

Striate Palmoplantar Keratoderma in a Nigerian Child: A Rare Disorder

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ABSTRACT

Hereditary focal palmoplantar keratodermas (PPK) are uncommon disorders occasionally seen by the dermatologist. Striate PPK is even more uncommon and may or may not be associated with organ abnormalities. An isolated case of striate PPK in a child with morbidity related to ambulation is reported here.

INTRODUCTION

Palmoplantar keratoderma (PPK) is a general term for focal or diffuse thickening of the palms and soles due to hyperkeratosis. It could be acquired or inherited. Hereditary focal PPK is a heterogenous group of disorders characterised by abnormal keratinisation. It could present with clinical features that are limited to the skin or manifest as part of a more generalized disorder.

CASE REPORT

A 13 year old girl presented to the Dermatology clinic with irregular thickening of the soles of the feet appearing since the first year of life. It was associated

with pain and resultant difficulty in walking. There was progressive thickening of the lesions over the years with recurrent fissuring and bleeding. Thickening on the palms were noticed by the age of 3 years, appearing in a distinct manner which were painful especially when using the hands for manual work like washing. There was associated itching with foul smelling discharge and hyperhidrosis. There were no other systemic symptoms. No abnormalities of the teeth, hair and nails. There was no history to suggest cardiac or oesophageal disease, neither was there any complaint of hearing loss. Her pregnancy, birth and neonatal history were essentially normal, she had no developmental delays and there was no family history of similar problems. Prior to presentation, she had used



Figure 1: Linear hyperkeratotic plaques on the volar surface of fingers and palms



Figure 2a: Hyperkeratosis of the sole with lack of transgrediens.



Figure 2b: Hyperkeratosis of the soles bilaterally ▶

various creams, drugs and herbal preparations.

Examination of the skin revealed hyperkeratotic plaques arranged in a linear pattern on the volar aspect of each of the 5 fingers of both hands. There were linear papules extending from the first and second digits to the palms and flexor aspect of the wrists. There was no transgrediens. There were fissures in between the plaques with hyperhidrosis of the hands (Fig 1). Finger nails had a yellowish hue with transverse lines and hyperkeratosis. Feet examination showed bilateral hyperkeratosis sparing the instep of the sole and loss of transgrediens (Fig 2). Subungual hyperkeratosis of nine toenails were seen. There was also nail dystrophy with destruction of the nail plate of the big and 2nd toes bilaterally. Hyperhidrosis of the dorsum of the feet was also seen. Other systems were essentially normal and a diagnosis of striate palmoplantar keratoderma was made. Full blood count was normal and patient was counselled on the need for a skin grafting. She has been scheduled for excision of the lesions and skin grafting as soon as funds are available.

DISCUSSION

The hereditary PPKs are a rare group of disorders with a wide spectrum of clinical severity. The lesions could be present in either a diffuse, focal or punctuate pattern. In the diffuse forms, hyperkeratosis is usually in a uniform pattern over the palmoplantar surface. While the focal forms present as localised lesions typically on weight bearing areas. Each of these forms can be as isolated dermatoses in which abnormalities are limited to the volar skin or in association with other systemic or organ abnormalities.

Striate PPK, also known as Brunauer-Fuhs-Siemens syndrome or Wachter-type focal PPK, is a rare focal palmoplantar keratoderma, with an autosomal dominant pattern of inheritance. It is characterised by linear hyperkeratosis on the digits and palms and focal hyperkeratosis of the soles. Pathogenesis has been linked to mutations in the genes for desmoglein 1 (18q11-12) and desmoplakin (6p21) - which are components of desmosomes and keratin 1 and 16. Phenotypic expression is variable and onset is usually early in childhood before the age of 2 years. Hyperkeratosis typically occurs on the pressure bearing areas and lesions on the soles are usually more diffuse than the palms. Lack of spillage of lesions to the dorsum of hands and feet with a sharp demarcation known as transgrediens, a common characteristic of the disorder was manifest in our patient. Hyperkeratosis in the palms is usually milder than the soles and is often exacerbated by manual labour. Hyperkeratotic plaques also may be observed on the elbows and knees. There are usually no other associated clinical features as seen in this patient, however some forms of focal PPK have

been associated with oesophageal cancer (Howell Evans syndrome), cardiomyopathy (Carvajal-Huerta syndrome) and sensorineural hearing loss. This patient did not have symptoms attributable to any organ and was managed as an isolated disease. The significant morbidity in the isolated forms arises from pain which may prevent patients from walking. They are also prone to excessive sweating of the palms and soles and secondary fungal infections.

Various forms of treatment like the topical keratolytics, topical and oral retinoids have been used with moderate success. Surgical debridement and excisions have been used as a last resort to reduce pain and improve ambulation and this is what we planned for our patient

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