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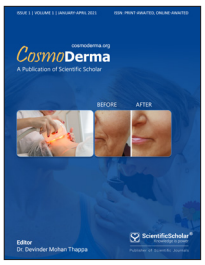

Letter to the Editor

Palmoplantar keratoderma and woolly hair

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Received : 12 September 2022
Accepted : 28 September 2022
Published : 12 October 2022

DOI
10.25259/CSDM_98_2022

Quick Response Code:

Dear Sir,

We report a case of a 23-year-old male who presented with palmoplantar keratoderma (PPK) making it difficult for him to walk and perform his daily activities due to stiffness of skin of palms with associated pitted keratolysis present on bilateral soles [Figures 1 and 2]. On close observation, coarse curly hair was noticed over the scalp [Figure 3a and b]. The patient was accompanied by his brother whose hair texture differed from that of patient but their sister reportedly had curly coarse hair. There were no complaints in his parents. On hair light microscopy, hair shafts were flat and irregular [Figure 4]. The patient was further investigated and ECG changes were consistent with fascicular ventricular tachycardia with the right bundle branch block pattern and 2D echocardiography showed right ventricular cardiomyopathy. Routine investigations were normal. The patient was counseled and symptomatically managed for PPK.

PPK is a heterogeneous group of genodermatosis, with hyperkeratosis of palms and soles. They are classified according to either their morphology (diffuse, focal, and punctate), mode of inheritance (autosomal dominant and autosomal recessive), and presence or absence of extracutaneous features. The classical clinical presentation of PPK, right ventricular cardiomyopathy, and woolly hair establishes the diagnosis of diffuse inherited PPK with extracutaneous features, most likely of Naxos disease, caused by mutation in plakoglobin gene. Therefore, whenever woolly hair presented with associated PPK, search for possible cardiac abnormalities is recommended, and regular follow-up is required.[1] Naxos disease is a rare genodermatosis with woolly hair, PPK, and

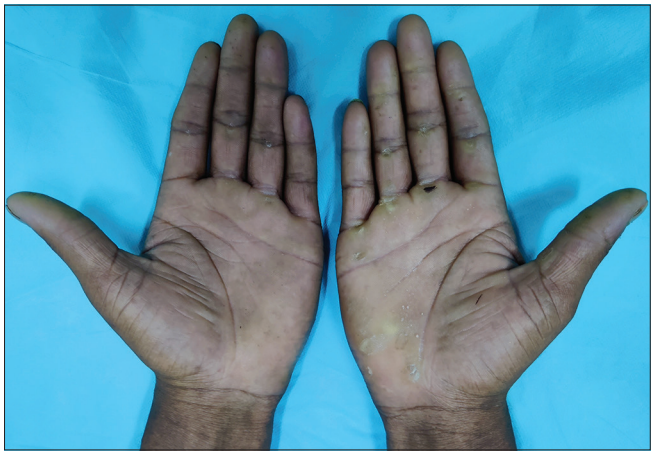

Figure 1: Palmar keratoderma with callosities.

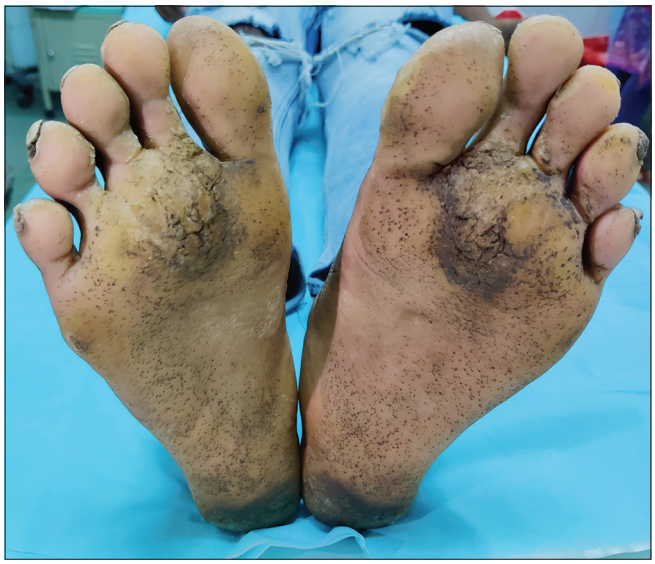

Figure 2: Plantar keratoderma with pitted keratolysis.

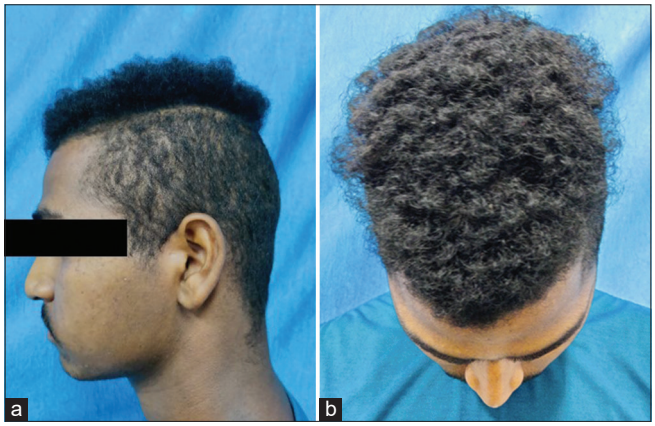

Figure 3: (a and b) Curly coarse hairs suggestive of woolly hair (lateral view).

cardiomyopathy.[2] In India, there are very few case reports on Naxos disease, which comes in view when the patient comes with recurrent syncope or rhythm abnormalities, so regular monitoring is required.

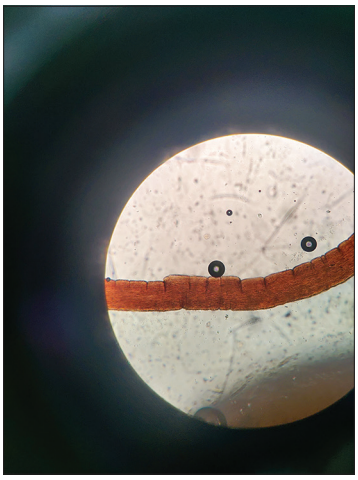

Figure 4: Hair microscopy (40 ×) showing flat and irregular hair shaft.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Kalyana T, Arora A, Thami GP. Palmoplantar keratoderma and woolly hair. CosmoDerma 2022;2:91.