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Disseminated Superficial Porokeratosis Associated with Gastro Intestinal Malignancy

Dr. Akshaya R C[1], Dr. Thillaikkarasi[2], Dr. Narasimhalu C R V[3], Dr. Sonti Sulochana[4]

[1]PG Resident, Department of Dermatology, Venereology and Leprosy, Saveetha Medical College and Hospital, Thandalam, Chennai-602105, India
Email: akkurens*[at]*gmail.com

[2]Assistant Professor, Department of Dermatology, Venereology and Leprosy, Saveetha Medical College and Hospital, Thandalam, Chennai-602105, India

[3]Professor, Department of Dermatology, Venereology and Leprosy, Saveetha Medical College and Hospital, Thandalam, Chennai-60210

[4]Professor, Department of pathology, Saveetha Medical College and Hospital, Thandalam, Chennai-602105, India

Abstarct: ***Porokeratosis is an uncommon keratinization disorder characterized by papules or plaque and elevated borders with distinct histological feature of cornoid lamella. Disseminated superficial porokeratosis is one of the variant of porokeratosis, which may be rarely associated with immune suppressive conditions and hematological malignancy*****but disseminated superficial porokeratosis in association with internal malignancy is uncommon.*****Herein we report a rare case of superficial disseminatedporokeratosissince childhood associated with gastrointestinal malignancy. A 46 year old female was referred to dermatology OPD with complaints of multiple dark raised lesions over the face, truck, back, upper limband lower limb since childhood. For the past 10 months the lesions started to worse by increasing size varying from 0.5 to 12cm and morphology with raised annular margin and central atrophy and it was associated with weight loss, decreased appetite and fatigue. Relevant investigations were done including histopathological examination of the colon and cutaneous lesion which confirmed the diagnosis of adenocarcinoma of colon and disseminated superficial porokeratosis respectively. This case is presented for its interesting features, that includes, the involvement of the cutaneous lesion on the u-v exposed areas as well as and the non-exposed areas, its association of adenocarcinoma of colon and the presence of family history of cutaneous lesion in her mother.***

Keywords: Disseminatedsuperficialporokeratosis, cornoid lamella, gastrointestinal malignancy.

1. Introduction

Porokeratosis is an autosomal dominant or a acquired conditionattributable to abnormal epidermal keratinisation and proliferation[1]. Porokeratosis is morphologically classified as six clinical variants includes Classic porokeratosis of Mibelli (CPM), disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminate, and linear porokeratosis [2] eruptive pruritic papular porokeratosis (EPPP) [3]. Apart from this six classical morphological variant, there is six more atypical morphological variant of porokeratosisfacial, giant, punched-out, hypertrophic, verrucous, and reticulate porokeratosis[4].Porokeratosis can present at birth and not develop until adulthood [5]. In some type of porokeratosis there is a genetic predisposition with a presence of similar cutaneous manifestation running in the family [6], but the histologic presence of cornoid lamella in all types of porokeratosis is typical.

2. Case Report

Forty six year old female presented with multiple dark colour skin lesion all over the body. These lesions initially started at the age of 10 years. Lesions wear gradually increasing in size and number with rapid progression since one year. Family history were present but in milder form. On dermatological examination she had multiple hyperpigmented patches with elevated borders and size varying from 0.5 to 12 cm in diameter over face trunk upper limb and, lower limb. Sparing of the areas in the axillary, inguinal fold, palm sole and mucous membrane were noted. Biopsy was planned to confirm the diagnosis, 3.5mm punch biopsy was taken from the elevated border of a lesion present over the right lower limb. Histopathology of skin confirmed it as porokeratosis with the presence of cornoid lamella.

There were associated history of abdominal pain, loss of weight and reduced appetite for the past 10 months. Complete blood count showedhaemoglobin 7.4 g/dl, total leucocyte count 10200 cells/cumm, absolute neutrophil count 7500 cells/cumm. Liver enzymes, electrolytes, urine routine were normal and kidney function test showed creatinine 0.8mg/dl urea 25mg/dl.Carcino embryonic antigen was 7.9 ng/ml which was higher than the normal range (less than 3.00 ng/ml). peripheral smearrevealed microcytic hypochromic red blood cell. The parameters for blood transfusion were worked up and she was delivered two packs of packed red blood cell before the surgical procedure. Colonoscopy revealed multiple polyps of colon. She underwent total abdomen colectomy with colostomy surgery. Specimen sent for histopathological examination. Histology confirmed adenocarcinoma of colon with histologic grade 1 pT3 pN0 and two adenomatous polyps, each measuring 1.5cm with microscopic invasion into the muscle layer. Skin lesions were treated with emollients. As the disease can inherit the patient's family was educated of possible future transmission of the disease in herprogeny.

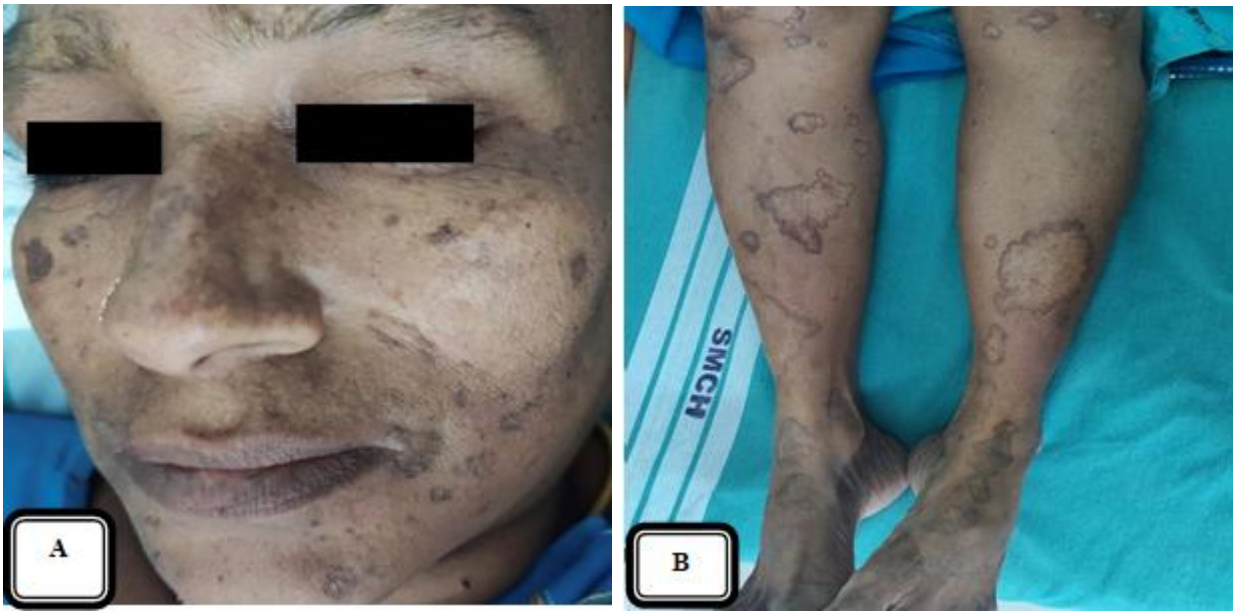

Figure 1: A. Multiple hyperpigmented plaque noted over face, few lesions have central hypopigmentation with surrounded hyperpigmented margin **B.** Multipleannular plaques with elevated borders and central hypo or hyperpigmentation with central atrophy. Its size varying from 0.5 to 12 cm in diameter noted over lower limb.

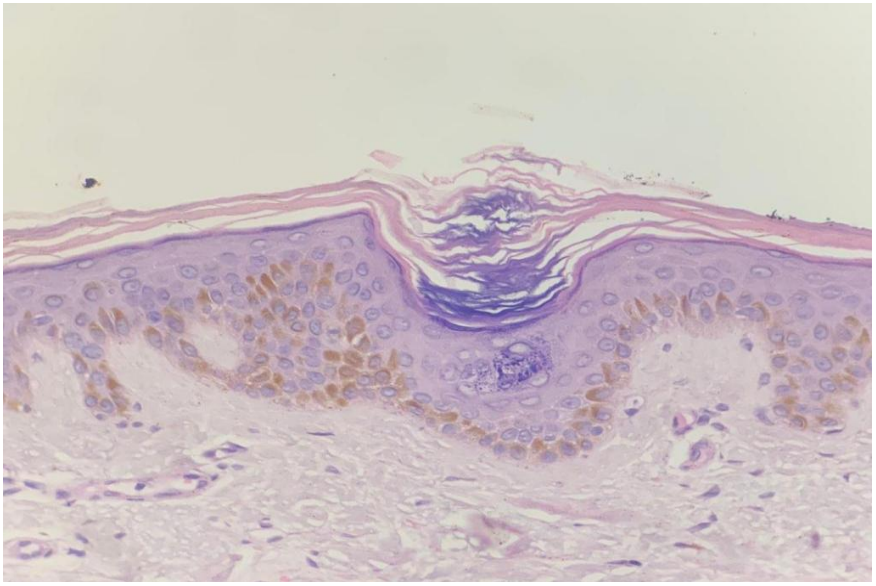

Figure 2: Section shows skin with keratotic plug with no granular layer and the presence of cornoid lamella. The underlying fibrocollagenous stroma show few capillaries

3. Discussion

Porokeratosis is a benign skin disorder with an autosomal dominant pattern of inheritance[7] or sporadic occurrence of the disease. In genetic studies, loci 18p11.37 has been identified for disseminated superficial porokeratosis[3]. Etiopathogenesis of porokeratosis is not clear. It has been suggested that the peripheral expansion of an abnormal mutant clone of epidermal keratinocyte at the parakeratotic column results in porokeratosis[8]. Risk factors contributing to porokeratosis are sun exposure, immune suppression and trauma [9]. Disseminated superficial porokeratosis is characterised by multiple sharply demarcated annular brown coloured lesions with atrophic centre and hyperkeratotic borders.The cutaneous lesion can occur anywhere in the body except the axillary vaults, inguinal folds, perigenital region, palms, soles and mucous membranes[10]. In histology porokeratosis reveals a parakeratotic column overlying a vertical zone of dyskeratotic and vacuolated cells within the epidermis, called the cornoid lamella[7]. Dermascopy examination of porokeratotic lesion shows double marginated white peripheral border representing cornoid lamella[11]. The major complication of a long standing disseminated porokeratotic lesion is development of squamous cell carcinoma, basal cell carcinoma within the porokeratotic lesion of elderly individual [12]. Clinical studies exposed association of disseminated superficial porokeratosis with immunosuppressive condition[8] like HIV, bone marrow transplantation, renal transplantation, hepatic transplantation, pregnancy[4] and also with internal malignancies like haematological malignancy, hepatic carcinoma, non-polyposis colon cancer, cholangiocarcinoma[13][14].

The treatment options of disseminated superficial porokeratosis includes sun protection, Keratolytic agents, topical 5-flourouracil, topical and oral retinoid agents, topical imiquimod, cryotherapy, photodynamic therapy, carbon dioxide laser ablation, excision, frequency doubled Nd: YAG[4], but attainingwhole resolution of lesions is challenging.

4. Conclusion

Highlights of our case report are: (a) Porokeratotic lesions involving almost all parts of the body from head to toe (b) presence of lesion since childhood with a positive family history. (c) Its association with adenocarcinoma of colon. Studies have been done in patients with pre-existing disseminatedporokeratosis individual and it's been said that 30% of them had a recently diagnosed malignancy [15]. Since incidence of disseminated superficial porokeratosis with internal malignancies have been reported, follow up is essential for early detection and management.

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