Palmoplantar keratoderma

What is it?

“Palmoplantar” means palms of the hands and soles of the feet, and “keratoderma” means thickened skin. So palmoplantar keratoderma is a disorder in which the skin of the palms and soles is abnormally thick. It is rare, and it often runs in families. There are different types, but all members of the same family will have the same type.

What does it look like?

The palms and soles may look normal at birth, or slightly whitish. Later the affected skin becomes thickened. Sometimes it first appears when the child starts to walk, and most types become more apparent with friction and with age. The affected skin may be red and flaky, or hard and yellowish. In some types the keratoderma affects the whole surface of the palms and soles, in others it is streaky or patchy like corns (calluses). Sometimes it extends round the sides to the backs of the fingers and tops of the toes – this is called a “trangrediens” pattern.

What problems does it cause?

Palmoplantar keratoderma can cause difficulty with walking, because of pain in the feet. The thick skin and sweating of the feet makes them particularly susceptible to odour, and to fungal infection (athlete’s foot). The thick skin on the palms may reduce sensitivity in the finger tips, impairing manual dexterity. All these problems, together with the unusual appearance can be stressful and lead to psychological difficulties.
Is palmoplantar keratoderma associated with other medical problems?

The vast majority of people with palmoplantar keratoderma are otherwise completely healthy. But some rare types of palmoplantar keratoderma are associated with other skin problems such as dry skin all over (“ichthyosis”), problems with the nails, hair or teeth, deafness, or other medical problems.

What causes palmoplantar keratoderma?

The palmoplantar keratodermas are caused by abnormal proteins in the superficial layer of skin (epidermis). We have several genes responsible for producing these proteins. A mistake (mutation) in any one of these genes causes production of a faulty protein, and this in turn affects the structure of the skin of the palms and soles. Most cases of palmoplantar keratoderma are caused by mutations in the gene that produces an epidermal protein called keratin 9. But there are several other genes and mutations responsible for the rarer types of palmoplantar keratoderma.

Where does the mutation come from?

Many patients with palmoplantar keratoderma inherit the condition from their father or mother. This is called “autosomal dominant inheritance”. They in turn are likely to pass it on to half of their children.

Some people with palmoplantar keratoderma cannot trace it back to their father or mother, but have an affected brother or sister, or other more distant relative. This is called “autosomal recessive inheritance”. Both their parents are “carriers” of the mutation but do not show the condition. People with autosomal recessive PPK do not pass it on to their children unless they marry another person carrying the same mutation, because the faulty gene has to come from both parents.

About half of people with palmoplantar keratoderma have no family history of the condition at all. In these cases the mutation has happened during their embryonic development. We do not know why it happens, and once it has appeared it can be passed on to future generations.

How is palmoplantar keratoderma treated?

At present there is no cure for palmoplantar keratoderma. The skin cells are programmed to make a faulty protein and this cannot be corrected. But the symptoms can be improved by the following measures:

- Regular use of moisturising creams and ointments
- The use of simple antiseptics if odour is a problem
- Antifungal cream or tablets if fungal infection is present
- Regular chiropody to pare down the thick skin
- Customised insoles to relieve pressure-points on the feet

In severe cases a dermatologist may prescribe the drug acitretin. This thins the affected skin. Treatment has to be closely monitored to avoid excessive thinning of the skin and various side-effects of the drug. It is a drug that must never be taken during pregnancy because it can damage the baby.

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