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RESEARCH ARTICLE

"WOOLY HAIR PALMOPLANTAR KERATODERMA : A CASE REPORT"

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Abstract

Wooly hair is a rare hair shaft abnormality characterized by extremely curly hair. It can occur in isolation or associated with palmoplantar keratoderma (PPK) and cardiac defects. We present the case of a 56-year-old female patient who initially presented with non-itchy, painful skin lesions over palms and soles since 5years and had abnormal scalp hair texture since childhood. She responded positively to treatment with topical application of 6%salicylic acid twice a day for 3 months on which the thickness and size of the plaques gradually reduced.

Introduction:-

WOOLY HAIR PALMOPLANTAR KERATODERMA syndrome encompasses a rare group of autosomal recessive diseases wherein hair and skin abnormalities are usually associated with potentially fatal cardiac complications. The four clinical syndromes described. Types 1-3 are associated with mutations of desmosomal proteins and present life-threatening cardiac abnormalities while type 4 occurs due to mutation in steroid receptor coactivator (SRC)-interacting protien (non-desmosomal) presenting without cardiac involvement.[1]

Case Report:

A 56-year-old female resident of Nellore presented with onset of non-itchy, painful skin lesions and abnormal scalp hair texture was observed which has been present since childhood. There was no history of similar skin and hair changes among siblings or other family members, no history of delayed milestones and mental retardation was elicited, no history of syncopal attacks, palpitations in the patient.

On Examination:

Multiple yellowish thick and hyperkeratotic discrete plaques with smallest being 1*2cm and largest being 5*6cm which were firm to hard in consistency with verrucous surface noted over bilateral palms and soles. Wooly textured hair noted over scalp.

The patient's general examination including oral cavity, nails, genitalia were normal, no pallor, cyanosis, icterus, lymphadenopathy, pedal edema. Systemic examination was unremarkable.

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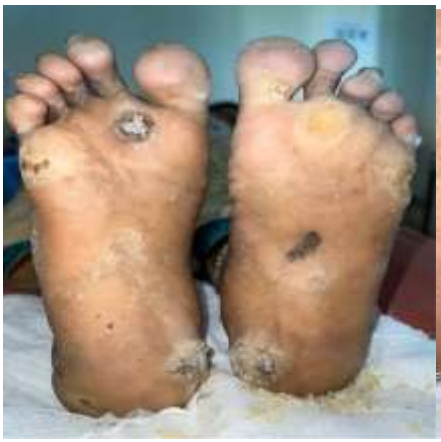
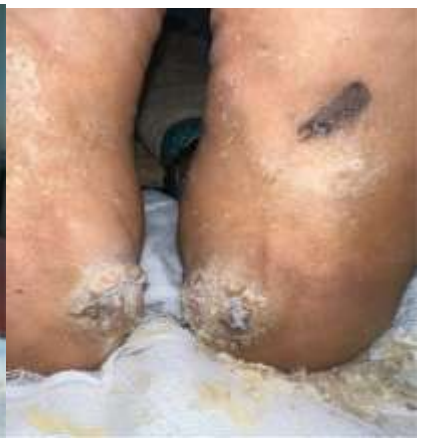


(Fig.1)



(Fig.2)

(Fig.3)

(Fig.4)

(Fig.5)

Multiple yellowish thick and hyperkeratotic plaques with verrucous surface noted over soles (Fig.1),(Fig.2)and palms (Fig.3), wooly hair over scalp(Fig.4),(Fig.5) noted.

Investigations:

Routine blood investigations were within normal limits, ECG and ECHO did not reveal any cardiac abnormalities.

Management and Outcome:
She was advised oral retinoid therapy - Tab Acitretin 25mg once a day and was explained both risks and benefits associated with oral retinoid therapy, but patient refused as she was planning for a baby. So she was given topical keratolytics - 6%salicylic acid twice a day for 3 months on which the thickness and size of the plaques gradually reduced following the treatment.

Discussion:-

There are four types of wooly hair, including dominant, recessive, acquired, and nevus.[2] Wooly hair usually appears as a solitary problem but has been reported in conjunction with a series of cutaneous and extracutaneous abnormalities. The evaluation of patients with diffuse congenital wooly hair includes the following: complete family history, skin examination to identify keratosis pilaris, PPK, webbed neck, facial dysmorphism, skin fragility, koilonychia, and hypoplastic nails and teeth; systemic examination to exclude cardiac involvement, progressive neurologic degeneration, osteoma cutis, short stature, deafness, and hypogonadism.[3] Wooly hair palmoplantar keratoderma are of 4 types of which in type 1 (NAXOS syndrome) has diffuse palmoplantar keratoderma, wooly hair and recessive form of arrhythmogenic right ventricular cardiomyopathy; type 2 (CARVAJAL syndrome) has striate keratoderma, wooly hair and dilated left ventricular cardiomyopathy; type 3 (NAXOS-like phenotype) has arrhythmogenic right ventricular cardiomyopathy, wooly hair and mild palmoplantar keratoderma; type 4 has wooly hair with palmoplantar keratoderma without cardiac abnormality.[1] Our patient had wooly hair in association with Palmoplantar keratoderma, without any other cutaneous or systemic abnormalities which fits in type 4. The prevalence of wooly hair with keratoderma worldwide is unknown.

Conclusion:-

Palmoplantar keratoderma (PPK) comprises a heterogenous group of disorders, which can be subdivided into hereditary and acquired forms. Classification of the hereditary PPK is difficult because of interindividual and intraindividual variations and differences in nomenclature.[4] The majority of the disorders in this group are hereditary, some are part of other dermatoses, and a few are acquired. The underlying genetic abnormalities have been identified for many of PPK; the involved genes encode intracellular structural proteins, desmosomal proteins, GJ components, and enzymes.[5] Evaluating a patient with PPK should include cutaneous examination of mucous membranes, extracutaneous manifestations like ophthalmologic, dental evaluation. A skin biopsy for histologic examination may provide additional diagnostic clues. Genetic analysis is helpful to establish the specific diagnosis, facilitate screening of family members, and enable prenatal diagnosis.

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