Original Research Paper

Volume - 11 | Issue - 10 | October - 2021 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Dermatology

A RARE CASE REPORT OF FAMILIAL SPECKLED ACRAL HYPOMELANOSIS

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ABSTRACT Familial speckled acral hypomelanosis is a rare entity with only few cases been reported so far. It falls under the category of acral speckled or mottled pigmentary disorders. The other disorders in this group are reticulate acropigmentation of Kitamura, acropigmentation of Dohi and acromelanosis albo- punctata which have a significant overlap in their features and often pose diagnostic challenge, as these disorders are variable phenotypic expression of similar gene defects. It presents as asymptomatic, speckled hypopigmented macules distributed symmetrically over the dorsa of hands and feet with prominent clustering of macules over the sides, with a similar family history. The patient's general health remains normal with no systemic involvement. There were no palmar pits or break in dermatoglyphics or hyperpigmented macules. There is no accentuation under Wood's lamp examination. Decreased or normal number of melanocytes with a striking number of macromelanosomes in melanocytes and keratinocytes are present in histopathological examination without any other remarkable changes in the epidermis or dermis.

KEYWORDS : acral, speckled, reticulate, hypomelanosis

INTRODUCTION:

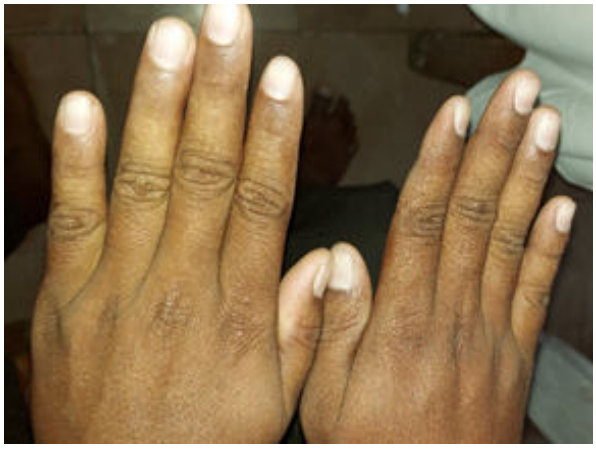
A wide range of skin conditions present with hypopigmentation, where morphology and distribution of lesions help in differentiating these conditions. Conditions which present with speckled or mottled hypopigmented macules in the acral areas are Familial Speckled Acral Hypomelanosis (FSAH), Reticulate acropigmentation of Kitamura (RAPK), Acropigmentation of Dohi and acromelanosis albo- punctata which pose a difficulty in diagnosis due to variable phenotypic expression of similar gene defects. Hereby, we report a case of FSAH in an otherwise healthy 25 year old male.

Case Report:

A 25-year-old man presented with asymptomatic small white spots in hands and feet since 15 years, which started as few spots on the dorsa of hands which then gradually increased in number to involve the dorsa of feet and palms and soles. There was family history of similar lesions in his father, who was not available for examination. There was no history of any preceding dermatoses, chemical exposure, drug intake or systemic illness.

On cutaneous examination, multiple, well defined, 1-2 mm discrete, speckled or guttate hypopigmented macules were present symmetrically in the dorsa of hands and feet and in palms and soles. The lesions were clustered more over the sides of dorsa of hands and feet. Other sites were uninvolved. Hair, nail and mucous membrane examination was normal. General and systemic examination of the patient was within normal limits.

Figure 1:



Multiple, discrete, speckled hypopigmented macules on the dorsa of hands.

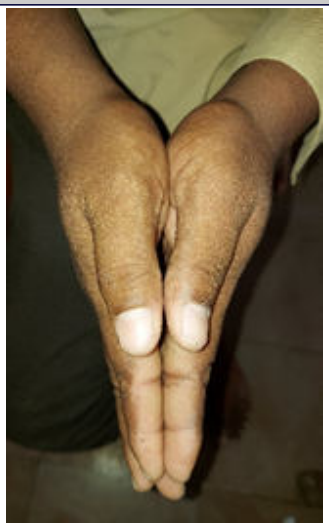
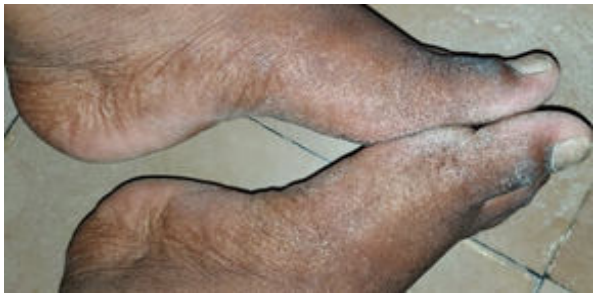


Figure 2: Clustering of speckled hypomelanotic macules on the sides of dorsa of hands.

Figure 3:



Multiple guttate hypomelatic macules over sides of dorsa of feet.

Under Wood's lamp examination, there were no accentuation of skin lesions. A skin biopsy was offered, but was refused by the patient. A diagnosis of Familial Speckled Acral Hypomelanosis was made based on the clinical findings.

DISCUSSION:

Familial Speckled Acral Hypomelanosis (FSAH) is first described in 2005 in West Bengal by Malakar et al. (Malakar et al., 2005) It presents with speckled hypopigmented macules on the sides of dorsa of hands and feet.

FSAH presents as multiple, asymptomatic speckled or guttate or confetti hypopigmented macules without atrophy in acral areas, with clustering of macules over the sides of dorsa of hands and feet. (*Acral Speckled Hypomelanosis - JAAD Case Reports*, n.d.) The word familial indicates the presence of strong family history of similar lesions.

Wood's lamp examination doesnot show any accentuation. Histopathological examination shows normal or reduced number of melanocytes in contrast to guttate vitiligo which shows complete absence of melanocytes. There is striking number of macromelanosomes in melanocytes and keratinocytes. (Singh et al., 2019). General health of the patient remains unaffected.

Reticulate acropigmentation of Kitamura (RAPK) can be differentiated from FASH by the presence of atrophic hyperpigmented macules over dorsa of hands and feet with palmar pits and break in dermatoglyphics. Histopathology shows hyperkeratosis with thinning and elongatation of rete ridges and increased melanin pigment in the basal layer of epidermis. Dowling Degos disease (DDD) is its close histopathological differential diagnosis which can be differentiated by the presence of antler- like branching of elongated rete ridges. (Rathoriya et al., 2016)

Acropigmentation of Dohi also called as Dyschromatosis Symmetrica Hereditaria (DSH) has both hypo and hyperpigmented macules on hands and feet which can extend to proximal extremities and face. (Sinha, n.d.)

Acromelanosis albo-punctata has generalized hyperpigmentation of skin with confetti-like hypopigmented macules over hands and feet which are also found in flexural areas to a lesser extent. Congenital symmetric acroleukopathy has large depigmented macules over peri-ungual areas.(Singh et al., 2019)

CONCLUSION:

We hereby report this case because of its rarity which presents with speckled hypopigmented macules over acral areas, which donot fit into other well defined reticulate acropigmentary disorders. It may be a new disorder or an unknown presentation of a previously defined disorder. A study of large number of cases is needed to solve this mystery.

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