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A CASE REPORT: LAMELLAR ICHTHYOSIS

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ABSTRACT:
Lamellar ichthyosis (LI), is otherwise called ichthyosis lamellaris and non-bullous inborn ichthyosis, is an uncommon acquired skin problem, influencing around 1 out of 600,000 individuals. Influenced children are brought into the world in a collodion film, a glossy, waxy-seeming external layer to the skin. This is shed 10–14 days after birth, uncovering the principle indication of the infection, broad scaling of the skin brought about by hyperkeratosis. The scales frequently tile the skin and may take after fish scales. The infant influenced with lamellar ichthyosis is likewise called as collodion infant. The eyelids and mouth may resemble being constrained open because of the snugness of the skin. There can be related eversion of the eyelids (ectropion). This condition is generally treated with emollients, creams, fundamental retinoids and eyedrops.

Key words: Lamellar Ichthyosis, erythroderma, dry eyes, waxy skin.

1. INTRODUCTION:

Lamellar ichthyosis (LI) is an autosomal passive problem that is clear upon entering the world and is available all through life. The infant is brought into the world encased in a collodion layer that sheds inside 10-14 days. The shedding of the film uncovers summed up scaling with variable redness of the skin. The scaling might be fine or platelike, looking like fish skin. In spite of the fact that the issue isn't perilous, it is very distorting and makes significant mental pressure influenced patients [1]. Patients with lamellar ichthyosis have quickened epidermal turnover with proliferative hyperkeratosis, as opposed to maintenance hyperkeratosis. This includes a transformation in the quality for transglutaminase 1 (TGM1). There are at any rate 14 distinguished diverse TGM1 transformations [2]. The transglutaminase 1 protein is engaged with the development of the cornified cell envelope. The arrangement of the cornified cell envelope is a basic platform whereupon ordinary intercellular lipid layer development in the layer corneum happens. Along these lines, changes in the TGM1 optionally cause abandons in the intercellular lipid layers in the layer corneum, prompting blemished hindrance capacity of the layer corneum and to the ichthyotic aggregate seen in lamellar ichthyosis patients and in transglutaminase 1 knockout mice. How much an imperfect cornified cell envelope alone adds to the hindrance variation from the norm in ichthyoses stays indistinct [3].The term swimming outfit ichthyosis depicts the trademark appropriation of the injuries, which include the storage compartment, the proximal pieces of the upper appendages, the scalp, and the neck, with saving of the focal face and

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furthest points. This type of lamellar ichthyosis is brought about by a homozygous missense transformation in TGM1 [4,5]. The scales are enormous on the lower limits, with the scales isolated by shallow crevices. Albeit present upon entering the world, during adolescence and into adulthood, erythroderma is ordinarily missing. Inclusion of the acral surfaces can fluctuate from mellow hyperlinearity to serious hyperkeratosis of the palms and soles. The scalp may grow thick, disciple scale that may prompt scarring alopecia [6]. Nail dystrophy and thermoregulatory issues prompting potential seizures are additionally qualities seen in lamellar ichthyosis [7].

2. MATERIAL AND METHOD

The Patient visited MGM Hospital with scales, itching sensation and dry eyes. Caretakers consent was seeked and explained about this case report publication. The Protocol and Written acceptance of them was submitted and got approved from Institutional Human Ethics Committee (IHEC).

3. CASE REPORT

A female child of 4yrs was admitted in dermatology ward, patient is a K/C/O Lamellar Ichthyosis (colloidan baby) since birth, generalized scales which range from fine & white to thick, dark & platelike, scales arranged in Mosaic pattern, dry eyes, waxy skin, alopecia, and her parents' marriage is consanguineous. RBS, Blood urea & serum creatinine are normal. The patient was assessed to have Lamellar Ichthyosis. The patient was treated with acetretin, Xerina Lotion(Glycolic acid, Urea and cetylated fatty ester complex), Salysia KT shampoo (ketoconazole and salicylic acid), Ontears eye drops (carboxymethylcellulose), Cetrizine, Oint. 5% salicylic acid & liq. Paraffin.

Figure 1: Child with Lamellar Ichthyosis.

3. DISCUSSION

In light of history and actual assessment patients evaluation was made. Patients with lamellar ichthyosis have typical life spans .In the neonatal period, following the shedding of the collodion layer, the infant is in danger for auxiliary sepsis and hypernatremic drying out. As the kid ages, the hyperkeratosis can meddle with ordinary perspiration organ work, which can incline to warm bigotry and conceivable warmth stun. Ectropion may bring about the failure to completely close the eyelids and can cause presentation keratitis. Outer hear-able channel stenosis and tympanic film blunting may bring about a conductive hearing misfortune. Osseointegrated hearing gadgets may successfully sidestep this meeting deformity. More uncommon affiliations incorporate muscular anomalies, for example, genu valgum, other visual issues, for example, corneal hole, and rickets [8].Like different ichthyoses, lamellar ichthyosis might be particularly inclined to broad, serious, and

persistent Trichophyton rubrum contaminations and viral diseases [9-11].

4. CONCLUSION

The treatment given to this patient is as per the rules. This generally happens because of hereditary imperfections. The avoidance angle is questionable; the nature of patients life in this condition is poor. Continuous visits to dermatologist is an unquestionable requirement, mental advising can improve patients perspective. Due to this LI patient can't perform the greater part of her everyday exercises, now and again vision may likewise be changed. Guardian is encouraged to keep up the skin hydrated for the duration of the day.

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6. CONFLICT OF INTEREST

The author(s) confirm that this article content has no conflict of interest.

7. AUTHORS CONTRIBUTION

Vineeth Reddy G worked in the Hospital in collection of data, Counseling the patient and their family, etc., Anila Reddy T designed the documents required for the work. Sharavana bhava B.S. discussed and conceived the idea of doing this work and prepared the Protocol.

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