Disseminated Superficial Actinic Porokeratosis

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Summary

A Case Report about a rare Geno dermatosis -Dissamenated Superficial Actinic Porokeratosis is presented.

Key Words

Classic porokeratosis of Mibelli (PM), disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminata (PPPD), linear porokeratosis,

Introduction

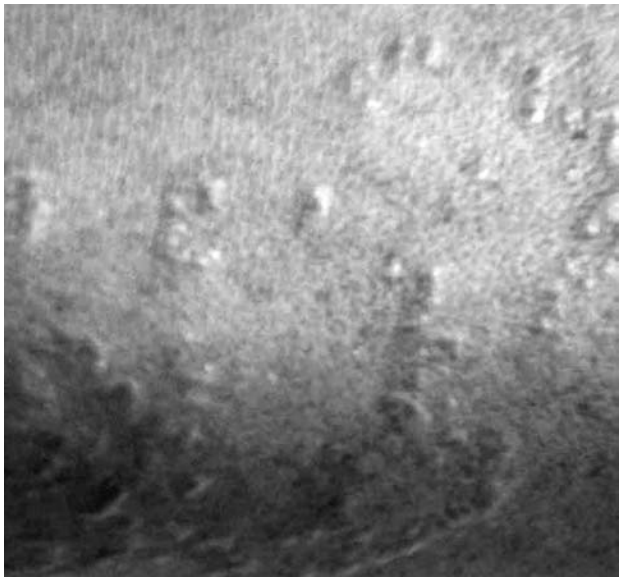
Porokeratosis is a clonal disorder of keratinization characterized by one or more atrophic patches surrounded by a clinically and histologically distinctive hyperkeratotic ridge like border called the cornoid lamella[1]. Ulcerative lesions have been described. Giant PM in a facial or acral location may cause destruction of underlying soft tissue or pseudoainhum with amputation[2].

Malignant degeneration has been reported in all forms of porokeratosis, with risks of 7.5% and 11% determined, large lesions, lesions of long-standing duration, and the linear type of porokeratosis were found to be at greatest risk.[3] A squamous cell carcinoma developing within a large PM lesion, associated with extensive metastases and hypercalcemia, has been reported.[4,5]

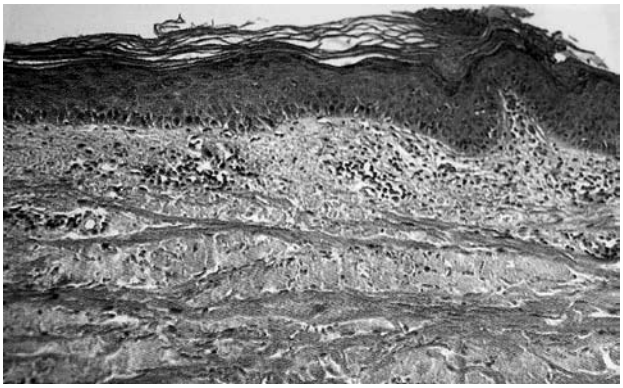
Case report

A 45 year old female, farmer, presented with multiple annular hyperpigmented raised papular and plaque lesions with central atrophy, bilaterally over forearms since one year.(Fig.1) The lesion began as progressive asymptomatic small keratotic erythematous papules, Spread peripherally to become brownish circinate plaque with central atrophy over the extensor surface of both forearm. There was no loss of sensation and no hair loss over the affected area. None of the Mucous membranes were involved. Patient is not known to be diabetic nor having any other significant systemic diseases.

General Physical & systemic Examination did not reveal any abnormality. Routine laboratory investigations were within normal limits including screening for Hepatitis B & C virus. Skin biopsy from lesion in forearm showed keratin filled invagination of the epidermis and invagination was less pronounced. Center of this keratin filled invagination showed parakeratotic column – coronoid leamella, features suggestive of: Actinic Porokeratosis- Plaque Type, with no evidence of malignancy (Fig 2)



Picture1 Skin lesion at Right forearm



Picture 2 Skin section from Lesion (H&E, 20X) showing Cornoid lamella

Discussion

Porokeratosis is represents a heterogeneous group of disorders of keratinization and has a wide variety of clinical manifestations. Clinically, the basic lesion is a sharply demarcated and hyperkeratotic plaque with central atrophy. Histopathologically it is characterised by the presence of cornoid lamella. The lesion of classical porokeratosis was described by Mibelli in 1893 and Cherosky described disseminated superficial actinic porokeratosis (DSAP) first time in 1966.

Five distinct clinical variants of porokeratosis are recognized: classic porokeratosis of Mibelli (PM), disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminata (PPPD), linear porokeratosis, and punctate porokeratosis. Several other variants have been described, including hyperkeratosis types, a pruritic papular variant, and an unusual verrucous variant that is localized to the buttocks and mimics psoriasis[6]. Occasionally, a patient may develop more than one type of Porokeratosis

Clonal hyperproliferation of atypical keratinocytes leads to the formation of the cornoid lamella which expands peripherally and forms the raised boundary between abnormal and normal keratinocytes. Local or systemic changes in immune function may allow the development of atypical clones of keratinocytes. An autosomal dominant mode of inheritance has been established for familial cases of all forms of porokeratosis.

Several risk factors for the development of porokeratosis have been identified; these factors include genetic inheritance, ultraviolet radiation and immunosuppression. Immunosuppression associated with porokeratosis may be secondary to a disease process such as HIV infection or lymphoma or an iatrogenic suppression such as with immune-modulating drugs used to prevent organ transplant rejection or to treat autoimmune diseases.[7]

Sun exposure is thought to cause DSAP, Excessive natural or artificial ultraviolet radiation, electron beam therapy, and extensive radiation therapy are well-established trigger factors. Immunosuppression may induce new lesions or cause pre-existing lesions to flare.

The formation of squamous or basal cell carcinomas has been reported in all forms of porokeratosis.[8] Chromosomal instability and reduced immune surveillance with overexpression of p53 are hypothesized to play a role in the development of cutaneous malignancies within porokeratosis[8,9]

DSAP is relatively common and other forms of porokeratosis are rare.Disseminated superficial (actinic) porokeratosis DSAP is 3 times as likely to develop in women compared with men. Skin lesions charecterstically are Multiple, brown, annular, keratotic lesions that develop predominantly on the extensor surfaces of the legs and the arms characterize DSAP. They are usually asymptomatic.Patients are typically women in their third or fourth decade of life with history of excessive ultraviolet exposure. Patients may have a history of phototherapy for psoriasis. Nonactinic forms may be seen following electron beam total skin irradiation, organ transplantation, hepatitis C virus–related hepatocellular carcinoma,[10] HIV infection, renal failure, or in association with other causes of immunosuppression.

Several modalities of treatment has been reported with variable success.In localised disease-topical application and in generalised disease systemic treatment is mandatory[11].

.Fluorouracil[12] 1 to 5% cream may be Apply to lesions.

Drug Preferentially taken up by cells that are dividing abnormally and It interferes with DNA and RNA synthesis leads to death of the abnormal cells.

.Vitamin D analogues-These agents regulate calcium-induced keratinocyte differentiation. Calcipotriene cream 0.005% is commonly used

Retinoids -These agents decrease the cohesiveness of abnormal hyperproliferative keratinocytes and may reduce the potential for malignant degeneration. Tretinoin -0.025%, 0.05%, and 0.1% creams available. for topical use.Isotretinoin can be given Orally to treats generalised conditions. isotretinoin therapy. 0.25-1.5 PO mg/kg/d . 20 mg PO qd combined with topical 5-fluorouracil has been successful Biological Response Modulator -Imiquimod 5% cream available for topical use. Drug Induces secretion of interferon-alpha and other cytokines but mechanism of action is not yet known.

Finally, the prognosis is generally excellent but regular monitoring of patients for the development of malignant transformation is essential, especially in the setting of immunosuppression. Squamous cell or basal cell carcinomas can be aggressive in patients who are immunosuppressed. And Prophylactic excision should be considered in appropriate situations.. Patients must practice strict sun protections. These measures include wearing protective clothing; applying sunblock; avoiding exposure to midday sunlight; and discontinuing exposure to artificial ultraviolet light, such as tanning beds and therapeutic phototherapy. Patients must periodically examine their skin for lesions suggestive of malignancy. A qualified physician should promptly evaluate any change in a porokeratosis lesion and family members should be examined for porokeratosis if familial porokeratosis is suspected.

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