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A CASE REPORT ON LINEAR POROKERATOSIS TRANSFORMING INTO SQUAMOUS CELL CANCER.

Dermatology

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ABSTRACT

A 27year old male presented with annular asymptomatic lesions in the past 10 years that first appeared along right upper arm following Blaschkow's line. It then progressed to entire right arm, upper chest, abdomen, groin area and medial aspect of right upper thigh The lesions were initially brown papules 1-3mm then transformed into plaque 8-10mm with atrophic centers and hyperkeratotic borders. In past three years he developed painless nodular growth with verrucous surface over lesions around elbow, groin area, and axilla. The lesions ulcerated with necrotic slough over the surface. He was histopathologically proven to be a case of porokeratosis progressing to squamous cell cancer. He was treated with oral acitretin and referred to oncology department for further management.

KEYWORDS

Porokeratosis, Coronoid Lamella, Keratinization.

INTRODUCTION:

Porokeratosis is a disorder of keratinization, characterized with keratotic papules or plaque having raised border and central atrophy. It can be congenital or acquired. The condition might occur at any age but more commonly seen in fifth decade of life. The characteristic histological feature is coronoid lamella. Dermoscopy of the lesion shows volcanic crater appearance.

Case Report:

A 27 year/male presented with multiple annular asymptomatic lesions that first appeared along Right upper arm following the Blaschko's line, then progressed to entire Right arm, upper chest and abdomen, groin area and medial aspect of Right upper thigh since 10 years, family history and any triggering factor – insignificant. The lesions were initially brown coloured papules, 1-3mm, then transformed to brown coloured plaques 8-10mm with thin atrophic centers and raised Hyperkeratotic borders. It was until 3 years back when patient noticed painless nodular growth with verrucous surface over porokeratotic lesions around elbow, groin area and axilla, it then gradually enlarged, became ulcerated at around 2 years back and presented with necrotic slough over the lesions with average size of cancerous growth of about 50×25 cm. complete blood count, liver function test, renal function test, lipid profile, serum electrolytes and protein were normal, viral markers were negativePunch biopsy samples from porokeratotic lesions and ulcerative growth over groin area was taken. On the basis of history, clinical examination and histopathological findings-diagnosis of: linear Porokerarosis with squamous cell cancer was made.

Patient was treated with oral acitretin at the dose of 25mg HS and topical sunscreen plus 5FU HS.After histopathological confirmation of squamous cell cancer patient was referred to oncology department.

DISCUSSION:

Porokeratosis is inherited as AD genetic condition with mutation in PMVK gene 12q24 characterized with asymptomatic keratotic or lichenoid papules or plaques, with distinct keratotic raised edges and hypo or hyperpigmented atrophic centers. Linear PK develops along Blaschko's line and it is more common among females than males [3]. Among variants of porokeratosis; linear porokeratosis is the rare one with incidence of 1 in 2,00,000[2]. Among other variants malignant potential of linear porokeratosis is maximum and the most common cancer being the squamous cell cancer [2]Malignant transformation has been reported in 6.9 to 11.6 % cases of porokeratosis [1] [2] [5]Other treatment modalities: topical calcipotriol, imiquimod, retinoides, Nd -Yag [3]

Differential diagnosis includes actinic keratosis, basal cell epithelioma, acrokeratosis verruciformis, lichen planus annulare, linear verrucous epidermal nevus.

Treatment –
first line includes topical imiquimod, 5-FU, vit D analogues, cryotherapy, FCo2 laser.

Second line treatment is oral retinoides.

Acknowledgement: none

Conflict of interest: none declared

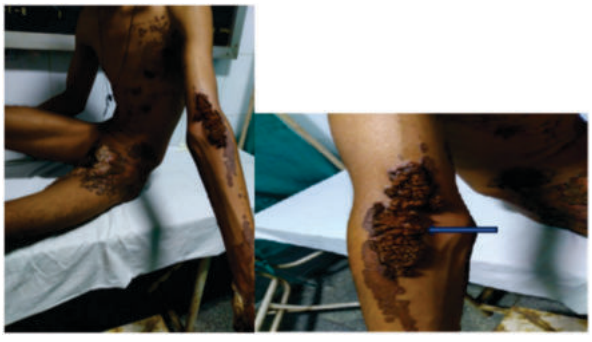

Fig 1: A. linear hyperkeratotic lesions along left fore arm following blaschkow's line, similar lesion on upper chest and groin with nodular swelling along right groin area with necrotic slough at the center. B. porokeratotic lesions with verrucous surface near elbow.

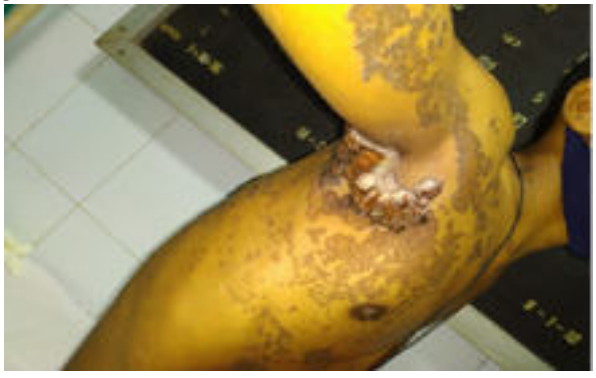

Fig 2: nodular growth over hyperkeratotic plaque along right axilla with necrotic slough on surface.

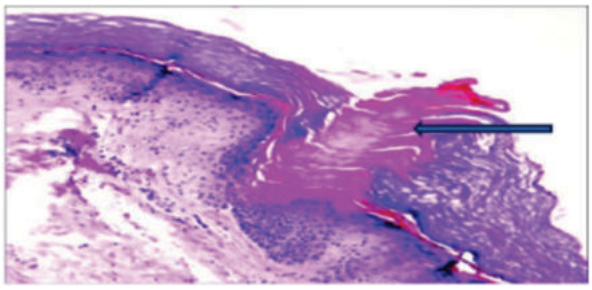

Fig 3: histopathological image showing coronoid lamella

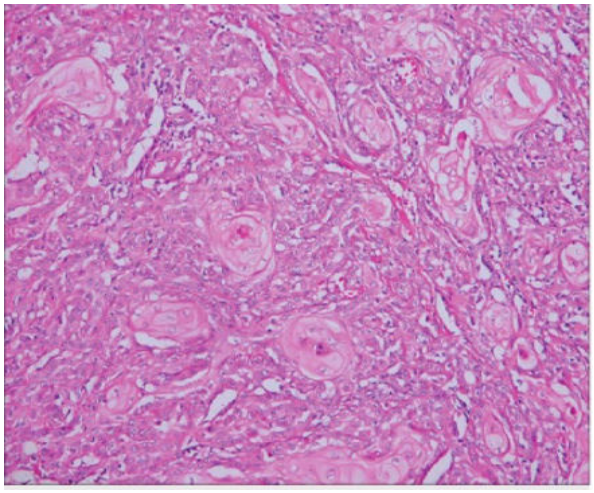

Fig 4: histopathological image showing features of squamous cell cancer

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