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Erythrokeratoderma

What is erythrokeratoderma?

Erythrokeratoderma, sometimes called erythrokeratodermia, is the descriptive name given to a rare group of disorders of keratinisation. This is the process that forms the different layers of the epidermis, the outermost layer of the skin. The various erythrokeratodermas are characterised by well demarcated plaques of erythema (redness) and hyperkeratosis (scaling).

Erythrokeratoderma



How do you get erythrokeratoderma?

Erythrokeratoderma is due to several genetically inherited disorders, mostly autosomal dominant. This means that the gene comes from one parent, and that an individual with the disease may pass it on to 50% of his or her children. But sporadic cases do occur due to new genetic mutations occurring at conception.

In most types of erythrokeratoderma, the underlying defect appears to be a mutation in one of the connexin genes. Connexins are gap junction proteins, found in the channels that connect adjacent cells. Different connexins are found in different tissues, accounting for variability in presentation.

How is it diagnosed?

Erythrokeratoderma is diagnosed by its clinical appearance. [Skin biopsy](#) can be performed for histology but there are no distinctive features.

Is there any treatment available?

Genetic counselling should be offered to affected individuals and their families of childbearing age. In time, genetic tests for the specific disorders may be available to some families.

There is no specific or curative treatment. Minimizing temperature changes and mechanical friction is important. Symptomatic improvement can be obtained by:

- [Emollients](#)
- Keratolytics such as [urea](#), [salicylic acid](#) or [alpha hydroxy acids](#)
- [Topical steroids](#)
- [Topical retinoids](#)
- Oral retinoids such as [acitretin](#) or [isotretinoin](#). These thin down the plaques and reduce scaling but the redness persists. It can be difficult to decide whether to take them longterm for erythrokeratoderma, because they may cause some adverse effects.

Specific types of erythrokeratoderma

Because erythrokeratoderma is rare, the classification of the different types is still evolving. But there are few well defined syndromes and some other atypical variants which are listed below.

- [Erythrokeratoderma variabilis](#) (Mendes da Costa's syndrome)
- [Erythrokeratoderma progressiva symmetrica](#) (Gottron's syndrome)
- [Progressive partially symmetrical erythrokeratoderma](#) with peripheral neuropathy and deafness
- Erythrokeratoderma en cocardes (Degos syndrome)
- Erythrokeratoderma with ataxia
- Annular migrating erythrokeratoderma
- Erythrokeratoderma-like lesions in KID syndrome (Keratitis, [Ichthyosis](#), and Deafness)
- Erythrokeratoderma with periorificial lesions
- Localised erythrokeratoderma

Erythrokeratoderma variabilis

This is the commonest of the erythrokeratodermas and is autosomal dominantly inherited or sporadic. More than 50% of the affected individuals show skin lesions at birth or during the neonatal period and 90% will show some evidence of the disease within their first year of life.

Erythrokeratoderma variabilis results in well-defined round or oval red scaly plaques (thick patches) that may join together to form map-like patterns. There are 2 types of skin lesion:

- Fixed plaques, usually occurring on the outer surfaces of the arms and legs
- Migratory plaques that can occur at any site, last for hours to days and either fade or move to other areas.

In severe cases the erythrokeratoderma can be generalised.

Some patients experience burning sensations and itching in the affected areas and others have no symptoms. The skin lesions may be triggered by internal and/or external factors, such as:

- Emotional stress
- Temperature changes
- Mechanical friction
- Hot or cold weather.

The hair, teeth and nails are not involved. Erythrokeratoderma does not affect physical and mental development and the general health of the individual is not affected.

After gradual progression throughout infancy and childhood, the disease tends to stabilise during puberty.

Erythrokeratoderma progressiva symmetrica

Erythrokeratoderma progressiva symmetrica is a very rare form of erythrokeratoderma that is also genetically inherited. Sporadic cases may occur.

Skin lesions are not present at birth and begin during infancy or early childhood. Affected individuals show fixed or slowly progressive red, scaly plaques symmetrically distributed on the body. The extremities, i.e., hands and feet, are often involved, which is uncommon in erythrokeratoderma variabilis.

Physical and mental development are normal.

Like erythrokeratoderma variabilis, the skin lesions of erythrokeratoderma progressiva symmetrica slowly progress and increase in number and size during childhood, and tend to stabilise after puberty. The inherited cases have symptoms lifelong but there are reports of spontaneous improvement after many years in the sporadic cases.

Other types of erythrokeratoderma

Other types of erythrokeratoderma are very rare and only few cases have been reported worldwide. Many of these had only skin lesions and were otherwise well.

Progressive partially symmetrical erythrokeratoderma results in deafness, muscle weakness, peripheral nerve damage, physical and mental retardation in addition to atypical peripheral erythrokeratoderma.

Related information

References:

- Rogers M. Erythrokeratodermas: A classification in a state of flux? Australasian Journal of Dermatology 2005 Aug;46(3):127-41. [Medline](#).

On DermNet NZ:

- [Scaly skin conditions](#)
- [Palmoplantar keratoderma](#)

Other websites:

- [Erythrokeratoderma Variabilis](#) – emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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