichen planus, angioedema, Langerhans cell histiocytosis (LCH) (Figure 7) and Mucha-Habermann disease (MHD) (Figure 8). There was one mortality in our study which was of LCH patient. LCH was multisystemic and patient had developed failure to thrive, pancytopenia, hepatosplenomegly with bone marrow infiltration. Before the treatment could be started, the child succumbed to death. Five cases of vasculitis included 4 cases of leukocytoclastic vasculitis (LCV) and 1 case of Henoch–Schönlein purpura (HSP).

Table 2: Comparison of various studies.

Parameters	Present Study	Sarkar et al[5]	Mathias et al[6]
Sample size	30	103	90
Duration of study	6 months	1 year	18 months
Male: Female ratio	2:1	1.51:1	1.4:1
Most common age group	School-age group	School-age group	Preschool age group
Most common diagnosis	1. Hereditary conditions (23%) 2. Primary cutaneous infections (20%)	1. Hereditary conditions in neonate group (37.5%) 2. Infections in preschool age group (42.1%) 3. Adverse cutaneous drug reactions in school age group (56.4%)	1. Primary cutaneous infections (40%) 2. Adverse cutaneous drug reactions (13.33%)

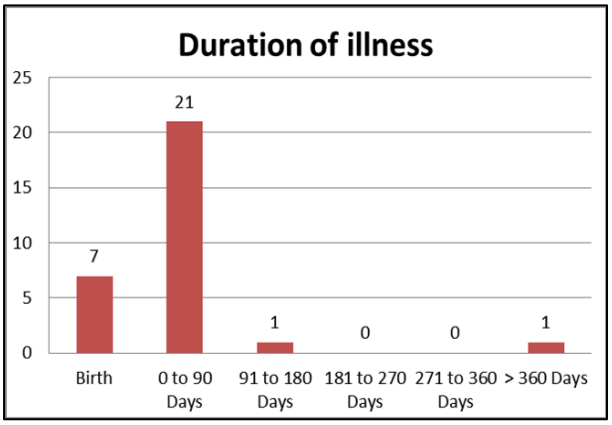

Figure 1: Distribution of cases as per duration of illness.

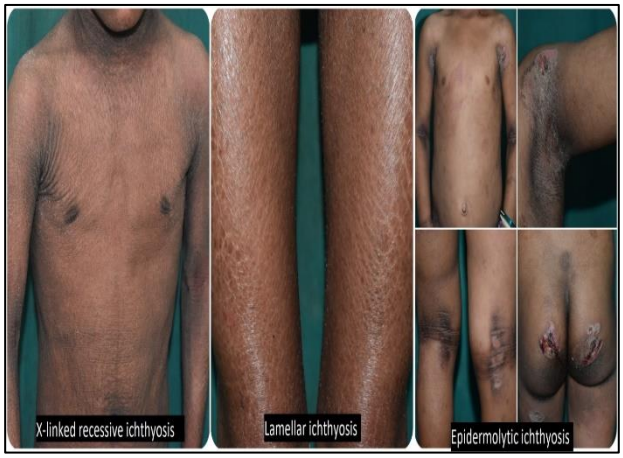

Figure 2: Spectrum of ichthyosis: X-linked recessive, lamellar and epidermolytic ichthyosis.

Figure 3: Epidermolysis bullosa simplex and dystrophica.

Five cases of CADR comprised: 3 cases of maculopapular drug rash, 1 case of erythema multiforme (EM) and 1 cases of Stevens-Johnson syndrome (SJS) (Figure 6). Out of the 3 cases of drug rash, 1 each was suspected due to beta-lactams and phenytoin and 1 was secondary to anti-Koch's treatment (AKT). 1 case of EM was due to cefixime and 1 case of SJS was due to amoxicillin-clavulanate.

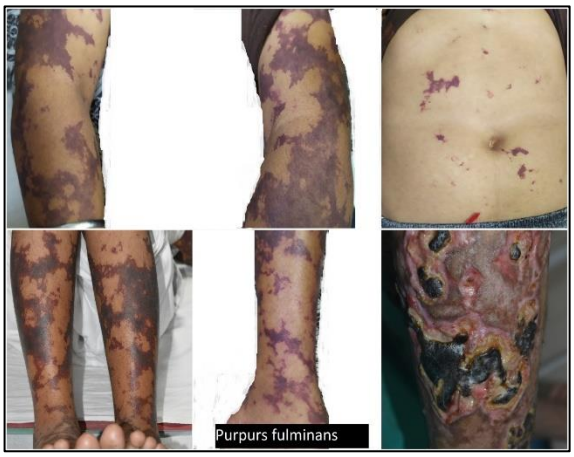

Figure 4: Sharply demarcated retiform purpura and necrotic eschar in a child with purpura fulminans.

Six cases of infections included 2 cases of varicella, 1 case each of herpes zoster, purpura fulminans (Figure 4), viral exanthem and necrotizing fasciitis (Figure 5). Purpura fulminans (PF) was acquired secondary to rickettsial infection with Weil Felix test positive for Ox-19 antigen. Necrotizing fasciitis (NF) was secondary to group A streptococcus.

DISCUSSION

The number of patients requiring immediate care in the Emergency Department due to dermatological conditions represents a significant percentage of the total, with an estimated 8-10%.[3,4] The spectrum of dermatological emergencies in all age groups has been studied in past. Also, the gamut of pediatric dermatological disorders presenting to the OPD has been studied in the past. But there are very few studies of PDE. Hence, our study focussed only on true PDE.

The male to female ratio in our study was 2:1 as compared to 1.51:1 in Sarkar et al and 1.4:1 in Mathias et al. The maximum number of children was in school-age group; constituting 40% of the total which was similar to Sarkar et al. (37.9%) while in Mathias et al. the maximum number of children were of preschool age group, constituting nearly 30% of the total.[5,6] Hereditary conditions (23%) were the most common condition in this study which was in comparison with Sarkar et al. (37.5%) whereas in Mathias et al., the most common condition was primary cutaneous infections (40%).[5,6] Comparison between various studies is shown in Table 2. True PDE includes an exhaustive list of various inherited and acquired conditions. This list can be divided into inherited conditions, infections, vascular lesions, drug reactions and miscellaneous diseases. Inherited PDE includes colloidion baby, ichthyoses and epidermolysis bullosa (EB) with its various variants.

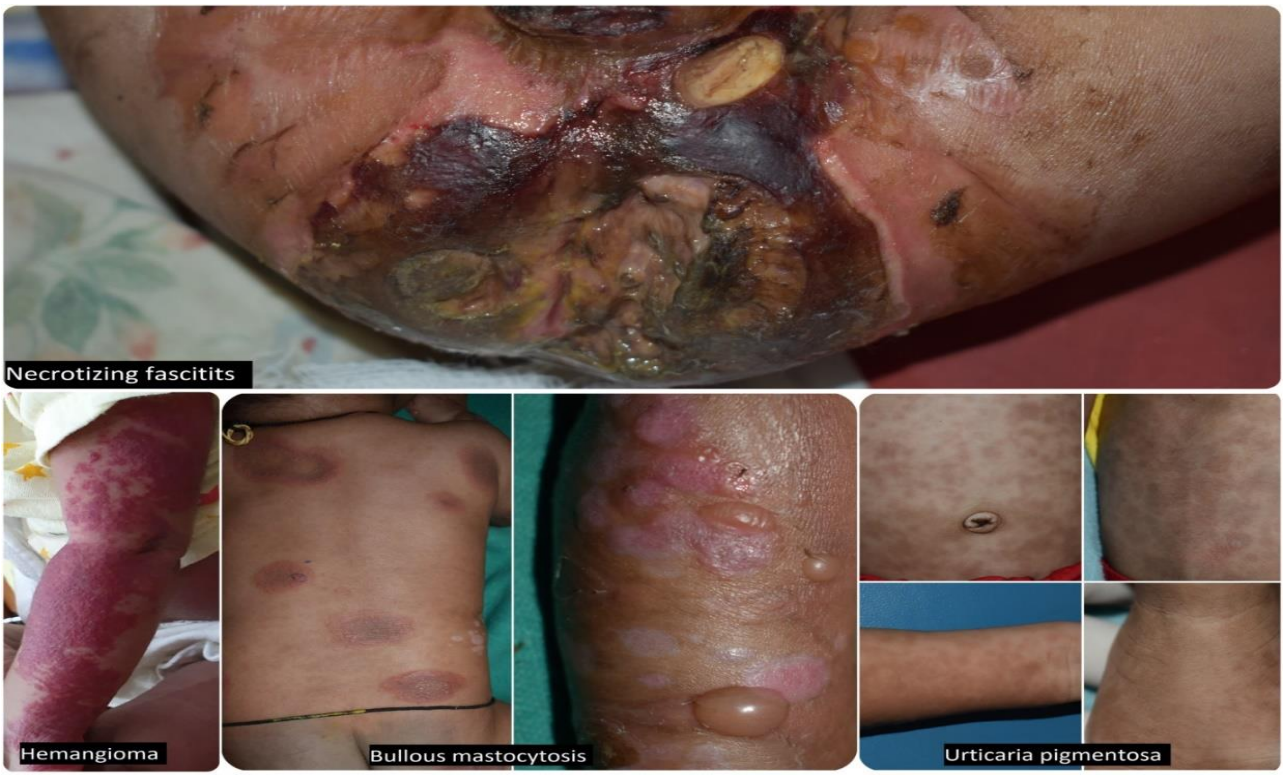

Figure 5: Necrotizing fasciitis: Early dermal necrosis and black eschar formation affecting scrotum and perianal area, extending over medial aspect of bilateral thighs; hemangioma; bullous mastocytosis and urticaria pigmentosa (doughy skin consistency).

Figure 6: Cutaneous adverse drug reaction spectrum: maculopapular drug rash, Stevens Johnson syndrome and erythema multiforme with typical target lesions.

The collodion baby presents at birth encased in a shiny, thickened, variably erythematous, cellophane-like membrane and is most commonly associated with non-bullous congenital ichthyosiform erythroderma (NCIE), lamellar ichthyosis, and self-healing collodion baby.[7] The collodion membrane acts as a poor barrier due to fissuring which results in increased water and electrolyte loss leading to increased risk of cutaneous infections and sepsis.[8] In ichthyoses and EB also, there in breach in barrier function leading to acute skin failure. Our cases of EBS and EBD had erosions over more than 50% of body surface area which leads to water and electrolyte imbalance and secondary bacterial infections. Hence, in these patient's skin care with proper barrier dressings play a vital role. Their parents should be counselled to avoid trauma to skin to prevent development of new lesions. Ichthyoses are associated with erythroderma if not treated and their proper management requires good emollient care and oral treatment in form of retinoids like acitretin.[9] We started acitretin in both cases of lamellar ichthyosis with emollient care to prevent erythroderma and both of them improved significantly.

Vascular tumors, including port-wine stains (PWS) and hemangiomas near vital structures may be life-threatening as they are associated with systemic involvement like

PHACES (Posterior fossa brain abnormalities, Hemangiomas, arterial malformations, coarctation of the aorta and other cardiac defects, and eye abnormalities) syndrome. Hence, they may require urgent management including diagnostic workup, referral for specialty evaluation, and/or early introduction of therapy.[10]

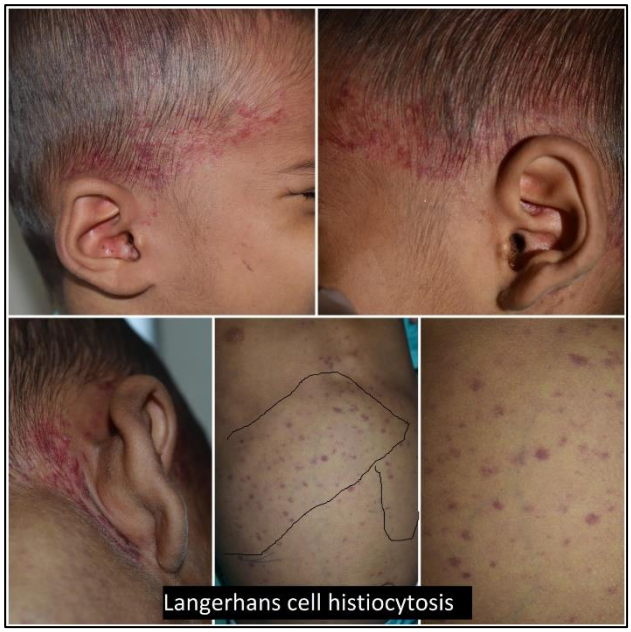

Figure 7: Langerhans cell histiocytosis of erythematous crusted papules in a seborrhoeic dermatitis distribution with petechiae over abdomen with hepatosplenomegaly.

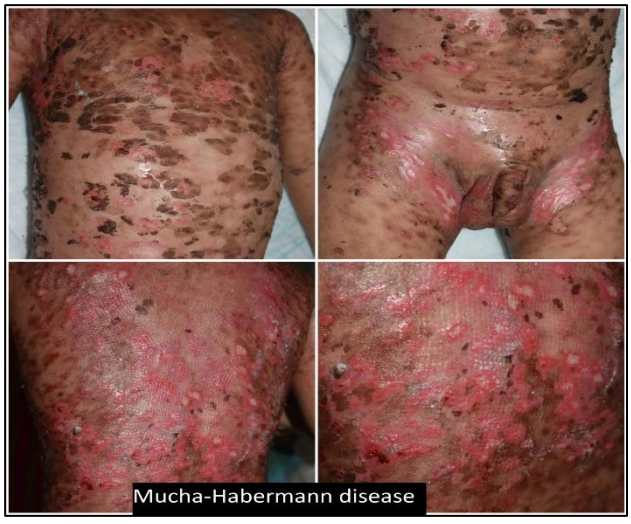

Figure 8: Multiple erythematous, purpuric papular, and vesicobullous lesions on trunk, face, and extremities with few necrotic plaques over abdomen.

Primary cutaneous infections like staphylococcal scalded skin syndrome (SSSS), NF, herpes simplex infections are PDE. We had one case of NF which was treated aggressively with antibiotics and fluid resuscitation with surgical consultation for debridement. The prime concern is early surgical debridement to remove all affected, necrotic tissue. Pediatric mortality rates are as high as 5.4% for NF and hence should be treated promptly.[11]

CADR are more common in boys younger than 3 years and girls older than 9 years.[12] EM is a hypersensitivity reaction occuring in response to infections, medications, or other illnesses. EM major is usually seen in 25–60% of patients, with mostly oral involvement.[13] SJS and TEN are life threatening immune-mediated hypersensitivity reactions involving the skin and mucous membranes. They are primarily caused by drugs, mainly sulfonamides, NSAIDs, allopurinol, penicillins, and anticonvulsants. Mortality estimates for SJS and TEN are as high as 1–5% and 25–35%, respectively and hence prompt management is required. Withdrawing the offending drug is essential and in extensive epidermal detachment, patients should be transferred to pediatric intensive care unit (PICU) to optimize wound care, prevent infections, and attend to hydration and nutrition.[14]

The rare cases seen in this study like PF, BM, MHD were difficult to diagnose and required strong suspicion and clinicopathological correlation. PF is a life-threatening emergency characterized by skin necrosis and disseminated intravascular coagulation (DIC) which requires rapid diagnosis and treatment. The mortality rate in PF patients is reported to be up to 50%, with the most common causes due to DIC and multi-organ failure and early aggressive supportive treatment and judicious use of surgical procedures improves outcome.[15] We treated our patient of PF with intravenous doxycycline 2.2 mg/kg twice daily for 10 days and the patient improved drastically. In bullous mastocytosis, even though skin is the most common organ involved, other organs like liver, spleen, bones (lytic lesions), gastrointestinal tract, etc., might also be involved with potential risk of experiencing shock and sudden death.[16] MHD is a severe form of pityriasis lichenoides et varioliformis acuta of unknown aetiology characterized by ulcerative and necrotic lesions with systemic manifestations. It has mortality rate of about 15%.[17] Our patient of MHD was treated with intravenous piperacillin and tazobactam at a dose of 60 mg/kg every 6 hours and oral prednisolone was started at 1 mg/kg/day and tapered down gradually. The patient responded well with complete healing of lesions resolving with post-inflammatory hyperpigmentation. LCH has been classified as single system (unifocal and multifocal) and multisystem (low risk and high risk). High risk multisystem is involvement of risk organs like liver, spleen and hematopoietic system and its overall survival rate is 84%.[18] Hence, management of multisystem LCH is challenging and requires early intervention.

Limitations

Nelson's score to assess severity of sick children could not be applied. Sample size of the study was small and duration of the study was short due to pandemic. Hence, in future more studies should be taken with large sample size

using Nelson's score system to assess the severity of paediatric dermatological emergencies.

CONCLUSION

To conclude, pediatric dermatological emergencies require an interdisciplinary approach for timely intervention. Also, standardized protocol should be followed in their management to prevent complications like acute skin failure. These patients should be admitted in PICU whenever possible for vital status monitoring especially in patients with multisystem diseases such as vasculitis, rickettsia infections. Early referral of these patients with primary skin conditions to the dermatologists might result in better clinical outcome and decreased mortality. Mortality rate of our study was only 3.33% which defines the role of early consultation and our prompt and effective management.

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Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

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