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Case Report on Management and Complication of Nonbullous Congenital Ichthyosis

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Abstract: ***Introduction: A diverse range of skin conditions known as ichthyosis are distinguished by widespread scaling. The T. G. M.1 gene, which produces the transglutaminase I protein, is mutated in the autosomal recessive disorder known as Nonbullous congenital ichthyosis. Changes in epidermal differentiation are reflected in the scales. One of the rarest types of ichthyosis, lamellar, typically causes a newborn to be born with a collodion membrane. An uncommon inherited skin condition known as lamellar ichthyosis causes aberrant skin cell production that causes severe scaling and drying of the skin. Patient information: A male 8-year-old child of a patient was hospitalized. The Patient's mother provided a history of the Patient's difficulty feeding, fluid loss (dehydration), loss of the body's mineral balance (electrolyte imbalance), and fluid loss. Breathing issues, a fluctuating body temperature, body-wide or skin infections. Therapeutic Intervention: Lamellar ichthyoids is a genetic disorder. Hence, there isn't a single therapy or treatment that can cure it. For people with lamellar ichthyosis, some medicines and drugs can help manage their symptoms and enhance their quality of life. Tazorotene is one of the retinoids that can aid in reducing skin thickness and scaling. A dermatologist may prescribe these drugs to assist in controlling the growth and shedding of skin cells. Nursing perspective conclusion: A male 8-year-old child of a patient was also hospitalized. The Patient's mother provided a history of the Patient's feeding difficulties, fluid loss (dehydration), loss of the body's mineral balance (electrolyte imbalance), breathing difficulties, an unstable body temperature, and skin or systemic infections. The medical intervention improved the Patient's condition. The Patient's health has improved, and his symptoms are less severe.***

Keywords: Ectropion, Lamellar ichthyosis, Photophobic

1.Introduction

Nonbullous congenital ichthyosisis a rare genetic skin condition that causes aberrant skin cell production, severe scaling, and skin drynessMutations in the genes that produce the proteins required for healthy skin development and function are the root cause. Nonbullous congenital ichthyosispatients are born with tight, thickened, and "fish-like" skin covered in broad, black scales. These scales can be very dry, which can be very uncomfortable and itchy. The illness affects every body part, including the face, scalp, and nails. Lamellar ichthyosis patients may also experience hearing loss, scarring alopecia, and ectropion, which is the outward turning of the eyelids and skin symptoms [(3)]. Lamellar ichthyosis has no known cause or effective treatment other than symptomatic relief. To avoid dryness and scaling, it is crucial to moisturize the skin frequently and gently. Among other topical drugs, emollients, keratolytics, and retinoids are commonly used to treat the symptoms. Infants with the disorder may need to be bathed using particular techniques to prevent infection and encourage smooth scaling of the scales.[1]

The genetic condition lamellar ichthyosis frequently has an autosomal recessive pattern. Finding out the likelihood of passing the illness on to future generations can be assisted through genetic counselling and testing. Affected individuals and their families also benefit from the resources and emotional support that support groups and counselling programs can offer. Lamellar ichthyosis patients may or may not have erythematous skin but have more significant, darker, platelike scales. Transglutaminase-1 gene (TGM-1) or ABCA12 ATP-binding cassette transporter gene defects have been linked to lamellar ichthyosis in patients.[6] The TGM-1 gene and other genes, including those linked to the lipoxygenase genes ALOX12 and ALOXE3, can also be linked to congenital nonbullous ichthyosiform erythroderma. The affected infant may be born with a collodion membrane in either state, and newborns may not be able to discern between the two conditions [(6).] A severe electrolyte and hydration imbalance could occur.[16] Some newborns' erythroderma may go away during childhood.[2]

Patient-specific information:

The primary concern and symptoms of the patient: A male 8-year-old child of a patient was also hospitalized. The Patient's mother described the Patient's history of eating difficulties, fluid loss (dehydration), loss of the body's mineral balance (electrolyte imbalance), and fluid loss. Breathing issues, unstable body temperature, skin infections or the entire body.

Medical, Family, and Psychology History: So far, two cast applications for the Patient have been finished. The Patient now promptly makes it to the hospital for extra care. He was born into a nuclear family, and no known hereditary diseases existed. The Patient engages in healthy practices such as watching television, playing sports, participating in extracurricular activities, and keeping things tidy

Relevant past intervention with the outcome: For the Patient, there have already been two applications for the cast. For further treatment, the Patient now arrives at the hospital immediately.

Clinical findings: Physical examination: During the physical test, he was oriented and measured 118 cm and 18 kg. The Patient's temperature was 99 degrees Fahrenheit, heart rate was 88 beats per minute, respiration was 18 breaths per minute, spo2 was 98%, Hb% was 11%, WBC count was 7300cumm, R. B. C. count was 4.42 million/cu mm, and platelet count was 3.08 millilitres.

Timeline: The Patient first complained of a congenital abnormality of the left foot after seeing an O. P. D. in a private hospital. Applications for the two-time cast have been submitted thus far. For additional care, the Patient now arrives at the hospital immediately.

Diagnostic assessment: During the physical examination, the Patient's temperature was 99°F, his pulse was 88 beats per minute, his respiration rate was 18 breaths per minute, his spo2 level was 98%, his haemoglobin level was 11%, his WBC count was 7300cumm, his RBc level was 4.42 million/cu mm, and his platelet level was 3.08 mcl. All statuses are also standard.

Therapeuticintervention: Lamellar ichthyosis is a genetic disorder. Hence, there isn't a single therapy or treatment that can cure it. For people with lamellar ichthyosis, some medicines and drugs can help manage their symptoms and enhance their quality of life. Tazorotene is one of the retinoids that can aid in reducing skin thickness and scaling. A dermatologist may prescribe these drugs to assist in controlling the growth and shedding of skin cells.

Followup and outcome: The Patient was the main focus of the follow up plan, as advised by the doctor. Both the Patient's health and the symptoms lessened. The Patient was encouraged to take the medicines that were recommended.

Intervention adherence and compliance:

The intervention needed to be followed.

The complication and adverse event:

There were no unfortunate occurrences reported.

2.Discussion

LI makes the skin dry, flaky, and peeling like parchment paper. Similar to what happened in our situation; the symptoms start at birth and affect the entire bod. These newborns have a collodion membrane that is glossy, taught, and transparent at birth. In a week or two, the membrane begins to desquamate. The mucosa is often unaffected, and there is no erythroderma Ectropion and eclabium occur due to thicker skin structure and pushing on the soft tissues near the lips and conjunctivae, the typical diagnostic signs in practically all instances. Ectropion and eclabium were both seen in our case. The most serious issues for which these newborns are in danger include overheating, increased insensible fluid loss, electrolyte abnormalities, skin infections, and sepsis because of decreased skin integrity. In the initial few days of life, pneumonia can be brought on by aspirating the squamous material in the amniotic fluid-possible difficulties with pulmonary breathing and sucking.[3]

The condition known as ichthyosis lamellaris, or Nonbullous congenital ichthyosis, is a rare hereditary skin condition that changes how skin cells are formed and causes the buildup of dry, thicker scales on the skin's surface. Around 1 in 100, 000 people are thought to be affected globally.

Typically, the disease is inherited autosomally recessively, meaning both parents must have the defective gene for their child to be afflicted. The transglutaminase one protein, crucial for proper skin development and function, is produced by genes often home to the genetic abnormalities that cause these conditions[4].

A collodion membrane, a thick, glossy outer layer of skin, is frequently present in newborns with lamellar ichthyosis. Once this membrane sheds during the first few weeks; they begin to acquire the typical symptoms of the illness, such as extensive, thicker scales covering most of the body. The scales, which can be brown or yellowish, might itch, feel tight, and hurt.

People with lamellar ichthyosis may also struggle with issues outside their physical symptoms. The disorder can impair a person's ability to control body temperature, raise their risk of infection via skin fissures, and harm their self-esteem and quality of life. Lamellar ichthyosis is currently incurable. Thus, the primary goals of therapy are symptom management and averting consequences. To keep the skin moisturized and avoid extreme dryness and scaling, it is essential to apply moisturizers and emollients often and heavily. Dermatologists may also recommend keratolytics, oral retinoids, and topical retinoids to assist in minimizing scaling and enhancing the texture of the skin. Infections may arise and necessitate the use of antibiotics to treat them[5].

3.Conclusion

In this instance, the Patient is an 8-year-old boy with congenital lamellar ichthyosis. Lamellar ichthyosis is an uncommon hereditary skin condition marked by the buildup of thick, dry scales on the skin's surface. It is a chronic illness that needs constant attention and management to lessen symptoms and consequences. There are techniques to manage lamellar ichthyosis symptoms even without a known cure. Frequent and robust moisturization, topical medications, and oral medications can help reduce scaling and improve the skin's texture. A diligent infection surveillance program and appropriate medical care are also crucial. Lamellar ichthyosis can cause both physical and emotional suffering. Affected people and their families must seek help through resources, support networks, and genetic counselling to understand the condition and get guidance for family planning.

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