

Title – A case of bullous ichthyosiform erythroderma

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INTRODUCTION

- Bullous ichthyosiform erythroderma is an autosomally dominant inherited disorder characterized by erythematous, erosive, and bullous skin lesions over the entire body at birth and abnormal hyperkeratosis on the palmoplantar surfaces as the patient grows older.
- However, there is autosomal recessive inheritance and a high frequency of spontaneous mutations and as many as half the cases have no family history and represent new mutations .
- It caused by a mutation in the keratin 1 (K1) and/or keratin 10 (K10) genes, and most pathogenic mutations are found within the helix initiation and termination motifs of the central helical rod domain (K1 and K10) or the upstream H1 homology domain (K10).
- Ridged hyperkeratosis at the flexures and presence of flaccid bullae is characteristic of this form.
- In the present case neither of the parents nor any other sibling is affected.



CLINICAL FEATURES



CONCLUSION

- A typical sporadic case of Bullous ichthyosiform erythroderma in 12 year old boy with erythroderma, erosion, and blisters on the entire body surface and increase skin markings of palms and soles. Teeth, hair and nails are normal.
- This case is managed by topical emollients and larges dose of vitamin A. Oral retinoids have not been started as there is no fusion of epiphyses yet.