CLINICAL INVESTIGATION

Erythrokeratoderma: Keratinization disorder

Erythrokeratoderma is a group of rare genetic skin diseases marked by reddish, dry, and thicker skin plaques. In erythrokeratoderma, there are two types of lesions: erythematous transitory patches and hyperkeratotic permanent plaques. There are no specific treatments or suggestions for Erythrokeratoderma. Erythrokeratoderma affects both men and women in about equal percentages. The disease's prevalence in the general population is unknown. The retinoids acitretin, etretinate, and isotretinoin, which are taken orally, have been demonstrated to be quite effective. Lesions return after therapy is stopped. Patients should be given a prognosis and be followed up on on a regular basis.

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Introduction

Erythrokeratoderma is a name used to describe a group of rare hereditary skin disorders characterised by well-defined reddish, dry, and thicker skin plaques. These lesions are usually symmetrically distributed throughout the body and gradually expand and grow over time. Furthermore, within the same family, the severity and progression of the disease might differ dramatically from one person to the next. Progressive Symmetrical Erythrokeratoderma (PSEK) and Erythokeratodermia Variabilis et Progressiva are the two most prevalent kinds of erythrokeratoderma (EKVP).

Causes

Erythrokeratoderma is linked to a variety of genetically transmitted illnesses, most of which are autosomal dominant. This shows that the gene comes from a single parent and that a person with the condition can pass it on to 50% of his or her offspring. Sporadic occurrences emerge as a result of novel genetic alterations at conception. The underlying problem in most erythrokeratoderma forms is usually a mutation in one of the connexin genes. Gap junction proteins are connexins, which are found in the channels that connect nearby cells. Different connexins are found in different tissues, allowing for appearance variety. According to a recent study, erythrokeratoderma can have a variety of causes and can be a symptom of a variety of inherited skin conditions. The following are the different types of erythrokeratoderma depending on hereditary variables.

- Autosomal dominant erythrokeratoderma
- Autosomal recessive erythrokeratoderma

Not all people with erythrokeratoderma have variations in the causative sequence of the epidermal gene discussed above, and more research is needed to identify and validate the specific genetic mutations that cause erythrokeratoderma, as well as the precise underlying mechanisms that lead to the disorder's onset.

Diagnosis

A diagnosis of erythrokeratoderma is obtained based on the detection of typical indicators, a full case history, a thorough clinical examination, and advanced exams, such as genetic testing or surgical removal (biopsy) and microscopic inspection of infected tissue. Histopathological examination only detects non-specific findings such as papillomatosis, mild to severe acanthosis, hypergranulosis with two to four layers of cells, compact hyperkeratosis or parakeratosis, and follicular plugging.

The papillary dermis contains dilated, elongated capillaries with a complicated inflammatory perivascular infiltration. Suprapapillary thinning is a potential presenting sign, and when paired with high papilloma-tosis, it can result with an epidermal "church spire" shape. Ultrastructural tests reveal a decrease in the number of lamellar bodies in the granular layer.

Treatment

There are no specific treatments or suggestions for Erythrokeratoderma. The different topical drugs used include emollients, retinoic acid, keratolytics, tazarotene, alpha hydroxyl acid, and topical corticosteroids. The retinoids acitretin, etretinate, and isotretinoin, which are taken orally, have been demonstrated to be quite effective. Lesions return after therapy is stopped. Patients should be given a prognosis and be followed up on on a regular basis.