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Clinical And Genetic and Immuno-Microbiological Aspects of Ichthyosis and Methods of Pathogen Etic Treatment

Eshniyazov Mahmudbek Maksudovich

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Chief Physician of the Khorezm branch of the Republican Specialized Scientific and Practical Medical Center for Dermatovenereology and Cosmetology Ministry of Health of the Republic of Uzbekistan

Abstract: This article is intended to identify a hereditary skin disease in which the process of keratinization of the epidermis is disturbed. The skin dries up and peels off, and in severe cases of the disease, the skin begins to appear on the skin. Often, ichthyosis appears in early childhood and is inherited from parents.

Key words: Sweat, inflammation, patient, surgery, treatment

Introduction

Ichthyosis is a very rare group of congenital skin diseases characterized by a certain degree of proliferation and thickening of the cornea with inflammation of the skin. Severe forms such as Harlequin and Lamellar forms may present with limb and organ involvement, respectively. Patients with sublamellar type have severe ectropion that can lead to corneal ulceration, tearing, and blindness. Treatment involves a multidisciplinary approach that allows for early intervention and treatment that optimizes outcomes. The time of intervention is not clearly defined in the literature. This is probably due to the paucity of data in these conditions, which are relatively rare. However, similar principles can be used to treat early ectropion in burn patients.

Materials and methods. Ichthyosis disease represents a very rare group of monogenetic diseases of cornification. This may be due to systemic symptoms. Clinical features vary considerably from mild to severe forms, but always include inflammation of the skin as well as scaling and thickened stratum corneum. Less common symptoms include pruritus, skin fragility, ectropion, and anhidrosis. includes, they are associated with rare species.

This group of diseases can manifest in mild forms, which usually require topical agents such as emollients for treatment. However, more severe forms that can lead to limb and limb loss usually require timely surgical treatment to save limbs, limbs and life. The Harlequin subtype may present with multiple constrictive plaques surrounding the limbs with decreased limb perfusion. However, patients with lamellar subtype may experience severe ectropion, corneal perforation, and blindness as a result.

Early recognition and surgical treatment are critical to survival and saving limbs and eyes. Most importantly, the timing of surgical intervention is critical for optimal results when working with patients with ichthyosis. Although nonoperative treatment is well established for this group, surgical intervention is less well documented. There are several concerns, including the ideal timing of surgical intervention, the ideal donor site for harvesting full-thickness skin grafts, and whether these patients have impaired wound healing due to skin disease.

It can be presented a patient with ichthyosis congenital with bilateral severe ectropion with whole-body enlargement who was successfully treated in collaboration between an ophthalmologist, dermatologist, and plastic surgeon. Treatment involves early intervention using full-thickness skin grafts from the medial arm.

Results. A 17-day-old male newborn with a history of severe bilateral ectropion presented to our local clinic on the recommendation of our ophthalmology colleagues. A 35-year-old mother with normal G2P1 booking parameters was born by cesarean section at 36 weeks due to fetal bradycardia. Her pregnancy was uneventful. An anomaly scan at 20 weeks' gestation was reportedly normal.

At birth, the child was noted to be grossly enlarged, with severe bilateral ectropion and flattened back ears consistent with lamellar ichthyosis. At birth, his APGAR scores were eight at 1 minute and nine at 5 minutes. He was discharged home on the 1st day of life with emollient, moisturizing eye drops and gel.

The image shows areas of severe dry itchiness across the skin. Note bilateral ectropion. On presentation to the outpatient department, the findings were similar to those noted at delivery, but there was slight improvement in skin proliferation. Notably, we noted a worsening of lid ectropion. The child could not close his eyes, while there was no erosion of the cornea.

Emergency ectropion release and full-thickness skin grafting were performed. After ectropion was removed, a full-thickness skin biopsy of the right medial arm was obtained under general anesthesia on day 23 of life. The flaps were then partially closed using Frost sutures over rubberized tubing from a sterile urinary catheter. Postoperatively, the patient did well and had complete correction of bilateral ectropion.

Ichthyosis represents a rare group of diseases characterized by varying degrees of cornification. These disorders are thought to be caused by mutations in the ABCA12 gene in an autosomal recessive pattern. Mutation of this gene causes defective lipid deposition in the stratum carenum of the epidermis, which manifests clinically as extreme hyperkeratosis. The most common form of ichthyosis is ichthyosis vulgaris, followed by X-linked ichthyosis. Ichthyosis vulgaris occurs in 1/300 live births.

Traditionally, these patients had a high mortality rate, but with improved care and advances in medicine, these patients are living longer and leading "normal" lives. A recent review by Rajpopat et al. Harlequin showed a total of 45 cases of ichthyosis, with an overall survival rate of 56%. They found that death occurred within the first 3 months and that 75% of cases were related to sepsis and/or respiratory failure.

Harlequin ichthyosis is a subtype of congenital ichthyosis (CI). This is a very rare but severe form of CI. It is characterized by thickened scaly plates, usually characterized by deep cracks. These plaques can cover the penis or the entire body from head to toe with skin folds and folds. Less commonly, these conditions may involve the entire body without sparing, as seen in our case.

Conclusion. In this article using the same basic principles as the management of burn injury ectropion are analyzed. Early intervention has been shown to produce excellent results and alleviate corneal abrasion. Relaxing incisions were made up to the orbicularis oculi. This provided an ideal recipient bed for a thin full-thickness graft from the medial arm. Frost stitches then allowed excellent grafting and softened against subsequent ectropion as the wound healed.

Ichthyosis represents a group of rare diseases characterized by thickening of the epidermis of the skin. The patient may present with rare variants, including thickened scales that can deform the limbs. In addition, variants such as lamellar subtypes usually present with severe ectropion. The multidisciplinary approach of dermatologist, ophthalmologist, ENT and plastic surgeons provides timely intervention, which allows to save limbs and eyes.

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