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ABSTRACT BOOK

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FATHER AND DAUGHTER WITH THICKENED PALMS AND SOLES IN PRIMARY CARE

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Introduction: Palmoplantar keratoderma (PPK) is characterised by excessive thickening of the palms and soles. Hereditary PPK is rare with prevalence in Asia estimated at 1 to 3 per 10,000. A careful assessment is needed due to this group of diseases' considerable clinical and genetic variation. **Case report:** We report a 3-year and 6-month-old girl who presented with skin thickening for two years. She has no extracutaneous symptoms. Her father and elder brother have similar dermatological manifestations. On examination, the child appears well, with no syndromic features. Her skin over both palms and soles is dry, coarse, hyperkeratotic, with trans gradient area involving web spaces of feet. There is no pseudo ainhum. The diagnosis of PPK is based on clinical findings. Laboratory investigations are not done. She was prescribed urea 10% LA cream BD and Vaseline as emollient LA QID. The skin thickening subsequently reduced within months. **Conclusion:** Hereditary PPK is a challenging condition to diagnose because it is uncommon, with a variety of presentations and classifications. More importantly, PPK can be associated with many other conditions and could be a feature of other syndromes, which the unknowing physician could miss. A thorough history and careful physical examination are important since patients with syndromic PPK will require a multidisciplinary approach as part of its management. The patient and her parents may be anxious about how the disease progresses. Ensuring life-long compliance with the medication is crucial as recurrence is common after discontinuing treatment.

Keywords: Palmoplantar keratoderma, Inherited, Keratoderma

Father and daughter with thickened palms and soles in primary care

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ABSTRACT

Palmoplantar keratoderma (PPK) is characterised by excessive thickening of the palms and soles. Hereditary PPK is rare with prevalence in Asia estimated at 1 to 3 per 10,000. A careful assessment is needed due to this group of diseases' considerable clinical and genetic variation. We report a 3-year and 6-month-old girl who presented with skin thickening for two years with her father and elder brother having similar dermatological manifestations. On examination, the child appears well, with no syndromic features. Her skin over both palms and soles is dry, coarse, hyperkeratotic, and trans gradient involving web spaces of feet. The diagnosis of PPK is based on clinical findings. Laboratory investigations are not done. She was prescribed urea 10% LA cream BD and Vaseline as emollient LA QID. The child remained well and active throughout the treatment. The skin thickening subsequently reduced within a couple of months.

INTRODUCTION

- Palmoplantar keratoderma (PPK) is one of the rarer dermatological conditions.
- Diagnosing PPK is challenging and requires a careful assessment.
- There are multiple classifications of PPK, based on the presence of other cutaneous or extracutaneous symptoms or genetic defects.
- A thorough history and careful physical examination can help distinguish between non-syndromic and syndromic conditions characterized by PPK and extracutaneous involvement.

CASE REPORT

- A 3-year and 6-month-old girl presented with skin thickening for two years, with her father and elder brother having similar dermatological manifestation.
- On examination, the child appears well, with no syndromic features. Her skin over both palms and soles is dry, coarse, hyperkeratotic, and trans gradient involving web spaces of feet.
- The diagnosis of PPK is based on clinical findings.
- With urea 10% LA cream BD and Vaseline as emollient LA QID, the skin thickening subsequently reduced within a couple of months.



CHALLENGES IN DIAGNOSIS

The diagnosis of a specific PPK type is challenging, due to its highly heterogenous phenotypes and genotypes. Ideally, diagnosis should be based on a combination of clinical, histopathologic features and genetic testing.

RECOMMENDATION FOR PRACTICE

- The diagnosis of PPK in practice is based on clinical findings.
- Presence of deafness, ichthyosis, periodontitis, cardiomyopathy and woolly hair, ectodermal dysplasia, PLACK (peeling skin with leukonychia, acral punctate keratoses, cheilitis and knuckle pads), guttate hypopigmentation and oesophageal cancer and squamous cell carcinoma may suggest syndromic PPK.
- Laboratory investigations such as tissue biopsy for histopathology examination or genetic testing for gene mutation detection help confirm the diagnosis however it is rarely done.
- The use of pharmacology therapies such as topical keratolytic, topical retinoid and oral retinoid are accepted.

IMPLICATIONS

- The detection of hereditary PPK may cause a patient and her parents to be concerned about how the disease progresses.
- Ensuring life-long compliance with the medication requires a good relationship between the patient and the doctor. As none of these treatments offer lasting effects, recurrence is common after discontinuing treatment.
- The potential risks have been minimised through early referral to a tertiary care facility in this case.

CONCLUSION

- A thorough history and careful physical examination have a propensity to reduce the gap for misdiagnosing or underdiagnosed of PPK.
- Adopting this recommendation can contribute to diagnose patients with syndromic PPK as they will require a multidisciplinary approach as part of its management.

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