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ORIGINAL ARTICLE

Childhood Erythroderma: A Prospective Clinical Study in a Tertiary Care Teaching Hospital

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ABSTRACT

In a prevalence study, 21 children presenting to the department of skin and v.d with symptoms of exfoliative dermatitis were subjected to a standardized diagnostic procedures including detailed history, clinical examination and laboratory investigations in order to access the etiological factors responsible in each case. The most common etiological factor was drug induced (19.04%) followed by infection (14.28%) and atopic dermatitis (14.28%). Most common complaint was pruritus followed by exfoliation. The girls were affected slightly higher than the boys with a ratio of (4:3). **Keywords:** Exfoliative dermatitis, Erythroderma, Papulo-squamous disorders.

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INTRODUCTION

Exfoliative dermatitis or Erythroderma is an inflammatory dermatosis characterized by erythema, scaling and thickening of skin involving greater than 90% of BSA. It was first described in 1868 by Von Hebra. It is a less common entity in childhood [1] and can be induced by many pre- existing dermatoses, drugs or any underlying systemic disorder. When it arises on normal looking skin due to underlying systemic disorder or due to drug reaction, it is called as primary erythroderma whereas it is called as secondary erythroderma when it arises from pre-existing dermatoses. Children may present at different age depending upon the aetiology [2]. The course is more prolonged and severe in childhood and there is a difficulty in diagnosis and management of this condition. So, it is imperative for the pediatricians and dermatologists to precisely evaluate and properly manage this condition. For reaching at the cause of erythroderma, thorough history regarding family history of psoriasis, Pityriasis rubra pilaris (PRP) or atopy; age of onset (may be congenital in cases of ichthyosis, infections or immunodeficiency disorders); history of pruritus, drug intake, blood transfusions, recurrent infections and topical applications should be asked for. Detailed cutaneous examination like morphology and distribution of lesions, spared areas, type of scales, palmoplantar thickening, scalp and mucous membrane involvement, skin tenderness. nikolsky's sign, lymphadenopathy, etc. One should also look for associated nail dystrophy and hair loss/dystrophy, although long standing erythroderma may also result in nail and hair changes irrespective of the etiology. Along with these it is also important to look for vitals like temperature, pulse, respiratory rate and blood pressure.

MATERIALS AND METHODS

It is a prospective clinical study. Children from 1-15 years of age clinically diagnosed with erythroderma are enrolled in our study. Study period was 2 years from December 2013 to December 2015 in the department of dermatology, in a tertiary care P.G teaching hospital. All patients were subjected to detailed history, general and dermatological examination. Routine investigations of complete hemogram, urine, skin and mucosal swabs for Gram's stain and KOH were done. Radiological tests of X-ray chest and USG abdomen and serological markers for HIV, Hepatitis and Herpes virus, Immunoglobulin and complement study, and skin biopsy were done where indicated.

RESULTS

This study is an observational clinical study of patients of pediatric age group with exfoliative dermatitis or erythroderma.

Total number of patients enrolled was 21 with 9 boys and 12 girls. 7 patients were between 1 to 5 years of age (3 boys and 4 girls), 10 patients were in the age group of 6 to 10 years (4 boys and 6 girls) and 4 patients were between 11 to 15 years of age (2 boys and 2 girls). Boy to girl ratio was found out to be 3:4. (Table 1)

Most common complaint was pruritus followed by scaling and skin irritation. On dermatological examination all patients had skin erythema, thickness and scaling with >90% body surface area (BSA) involvement. Face was seen as initial site of lesions in younger age group below 5 years of age irrespective of sex where as trunk was seen as initial site of lesion followed by scalp in children of higher age group of more than 5 years of age. On muco-cutaneous examination oral mucosa was affected in about 10% cases. Scalp showed diffuse scaling, crusting with or without alopecia in about 36% cases. Palms and soles involvement was seen in about 15% cases and nail changes in form of pitting, shining discolouration, beau's lines, paronychia was observed in about 22% cases.

Drug induced erythroderma was seen as common etiological factor (19.04%), followed by Atopic eczema, Psoriasis, Infections, PRP etc. (Table 2)

DISCUSSION

Erythroderma in children is an uncommon and difficult entity to diagnose and manage. The common causes in pre-school and school going children include ichthyosis & associated syndromes (Fig 1), atopic dermatitis, scabies infestation; infections like staphylococcal scalded skin syndrome, scarlet fever, toxic shock syndrome, etc; drugs; papulo-squamous disorders such as psoriasis, seborrheic dermatitis, Pityriasis rubra pilaris (PRP), etc; nutritional deficiency such as protein energy malnutrition, acrodermatitis enteropathica; and immunodeficiency syndromes [3].



 $Fig\ 1.\ Ich thyosis\ presenting\ as\ erythroderma\ with\ generalize\ erythema\ and\ scaling.$

A study conducted by Kalsy and Puri [14] included 14 children with erythroderma; drugs were the most common cause of childhood erythroderma accounting for 42.8% (6 cases), followed by psoriasis and ichthyosis 35.7% (5 cases), atopic dermatitis- 14.3% (2 cases) and crusted scabies- 7.1% (1 case). Whereas, in a study by Sarkar and Garg, [5] including neonates and infants also in their study, found infections (40%) were the most common cause of childhood erythroderma, followed by icthyosiform erythroderma(25%), atopic dermatitis (15%), infantile seborrheic dermatitis (10%) and the cause was unidentified in 10% cases.

Drug induced erythroderma was the most common cause in our study. The commonest offending drug was Dapsone causing Dapsone syndrome with fever, hepatitis, lymphadenopathy and skin rash of usually maculopapular / exfoliative dermatitis type; in its complete form (Fig 2). Interestingly, in almost half of

the patients one or more manifestations may be missing. This syndrome develops usually 4-6 weeks after starting dapsone. Continued drug administration results in rapid deterioration while recovery occurs on its discontinuation [6]. The second most common offending drugs were antiepileptic given for seizure disorder [7, 8].

Atopic dermatitis is a frequent manifestation of atopy. It is a chronic inflammatory condition that usually begins in infancy and childhood. It presents with intense pruritus, dry skin, eczematous (erythematous oedematous papules and plaques) or vesicular lesions. Clue for diagnosing erythrodermic atopic dermatitis is personal or family history of atopy i.e. asthma, allergic rhinitis and or conjunctivitis or atopic dermatitis. It typically affects face and extensor surfaces below 2 years of age with sparing of axillae and groins. Sometimes, there may be involvement of area around the mouth because of irritation caused by saliva. As the age progresses, there is more of flexural accentuation [9].

Among the papulo-squamous disorders highest incidence are of PRP and psoriasis (Fig 3). Congenital erythrodermic psoriasis is an extremely rare condition, the incidence being directly proportional to increase in age. There may be history of localized disease or positive family history. Psoriatic nail changes and well circumscribed plaques may be seen at the margins of erythrodermic skin [10]. Unlike adults, plaques are softer, thinner and less scaly in children and it usually begins as recalcitrant diaper dermatitis.

Pityriasis rubra pilaris presents as erythematous papulo-squamous dermatosis characterized by reddish follicular papules and/or plaques with thick, dry scales. The pathognomic finding of PRP is 'island of normal areas within the erythrodermic skin (Fig 4). It is inherited as an autosomal dominant trait with variable expression and reduced penetrance. Most of familial cases are of type 5 (atypical juvenile), rest belong to either type 3 (classic juvenile) or type 4 (circumscribed juvenile) [11].



Fig 2. Dapsone Syndrome presenting with diffuse erythematous eruption on face and hand.



Fig 3. Children with erythryodermic psoriasis.







Fig 4. A girl child with manifestations of pityriasis rubra pilaris (PRP).

Table 1: Age and sex wise distribution of patients

Age group	Number of patients	Boys	Girls
1-5 years	7	3	4
6-10 years	10	4	6
11- 15 years	4	2	2

Table 2: Etiological factor in patients

Cause of erythroderma	Number of patients	Percentage
Drugs	4	19.04%
Atopic eczema	3	14.28%
Psoriasis	3	14.28%
Ichthyosis	2	9.52%
Pityriasis rubra pilaris	2	9.52%
Infections	3	14.28%
Nutritional	1	4.76%
Seborrheic dermatitis	1	4.76%
TEN	1	4.76%
Idiopathic	1	4.76%

CONCLUSION

It is essential for dermatologists and pediatricians to recognize erythroderma and distinguish it from eczema and psoriasis. The aim should then be to search for underlying disorders, some of which have a specific treatment. There is a favorable response to treatment in cases where erythroderma is caused by drugs, nutritional deficiencies, seborrheic dermatitis or infections like Staphylococcal scalded skin syndrome. Erythroderma in children is a well-established entity but has only occasionally received attention. It could be a potentially life-threatening condition if due care is not given at the initial stages of the disease. However, diagnosing this condition remains a challenge due to poor specificity of clinical and histological signs. Since our study found drugs as most common etiological factor in children from 1 – 15 years of age, a detailed drug history is of great importance in diagnosing the condition early which can reduce the overall outcome in many patients. Careful monitoring of the patient and correction of the hematologic, biochemical and metabolic imbalance when required would improve the final outcome in these patients. Moreover the parents should be advised to avoid any kind of self medication including local application of herbal preparations.

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