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Bullous disease of childhood in Srikakulam district

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Abstract:
Background & objectives: *Chronic bullous disease of childhood is a rare autoimmune skin condition which results in clusters of blisters developing in rings often on the face or genitals. The aim of the present study was to estimate the incidence of the bullous dermatoses in childhood*
Materials & methods: *All the patients (n=30) were subjected to detailed history taking and clinical examination. Their particulars regarding age, sex, occupation, personal and family history, presenting complaints, duration, general condition and finding on clinical examination were recorded.*
Results: *The incidence of Bullous dermatoses of childhood is 0.1% of the total new child dermatological cases seen in Rajiv Gandhi Institute of Medical Sciences (RIMS)/ Government General Hospital, Srikakulam, Andhra Pradesh in 18 months. In most of the bullous dermatoses of childhood, the underlying cause is infections (52.8%). The underlying cause is undetectable in the remaining bullous dermatoses. Associated diseases include, scabies, Papular Urticaria, and rarely Pulmunary Tuberculosis. The histological changes are consistant with clinical diagnosis in majority of the cases except in the variants of epidermolysis bullosa (EB).*
Conclusion: *The underlying cause is undetectable in the remaining bullous dermatoses. Associated diseases include scabies, URTI, Papular Urticasia, and rarely Pulmunary Tuberculosis.*
Key words: *Chronic bullous disease, scabies, Papular Urticaria*

I. Introduction

Chronic bullous disease of childhood is a rare autoimmune skin condition which results in clusters of blisters developing in rings often on the face or genitals. In autoimmune disorders, antibodies – which usually seek and destroy foreign invaders to the body, such as viruses – attack the body's own cells instead. This causes inflammation and damage. Other autoimmune disorders include some types of arthritis and inflammatory bowel disease. Chronic bullous disease of childhood is also known as chronic bullous dermatosis or linear IgA dermatosis of childhood. There is a similar condition called IgA dermatosis which affects adults but its development and treatment are different from that in children (1).

The main feature is the development of clusters of blisters in specific areas of the body. These blisters are usually itchy and uncomfortable. Some children have many of these clusters, others have only a few. They tend to appear in phases, with new clusters of blisters appearing in the same area as previous ones (1).

Srikakulam is a district head quarters town in Andhra Pradesh, INDIA on the east coast with a population of 2,703,114 (2011 Census). It has about 5,837 Sq.Km. area. The child population of the district is 281,037. (2011 Census). As the clinical and histopathological reports of Bullous dermatoses of childhood is scarce in India, it is thought to be worthwhile to study in detail the clinical and histopathological features.
Therefore, the aim of the present study was to estimate the incidence of the bullous dermatoses in childhood period in patients attending a teaching Department of Dermatology, venereology and leprology.

II. Materials and methods

The study was approved by Institute ethics committee of Rajiv Gandhi Institute of Medical Sciences (RIMS), Srikakulam, Andhra Pradesh, India. A total of 30 cases clinically diagnized as Bullous dermatoses of childhood in both sexes attending the OPD of Dermatology, Venneology and leprology, Government General Hospital, Srikakulam were selected for this study.

The study was conducted for a period of eighteen months from January, 2014 to june, 2015 at Government General Hospital, RIMS, Srikakulam. All the patients were subjected to detailed history taking and clinical examination. Their particulars regarding age, sex, occupation, personal and family history, presenting complaints, duration, general condition and finding on clinical examination were recorded.

Routine haemetological and biochemical investigations were done and reports recorded. Skin biopsy was done for all cases and the specimens were sent to Department of Pathology, RIMS, Srikakulam and histological features were studied and noted. Following treatment some patients had remission. There were relapses, but they were recorded only once.

The most characteristic early lesion of the disease was choosen for biopsy.

iopsy was performed taking our adequate bit of the skin with infact early bullous lesion including a part of the subcutaneous tissue.

The area selected is cleaned with spirit and infiltrated with 2% Xylocaine locally after a test dose previously. An elliptical incision of about (12mm long .5mm width) the size of the early lesion including a small portion of non blistering area and the bit along with subcutaneous tissue is removed and immediately placed in a labeled bottle containing 10% formalin solution.

The specimen along with requisition form containing particulars & clinical diagnosis was transported for histopathological examination to the Department of Pathology, RIMS, Srikakulam.

The sectioning, staining and microscopic examination were carried out in the Department of Pathology. The sections were stained with Hematoxylin and Eosin and examined under the Microscope.

III. Results

The age, gender and clinical characteristics like sites, symptoms and duration of disease is depicted in Table 1.

Table 2 shows the cutaneous findings and etiological diagnosis of study participants.

IV. Principal Findings

1. Bullous impetigo:- 15 out of 30 patients

In all 15 cases, the epidermis splits were present just below the stratum granulosum large blisters. In two cases, Acantholytic cells were observed. The upper dermis was studded with inflammatory infiltrate of neutrophils and lympho cytes. In some cases, sections of sarcoptes Scabiei and its eggs were observed. These 15 Cases histopathologically confirmed the diagnosis as bullous impetigo with some were with scabies.

2. Chronic Bullous dermatisis of childhood:- (5 out of 30 patients)

In 5 cases, the bulla formation observed. Subepidermally 1 contain numerous eosinophils suggestive of pemphigoid, in some neutrophils predominate and dermal capillary micro-abscesses, suggesting, dermatitis herpitiformis of childhood.

3. Epidermolysis bullosa (EB):- (5 Cases out of 30 patients)

In 5 cases, the bullous formation is within the upperdermis and that some dermal components may be attached to the roof of the blister. In these cases, histologically confirmed the diagnosis as Epodermolysis bullosa. There were small keratin cysts lined by a stratified epithelium in connection with ecrine sweat ducts were observed. These were histologically diagnosed as milia. Clinically milia were observed in dystrophic EB in areas of scarring.

4. Staphylococcal Scalded Skin Syndrome (SSSS):-

In one case, the intra-epidermal split was observed just below the Granular layer,there was acantholysis clinically it was staphylococcal scalded skin syndrome,histologically it was confirmed.

5. Toxic epidermal necrolysis (TEN) :-

In one case the whole epidermis necrosed and there was subepidermal blister formation, with damage to basal cells. Histologically it was confirmed as TEN.

6) **Dermatitis Hapetiformis (DH)** :-

In one case, the bulla formation was observed subepidermally with micro-abscesses at the tips of the dermal papillae with mentrophils and eosinophils accumulation Histologically the diagnosis of DH was confirmed.

7) **Pemphigus Foliaceus** :-

In one case, bulla formation was observed subcorneally and dyskeratotic cells, in the grameler layer. In some fields of the section 1 there is acanthosis and parakeratosis were observed. This indicates the presence of older lesions. Histologically confirmed the clinical diagnosis of Pemphigus Foliaceus.

8) Bullous icthyosiform Erythromderms(BIE):-

In one case, the epidermis was thickened, hyper keretosis and acanthosis, with bizarre vacuolization of the epidermal granular cells.

In one field, there was a subcorneal blister formation was observed. Therefore histologically, the case was confirmed as Bullous icthyosiform erythroderma.

V. Discussion

The primary bullous dermatoses of childhood are a well recognized group of skin diseases of fairly occurance(2). The skin of the infant in greatly increased suscptability to superficial bacterial injections. Particularly by micrococci. This is seen in children and manu born suffering from impetigo. The cohesion at the dermo-epidermal junction in infants is apparently less firm than in adults, and certain diseases with cohesion rarely or fever produce bullous reactions in the adult may do so in the child (3).

The incidence of Bullous dermatosis of childhood group who attended the OPD, Government General Hospital, Srikakulam, Andhra Pradesh during the period of January, 2014 to june, 2015 was formed to be 0.1%.

In Britain, EB all dominant types, constitutes an incidence of 1 in 50,000 whereas recessive types, the incidence is 1 in 3,00,000 (4). The incidence of DH in 20-25 years age group is 1 in 800 (5). Bullous dermatosis like incontinentia pigmenti are almost always seen in females.

DH is more common in males. The remaining bullous dermatoses are observed more or less equal in both sexes. The exact sex ratio for Bullous impetigo in children is not available in the literature. In the present study 60% cases are female children and 40% are male children. In Chromic bullous dermatosis of childhood (CBDC) the ratio of male, female patients was 4% In EB male, female ratio was 3:2. Females are less affected. The pemphigus group affects both sexes almost equally, although under the age of 20 there is a predilection for women.

In the present study, one case of pemphigus foliaceus was a female child aged 8 years.

VI. Consanguineous Marriage:-

In EB, both autosomal dominant & recessive types are present.

In the present study, there is a history of Consanguineous marriage in the parents of four (4) cases out of five cases.

Two cases are brother and sister and the rest are two sisters.

BIE is inherited as a dominant disease. In this study, one male child is a product of Consanguineous marriage, suffering from Bullom icthyosiform Erythroderme (BIE). This is the only case recorded in this study.

Age:

In this present study the bullous dermetoses were observed in a large majority of cases in 4-5 years.

The mean age was 4 – 7 Years.

The median age is 4 – 5 Years.

Occupation

Child labourers constituted a large group with mechano-bullous disorders. Bullous impetigo is more common in school going students due to over crouding and contacts.

Duration Of Illness

In Bullous impetigo, the bullae, persists for 2 to 3 days (6). In this study 1 the bullae persisted for 2 days an average and ruptured spontaneously, leaving behind brownish yellow crusts. Most of the lesions subsided within a week.

In EB. The duration of illness depends upon the type. In lethal variants, the children die within two years.

Lesions of non lethal variants starts from birth and persist into adult life.

In this study, in 3 out of 5 cases of Dystrophic EB, the bullae started at birth and they are occuring over since following Trauma.

In two other Dystrophic EB cases with localized congenital absence of skin (Bart's Syndrome), the lesions started at birth. In CBDC Spontaneous remission known to occur in a majority of patients in about 3 – 4 years (7). In this study the lesions subsided within a week. Dapsone Therapy and some with systemic steroids. No relapses were found during the study period.

In TEN, the mortality rate reported in the drug induced group is about 25%. In this study, one case of TEN was an anti TB (ATT) treatment for Pul TB for the past one month.

Ethambutol is one of the known drugs to produce TEN (8). In this case, INH, Rifampoicin, Ethambutol were being administered. The Child was managed with systemic steroids, appropriate antibiotics to prevent secondary infection, eye- care, and adequate hydration. The child recovered completely from TEN.

SSSS if adequately treated, carries an excellent prognosis (9). In this study, one child had SSSS due to Umbilical Sepsis. The child was treated with Penicillin, and recovered completely within a week.

Penphigus Foliaceus (PF) the onset is slow the disease remains limited for long periods of time with periodic exacerbations, which may or may not become generalized . In this study, one female child, with pemphigus foliaceus of one year duration was treated by General practitioners with inadequate doses of oral steroids and sudden withdrawal precipitated Erythroderma. She was admitted in our hospital and treated with adequate doses of steroids. The disease was remitted.

Bullou Icthyosiform Erythrodema (BIE) manifests shortly after birth and may persists into adult life(10). In this study, in a child aged one year, the blistering was developing on and off. The blistering process was responding to systemic steroids.

In Dematitis hepitiforms (DH), the duration of the disease runs very long course with exacerbations and remissions. In this study are child developed the disease at the age of 9 years had frequent attacks of blisters of DH.

Symtoms

The common symptoms in the present study include blistering (100%), Crusting (100%), erosions(62.7%), Scaling (56%), Itching (46.2%), Ulceration(36.3%) and others (10%). Loss of nails & caries tooth constituted 2% cases.

Site Of Involvement

The site of initial, involvement from the history was a useful clue in arriving at a diagnosis. In cases of EB the initial sites are commonly Trauma prone areas, like palms & soles, oralcavity due to milk sucking from the mother.

In this study, in all 5 cases the initial involvement was Trauma Prone Areas.

The face is the commonest site for Bullous impetigo. In this study, 10 out of 15 cases presented with bullous lesions over the face.

TEN commenced from face and chest and rapidly progressed to involve the rest of the body.

Pemphigus foliacius initially appeared over the lower part of trunk.

Signs

Blistering and Crusting	-	100%	
Erythoma		-	62.7%
Bulls spread sign	-	60%	
Scaling	-	58.1%	
Pigmentation		-	50%
Nikolsky's Sign	-	10%	
Epidermal necrosis		-	1 Case.

Nail changes secondary to Dystrophic – EB was observed in 2 Cases. In one case, there was complete loss of 20 nails. The nail changes were also observed in TEN. In 7 out of 30 cases there was preexisting dermatoses in this study. The pre-existing dermatoses predispose to Impetigo.

VII. Conclusion:

From this study, it is concluded that, The incidence of Bullous dermatoses of childhood is 0.1% of the total new child dermatological cases seen in Rajiv Gandhi Institute of Medical Sciences (RIMS)/ Government General Hospital, Srikakulam, Andhra Pradesh in 18 months. The dermatoses occur mainly in the 4 – 5 years age group and the mean age is 4.7 years and the median age is 4.5 years. The dermatoses occur more or less equally in both sexes. Occupation has no major role over the causation of blisters except in mechano-bullous disorders. The mean and median duration of illness are 1.1 year and one month respectively. Presenting symptom in majority of child patients is blistering with or without erythema. The description of early lesions and the site of initial involvement gives a clue to the underlying dermatoses. Injudicious administration of systemic steroids and its sudden withdrawal in the management of pre-existing dermatoses makes the disease process become generalized. The clinical features of Bullous Dermatoses of Childhood includes erythema, blistering, crusting, scaling etc. In most of the bullous dermatoses of childhood, the underlying cause is infections (52.8%). The underlying cause is undetectable in the remaining bullous dermatoses. Associated diseases include, scabies, URTI, Papular Urticasia, and rarely Pulmunary Tuberculosis. The histological changes are consistant with clinical diagnosis in majority of the cases except in the variants of epidermolysis bullosa (EB).

Conflicts of interest: None.

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Table 1: Clinical characteristics of the study participants.

Sl.No	Parameter	Observation (n=30)
1	Incidence	0.1% observed out of 26,208 child patients
2	Age (Years)	4.5 (mean)
3	Gender Male Female	 15 15
4	Duration of disorder	1 month to 5 years
5	Symptoms Blisters Crusting Erosions	 100% 100% 62%
6	Sites Face Hands & Feet Trunk – Buttocks & Thighs	 23% 20% 16.5% 13%

Table 2: Cutaneous findings and etiological diagnosis of study participants.

Sl.no	Parameter	Observation (n=30)
1	**Cutaneous findings** Bulla Crusting Erythema Bulla Spreading Sign Scaling Pigmentation Excoriation Papules & Papulo Pustules	 100% 100% 62.7% 60% 56% 50% 33% 23%
2	**Etiological diagnosis** Chronic Bullous Dermatosis of Childhood Bullous Impetigo Staphylococcal Scalded Skin Syndrome Toxic Epirdamal necrolysis Bullous iethyosiform Erythroderma Epidermolysis Bullosa Pemphigus Foliaccous Dermatitis herpitiformis	 5 Cases 15 Cases 1 Case 1 Case 1 Case 5 Cases 1 Case 1 Case