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Commentary

Commentary on Erythrokeratoderma

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ABSTRACT

Erythrokeratoderma is a category of unusual hereditary skin conditions characterized by reddened, dried and thickened skin plaques. Two characteristic forms of lesions exist in erythrokeratoderma: Erythematous transient patches and Hyperkeratotic stable plaques. No clear therapies or recommendations for Erythrokeratoderma are available. Males and women are affected in similar numbers by Erythrokeratoderma. The occurrence in the general populace of the disease is uncertain. The oral retinoids acitretin, etretinate, and isotretinoin were shown to be very effective. When therapy is discontinued, lesions reappear. Prognosis and regular follow-up can be recommended for patients.

Keywords: Erthroketoderma; Skin condition; Hereditary

INTRODUCTION

Erythrokeratoderma is a term used for a category of unusual hereditary skin conditions characterized by reddened, dried and thickened skin plaques that are well demarcated. Typically, these lesions are distributed symmetrically on the body and tend to slowly expand and progress over time. Also, within members of the same family, the severity and development of the condition can vary significantly from one person to another. The most common types of erythrokeratoderma are Progressive Symmetrical Erythrokeratoderma (PSEK) and Erythokeratodermia Variabilis et Progressiva (EKVP).

CAUSES

Erythrokeratoderma is attributed to many, often autosomal dominant, genetically transmitted diseases. This suggests that the gene comes from one parent and can be passed on to 50 percent of his or her offspring by a person with the disease. Owing to new genetic mutations at conception, sporadic cases occur. In most erythrokeratoderma forms, a mutation in one of the connexin genes tends to be the underlying defect. Connexins, located in the channels binding neighboring cells, are gap junction proteins. In different tissues, different connexins are detected, allowing for appearance heterogeneity. Latest study has demonstrated that erythrokeratoderma can have multiple causes and can be a characteristic of a number of inherited skin disorders. Erythrokeratoderma forms have been described based on hereditary factors as follows.

1. Autosomal dominant erythrokeratoderma
2. Autosomal recessive erythrokeratoderma

Not all individuals with erythrokeratoderma characteristics have variations in the causative sequence of the epidermal gene discussed above, and more study is required to define and validate the particular genetic mutations that cause erythrokeratoderma and to establish the precise underlying mechanisms involved in the disorder's development.

Males and women are affected in similar numbers by Erythrokeratoderma. The occurrence in the general populace of the disease is uncertain. In 1911, the condition was first identified by Darier. More than 100 cases have been identified in the medical literature since then. In Middle Eastern populations with a higher degree of marriage between blood-relatives, autosomal recessive inherited variants of erythrokeratoderma have been reported more commonly (consanguinity). Two characteristic forms of lesions exist in erythrokeratoderma: Erythematous transient patches, Hyperkeratotic stable plaques.

DIAGNOSIS

Based on the recognition of characteristic signs, a detailed case history, a rigorous clinical examination and advanced examinations, including genetic testing or surgical removal (biopsy) and microscopic evaluation of infected tissue, a diagnosis of erythrokeratoderma is made. Only non-specific observations, including papillomatosis, mild to extreme acanthosis, hypergranulosis consisting of two layers to four layers cells,

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compact hyperkeratosis or parakeratosis and follicular plugging, are seen by histopathological review. Dilated, elongated capillaries with a complex inflammatory perivascular infiltrate are found in the papillary dermis. A potential presenting symptom is an aspect of suprapapillary thinning, and when it is combined with extreme papilloma-tosis, this can manifest in an epidermis "church spire" configuration. A drop in the number of lamellar bodies in the granular layer is seen by ultrastructural tests.

TREATMENT

No clear therapies or recommendations for Erythrokeratoderma are available. Emollients, retinoic acid, keratolytics, tazarotene, alpha hydroxyl acid, and topical corticosteroids are the various topical medicines utilized. The oral retinoids acitretin, etretinate, and isotretinoin were shown to be very effective. When therapy is discontinued, lesions reappear. Prognosis and regular follow-up can be recommended for patients.

REFERENCES

1. https://rarediseases.org/rare-diseases/https-rarediseases-org-rare-diseases-erythrokeratoderma/#:~:text=Erythrokeratoderma%20is%20an%20umbrella%20term,expand%20and%20progress%20over%20time.
2. https://dermnetnz.org/topics/erythrokeratoderma/
3. https://emedicine.medscape.com/article/1110820-overview
4. https://www.ijpd.in/article.asp?issn=2319-7250;year=2016;volume=17;issue=3;spage=202;epage=205;aulast=Srinivas
5. https://www.dermatologyadvisor.com/home/decision-support-in-medicine/dermatology/erythrokeratodermia-variabilis-erythrokeratodermia-figurate-variabilis-erythrokeratoderma-congenitals-progressiva-symmetrica-erythrokeratoderma-progressive/
6. https://www.ijpd.in/article.asp?issn=2319-7250;year=2016;volume=17;issue=4;spage=309;epage=311;aulast=Patro
7. https://www.idoj.in/article.asp?issn=2229-5178;year=2019;volume=10;issue=2;spage=183;epage=186;aulast=Tiwary