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RESEARCH ARTICLE

MENDES DA COSTA SYNDROME MASQUERADING AS PSORIASIS VULGARIS- A CASE REPORT

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Abstract

Erythokeratodermavariabilis(EKV) is rareAutosomal dominantdisease with two types of presentations.Onset is usually in infancy. Two types of lesions:

1) Relatively fixed well demarcated keratotic and erythematous plaquesover extensor surfaces.
2) Transient erythematous, polycyclic or comma shaped macular lesions occurring at any site. We arereportingmale patient presenting withasymptomatic hyperkeratotic plaques over extensors,withoutseasonal variationsand withoutpositivefamily historybut clinically mimicking as psoriasis vulgaris.

Introduction:-

Erythrokeratodermiasarea heterogenous group of inherited cornification disorders. EK wereclassified asi) confluent and reticular EK of GougerotandCarteaud, ii) variable EKof Mendes da Costa and iii) symmetric progressive EK. Erythrokeratodermavariabilis(EKV) is a rare,ichthyosiformdisorder, first, described by Mendes da Costa in1925[1].

Onset isusually ininfancy. Two typesof lesions: 1) Relatively fixed welldemarcated keratotic and erythematous plaques over extensorsurfaces. 2) Transient erythematous, polycyclic or commashapedmacular lesions occurring at any site. Progressive symmetricalerythrokeratoderma(PSEK), clinical Variant ofEK with symmetrical appearance; with little pruritus, lesionsare non migratoryin nature[2].The lesions slowly become progressive in number and size, which increases throughoutthe year[3].

Case Report:

A 59year old male presented with hyperpigmented plaquesover hands and legs since 10years, lesions areasymptomatic. On examination, multiple hyperpigmented, hyperkeratoticplaques noted over bilateral extensor aspectsof elbows, knees, ankles, dorsa of hands. Plaques are of variable sizes having irregular margins.There is

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no exacerbationof lesions on exposure to cold, heat or emotional stress. Patient isnot on any medications. No history of similarcomplaints in the family.Palms andsoles normal.Teeth, mucusmembranes, hair are normal; nails normal.Histopathological examinationrevealed epidermis-focal spongiosiswith normal granular layer and moderately thickened stratumcorneum with parakeratosis and orthokeratosis.Sparse superficialperivascular lymphocytic infiltrate withepidermal hyperplasia is noted. Complete blood count, liverfunction test, renal function test,urine analysis were withinnormal limits.

Figure 1:- Hyperkeratotic plaques over elbows, knuckles, interphalangeal joints.

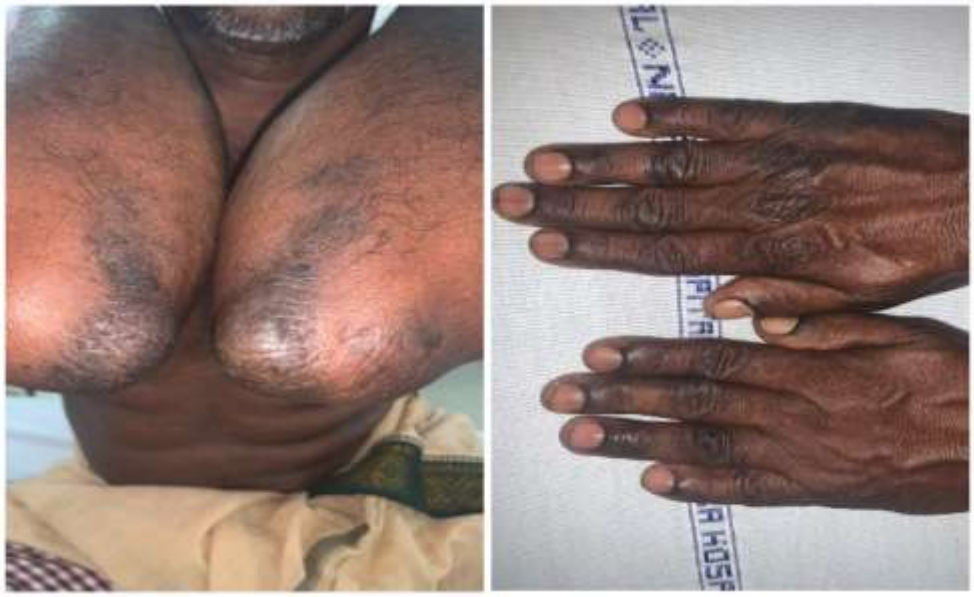


Figure 2:- Hyperkeratotic plaques over elbows & ankles.

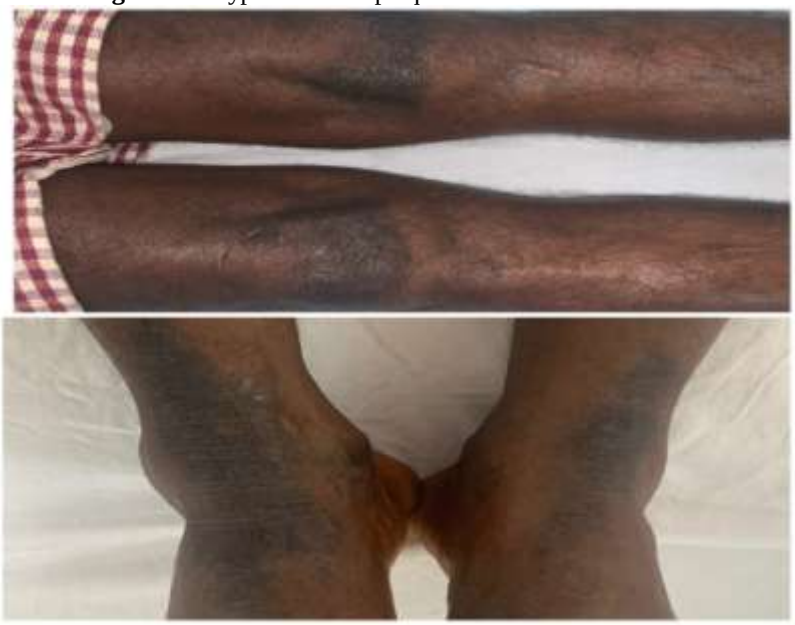
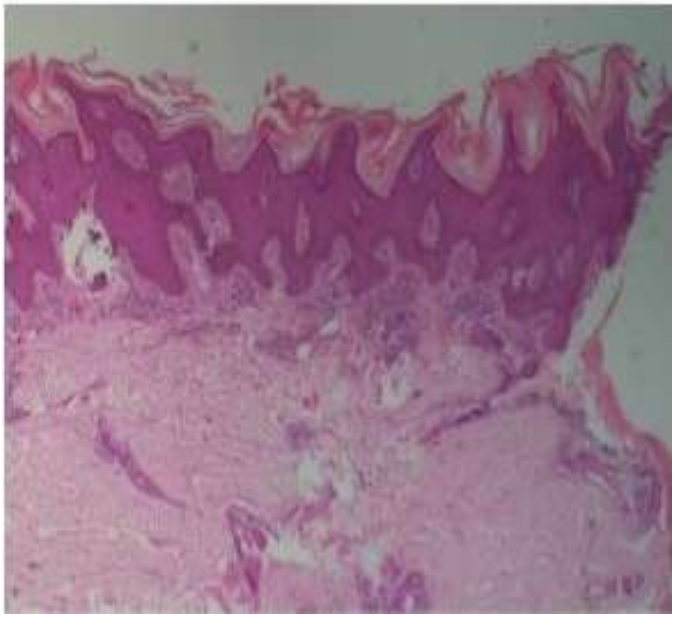


Figure 3:-Histopathology.



Discussion:-

The classicalEKV initially described by Mendes daCosta is characterized by two types of skin lesions: (a) figurate hyperkeratotic plaques and (b) transient erythematous areas. Inheritance is usually autosomal dominant. There is a defect in the GJ connexin proteins Cx31 and Cx30.3 expressed in the granular layer of the epidermis[4].These subtypesare independent, and their shapesand distribution can be changeable at any time[5].Disease ischaracterized by well demarcated, keratotic and erythematousplaques over extensor surfaces. We are here reporting a case of 59 years old male with hyperpigmented plaquesover hands and legs since 10years, lesions areasymptomatic. On examination- multiple, hyperkeratotic, hyperpigmentedplaques noted over bilateral extensor aspectswith presentation similar to psoriasis vulgaris, lackspositive family history but diagnosed as EKV on histopathology.

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