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Case Report

UNUSUAL PRESENTATIONS OF ICHTHYOTIC DISORDERS IN CHILDREN - A CASE SERIES

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ABSTRACT
Ichthyosis are disorders of cornification (Gabriele and Franzika (2012). Clinically characterized by wide spread scaling and varying levels of erythema (Bale and Richard (1993-2001). Both males and females are equally affected. Here by we report a case series of congenital ichthyosiform erythroderma (CIE), Lamellar ichthyosis (LI), Collodion baby at our hospital due to rarity in our population.

Keywords: *Genetically Inherited, Ichthyotic Disorders, Congenital Ichthyosiform Erythroderma, Lamellar Ichthyosis, Collodion Baby*

INTRODUCTION
In ichthyotic disorders abnormal differentiation and desquamation of epidermis result in a defective barrier function (Gabriele and Franzika (2012). It may be inherited or acquired. Autosomal recessive congenital ichthyosis (ARCI) is a heterogenous group of inherited keratinization disorders (Bale and Richard 1993-2001) which were reported at our hospital.

CASE SERIES
Case 1:
15 years old female came with chief complaints of redness and scaling all over the body since birth. History of birth with collodion membrane, history of 3rd degree consanginous marriage, history of decrease in sweating. On examination - diffuse erythema and scaling all over the body. Scales are thin, semiadherent with powdery consistency. Pityriasis amiantacea over scalp. On histopathological examination- mild focal spongiosis, mild parakeratosis, slight hypergranulosis, lamellated orthohyperkeratosis, superficial perivascular lymphocytic infiltrate seen. According to history, clinical features, histopathology diagnosed as Non bullous congenital ichthyosiform erythroderma. Treatment- Emollients, Iron supplements were prescribed, advised intake of more fluids, calories and proteins. For scalp- topical mineral oil, daily salicylic acid shampoo, clobetasol solution twice daily application, tab. cephalexin 250mg twice daily for 2 weeks.

Figure 1

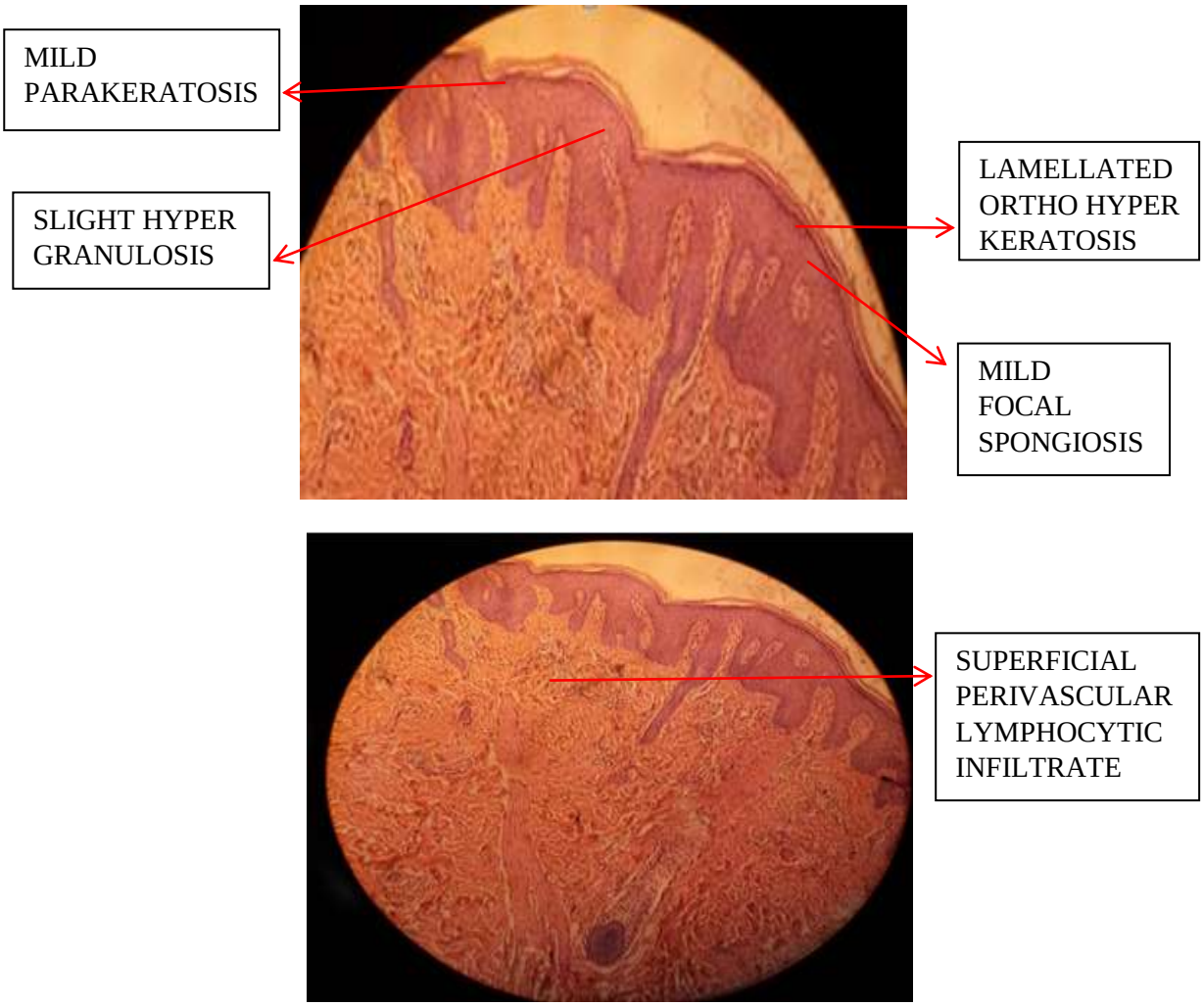

Figure 2

Case 2:
6 years old male. History given by mother. Came with chief complaints of scaling all over body since birth. History of collodion membrane at birth. History of 2^{nd} degree consanginous marriage. On examination-large plate like scales resembling like fish skin all over the body including palms and soles, eclabium present, ectropion and corneal opacity of right eye. On histopathological examination-hyperkeratosis, hypergranulosis, acanthosis seen. By history, clinical examination, histopathological examination diagnosed as lamellar ichthyosis. Treatment- emollients, lubricant eye drops, advised corneal surgery for corneal opacity but parents were not affordable.

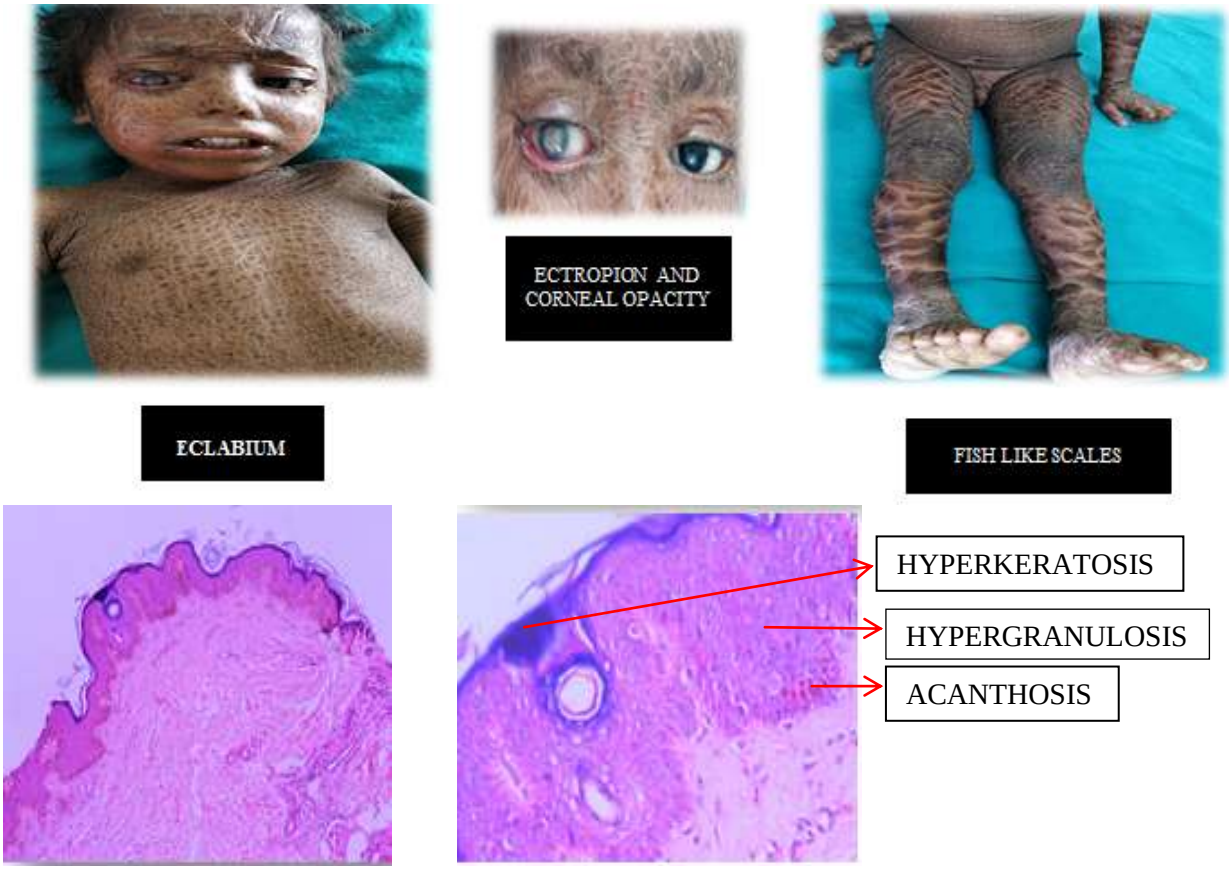

Figure 3

Case 3: Baby girl sibling of case 2. Presented at birth with collodion membrane. Born at our hospital. On examination – shiny, taut, yellowish membrane over body present. Investigations were found to be normal. By clinical examination diagnosed as collodion baby. Treatment- In NICU baby was managed with iv fluids, electrolytes, emollients.

Figure 4

Case 4:

6 months old male baby. History given by mother. Presented with chief compliants of scaling all over body since birth. History of collodion membrane at birth. No history of consanguinity. On examination-

diffuse scaling all over the body including palms and soles, Ectropion of right eye. By history and clinical findings- diagnosed as non bullous congenital ichthyosiform erythroderma. Treatment- emollients, vitamin A supplements, iron supplements, lubricant eye drops for ectropion.

Figure 5

DISCUSSION

The spectrum of autosomal recessive congenital ichthyosis ranges from fatal harlequin ichthyosis to less severe disorders such as LI and CIE (Bale and Richard 1993-2001). Incidence: CIE- 1:1,80,000 Population (Lippincott and Wilkins, 2005). LI- 1:3,00,000 population (Lippincott Williams and Wilkins, 2005). Both LI and CIE have normal life span and associated most commonly with transglutaminase-1 mutations (Oji and Traupe, 2006; Luu *et al.*, 2010). LI symptoms remain severe throughout the life, CIE improve at puberty (Rimoin *et al.*, 2002).

CONCLUSION

It is important to recognise the presentation of congenital ichthyosis (Taieb and Labreze, 2002). Adequate treatment must be initiated in NICU (Lippincott and Wilkins, 2005; Taieb and Labreze, 2002). Health care providers should be aware about spectrum of collodion baby (Taieb and Labreze, 2002). Prognosis of underlying conditions is incredibly diverse. ARCI are rare group of disorders.

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