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Research Article

INCIDENCE AND CHARACTERISTICS, PATHOLOGICAL CONDITION OF BULLOUS PEMPHIGOID IN PEDIATRIC GROUP

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ABSTRACT

Bullous problems are a complicated organization of disorders which remain an enigma and a project the clinical profession in trendy and the dermatologists and pediatricians specially. Pediatric cutaneous bullous disorders involve extra consideration due to the fact the connection between epidermis and dermis isn't well residential, pores and skin is thinner with better permeability of stratum corneum and absence of first hand history in newborn and infants. The present study aims to incidence and characteristics, pathological condition of Bullous pemphigoid in pediatric group. The study group comprises children in age group between 0 and 14 years, presenting with intact blisters attending the Skin and VD outpatient Department, Sri lakshmi Narayana Institute Of Medical Sciences and SreeBalaji Medical college and Hospital . All cases were evaluated by means of standard proforma which included a detailed history, thorough clinical examination with appropriate investigations where ever required. Majority of the cases were in the age range of 5–10 years (63% or 19 cases) followed by 11–14 years (20% or 6 cases), 2–6 years (10% or 3 cases), and 0–2 years (7% or 2 cases), respectively. The mean age of the study population was 9.36± 3.62 years (mean ± SD). A positive family history was associated with all the subtypes of EB. Rest of the diseases had proven no circle of relatives history of similar diseases. Total 17 (57%) were male and 13 (43%) were female. Male: female ratio was 1.3:2, which reflected an overall male prevalence.DIF became no longer an alternative to histopathology however complementary to the latter. A right correlation between clinical, histopathological, and DIF findings is vital for the very last analysis of VB problems.

Keywords :-Direct Immunofluorescence, Cicatricial Pemphigoid, Epidermolysis Bullosa.

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INTRODUCTION

Skin diseases are not unusual in infants and children account for at the least 30% of all outpatient visits to pediatricians to a dermatologist. Pediatric cutaneous bullous disorders involve extra consideration due to the fact the connection between epidermis and dermis isn't well residential, pores and skin is thinner with better permeability of stratum corneum and absence of first hand history in newborn and toddlers.[1]Blistering issues are heterogeneous. It can be both immunobullous (which happens because of autoimmune etiology) and mechanobullous (springing up due to genetic structural illness). Immunobullous problems include pemphigus group of illnesses, bullous pemphigoid (BP), cicatricial pemphigoid (CP), dermatitis and herpetiformis (DH). Mechanobullous problems consist of one of kind classes of epidermolysis bullosa (EB). According to the plane of blister formation, it is able to be intraepidermal and subepidermal.

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Clinical characteristic of formative years BP seems to extensively have age peaks: infantile and childhood forms. The infantile shape happens within the first year of existence and offers predominantly with palmoplantar lesions with or without universal blistering and infrequently with any genital lesions.[2]

The early life form peaks around the age of 8 and has a less identical presentation of lesions with an elevated involvement of the outside genitalia in up to 44% of cases. The two key distinctive functions of childhood BP from maturity BP consist of acral involvement and mucous membrane involvement. [3]

Diagnostic outcomes in infantile and person BP are comparable, but serological exams were not performed systematically in a number of the said instances. [4] The gold fashionable for diagnosis is direct immunofluorescence microscopy (DIF). However, little statistics is to be had on the interpretation of ELISA stages, inflammatory markers or blood cellular counts in babies. Further expertise, specifically approximately the relevance of ELISA degrees might help to evaluate sickness severity and therefore influence the choice of medication or duration of remedy. [5]

Concerning the remedy of infantile BP, first line treatment commonly includes topical or systemic corticosteroids. However, there aren't any stringent healing standards and there was very little discussion on the distinctive options for 2d line remedy. Furthermore, in scientific consensus hints on remedy of BP, there is little or no, if any, information on remedy in toddlers. [6]

Only limited information is available about frequency of specific skin diseases in children and due to the rare incidence of each of these diseases in children, most of the cases have been reported as case reports and there are no previous studies regarding the pattern of Bullous pemphigoid diseases exclusively in pediatric age group. The present study is an attempt to Incidence and characteristics, pathological condition of Bullous pemphigoid in pediatric group.

MATERIAL AND METHODS

The present study was undertaken in the Department of dermatology Sri Lakshmi Narayana Institute Of Medical Sciences and Balaji Medical college and Hospital.We have included 50 children on an outpatient basis below the age of 18 years who had presented with the clinical features suggestive of BP disorders. We have excluded the patients who were either moribund or having the history of trauma, burn injury, allergic dermatitis, insect bite, or bulla formation due to infections like herpes or impetigo. Inclusion criteria: Only children between age groupof 0-14 years, presenting with intact blisters, bothsexes, parent/guardian giving verbal consent forthe study

All the cases were subjected to a thorough history taking including name, age, sex, address, religion, economic status of the family along with chief complaints, total duration of disease, related past, family and treatment history, complete general physical, local and systemic examination. Routine investigations were carried out in all the cases, while special investigations like Tzanck smear (cytology),bacterial smear and culture, histopathological examination and IMF have a look at became restrained to simplest few cases in which they carried diagnostic significance.

Skin biopsy specimens were obtained maintaining strict asepsis by 3–5 mm punch biopsy from the lesional and perilesional areas under local anaesthesia with 2% xylocaine injection. Lesional samples for histopathological examination were collected in a clean container filled with 10% neutral buffered formaldehyde solution. Perilesional skin biopsies for DIF were taken in a clean container filled with normal saline. In suspected cases of EB, an additional biopsy was taken from an artificial bulla formed by rubbing a normal area of skin with an eraser. All skin biopsy specimens were examined by two trained pathologists at our institute using light microscope & immunofluorescence microscope.

Hematoxylin & Eosin (H&E) stain was used for light microscopy. Histopathological findings were noted in an algorithmic manner moving from superficial to deep dermal layer including level of blister, blister content, row of tombstone appearance, festooning of dermal papillae, intradermal perivascular inflammation, etc.,

Specimens for DIF study were stained using fluorescein isothiocyanate (FITC)‑conjugated polyclonal rabbit antisera against human IgG, IgM, IgA, and C3 (DAKO Denmark). The nature, site, and pattern of immune deposit were noted under an immuno fluorescence microscope, and the findings were interpreted in an algorithmic manner

RESULTS

We got total 30 cases of Cutaneous BP disorder in pediatric age group, and the incidence was 27.7% of total cases with VB lesions. The youngest was a 1‑year‑old boy and 14‑year‑old girl was the oldest. Majority of the cases were in the age range of 5–10 years (63% or 19 cases) followed by 11–14 years (20% or 6 cases), 2–6 years (10% or 3 cases), and 0–2 years (7% or 2 cases), respectively. The mean age of the study population was 9.36± 3.62 years (mean ± SD). A positive family history was associated with all the subtypes of EB. Rest of the diseases had proven no circle of relatives history of similar diseases. Total 17 (57%) were male and 13 (43%) were female. Male: female ratio was 1.3:2, which reflected an overall male prevalence.

After considering the clinical data, histopathological findings, DIF findings including the SST and IFM, it was concluded that LABD was the most common bullous disorder constituting BP 50% (15 cases) ,PF 17% (5 cases), DEB 20% (6 cases) , EBS 7% (2 cases) and PNP 6.6% (2 cases) .

Between the whole study population, the disease appearance was insidious in nature in 73% cases (22 cases) and acute in only 27% cases (8cases).

There was maximum involvement of the Trunk involvement was observed in cases of BP 56% (17 out of 30 cases) followed by PF23% (7out of 30 cases).Lower limbs were the most common site of blister formation in BP 20% (6 out of 50 cases). Diffuse involvement of the whole body was characteristically found in all cases of EB. The commonest site of mucosal involvement was oral mucosa [Figure 7b], which was involved in 43% of all cases

In total, 70% (7 out of 20) cases of PV and 55% cases of BP showed oral mucosal involvement. In total, 25% (5 out of 20) cases of PV and 100% cases of PNP displayed nasal mucosal involvement. Conjunctival involvement was found in two case of PV.

On light microscopic examination, all cases of PV presented with suprabasal bullae containing acantholytic cells. Subcorneal blisters have been located in all instances of pemphigus foliaceous. The blister cavity was filled with acantholytic cells in 75% (3 out of 9) cases and proteinaceous fluid in 25% (1 out of 9) cases. All cases of PNP provided with suprabasal bullae containing mixed factors, viz. acantholytic cells, neutrophils, eosinophils, etc. “Row of tombstone” appearance was seen in all cases of PNP. Subepidermal bullae were found in all cases of BP and In cases of BP, blister cavity was filled 10% (1 out of 10) cases. In cases of LABD, blister cavity was filled with neutrophils in 66.6% (8 out of 12) cases and proteinaceous fluid in 33.3% (4 out of 12) cases. The most effective case of CP, the blister cavity changed into packed with a aggregate of acantholytic cells, neutrophils, and eosinophils. Festooning of dermal papillae was seen in all cases of BP and CP. Perivascular inflammatory infiltrate was predominantly found in LABD and CP (100% cases) followed by PNP (50% cases) and PV (37.5% cases). Light microscopic findings had been discovered to be statistically vast concerning all of the parameters.

Direct immuno fluorescence (DIF) study findings are shown in. It reveals that the predominant type of antibody deposited was IgG in cases of PV and PF. The site of deposition was intercellular space (ICS) between keratinocytes in a lace‑like Complement C3 was also deposited in three cases (23.7%) only. The cases of PNP showed deposition of IgG and complement C3 predominantly. IgG was found to be deposited in the ICS showing fishnet pattern, whereas C3 was deposited along the dermoepidermal junction (DEJ) in a granular pattern. The subepidermal bullous lesions predominantly showed deposition of IgA observed by using IgG and C3.The pattern and site of deposition were linear and along DEJ, respectively. Subepidermal bullae showing linear IgA deposition along DEJ were finally diagnosed as LABD.

DISCUSSION

VB illnesses are a different group of problems characterized with the aid of the development of vesicles or bullae springing up over the skin and/or mucous membrane. In spite of comparable medical presentations, they're remarkably unique from each different each etiologically and pathologically. Appropriate diagnosis is important to prevent the fatal outcome if untreated especially in pediatric populace. Very few studies in India have dealt with the clinic-pathological functions of pediatric VB illnesses. In the prevailing have a look at, a complete analysis of clinical, pathological, and DIF microscopic features of intraepidermal bullous diseases became completed.

In the present study, most of the cases were in the age range of 5–10 years (63% or 19 cases). The mean age of the study population was 9.36± 3.62 years (mean ± SD). Which is correlated with [7]study of 196 patients with immunobullous disorders, the mean age of the patients at the time of diagnosis was 7.72 ± 5.66 years.

where there was slight male preponderance, which is similar to our study had male: female ratio of 1.3:2 , which reflected an overall male preponderance. In the study 17 (57%) were male and 13 (43%) were female. but this study included immunobullous lesions only.Lower limbs were the most common site of blister formation in BP 50% (15out of 30 cases). A case report on an 9‑year‑old girl with chronic generalized PV showed multiple eruptions and blisters on face, limbs, and trunk. These is compared to [8] study on 31 cases of LABD in Tunisian children revealed that most of the children had a generalized eruption (93%), face was involved in 53% cases, and only two patients had localized lesions on their legs A study done.[9]revealed that the clinical presentation was similar to adults but facial involvement was common in children suffering from BP, contrary to our study.

De Pablo-martinez[10] study on PV, clinical presentation in children was essentially similar to cases on four infants with BP revealed that one out of the four cases had tense bullae with erythema around the blisters, which was similar to present study.

Similar to current study,[11]also found that in blisters of BP, DIF staining of perilesional skin shows linear IgG deposits along BMZ in all patients. Deposition of complement C3 may accompany IgG.

Other immuno reactants such as IgA, IgM, and IgE may also be found rarely. In a case report done on

the LABD of childhood, DIF examination showed strong linear deposition of IgA, IgG, and C3 at the DEJ with similar but moderately strong deposition of IgM (2+) in the same region. Though the cases described by [12-13] had only deposits of IgA with associated IgG appearing in the early stages of the illness but our there is no other immuno reactant likes IgA, IgM, and IgE.

CONCLUSION

This study portrayed that Linear IgA bullous dermatosis become located to be the maximum commonplace kind of bullous disorder in pediatric population, followed by means of PV. Among the genetic bullous problems junctional EB become the commonest one. Majority supplied with cutaneous blisters together with other mucosal involvement, oral mucosal involvement being the most common. Histopathological examination showed the everyday hallmark capabilities that had been conclusive in maximum of the instances except in LABD and EB, necessitating the use of DIF. For subcategorizing EB, ordinary DIF remained inconclusive. IFM proved vital in these cases. Thus, DIF became no longer an alternative to histopathology however complementary to the latter. A right correlation between clinical, histopathological, and DIF findings is vital for the very last analysis of VB problems.

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