EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 5
Issue: 9
578-580
Year: 2018

GIANT POROKERATOSIS ON FACE- AN UNUSUAL SITE

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Article Received on 12/07/2018 **Article Revised on 01/08/2018** **Article Accepted on 21/08/2018**

ABSTRACT
Giant porokeratosis is a rare disorder of epidermal keratinisation characterised clinically by a large plaque of size varying from 10-20cm with an atrophic center and surrounding raised hyperkeratotic rim of size varying from 1-2 cm. There are very few case reports of giant porokeratosis in the literature. Giant porokeratosis has been reported on genitalia, buttocks, hand and feet and we hereby discuss a rare case of giant porokeratosis present over face in a 21- year old male patient. To the best of our knowledge this is the first case report of giant porokeratosis on face without any other association.

KEYWORDS: Porokeratosis, cornoid lamella, keratinisation.

INTRODUCTION
Porokeratosis is a clonal expansion of keratinocytes differentiating abnormally with a thin column of parakeratosis, cornoid lamella, which represents the raised active border histopathologically. Giant porokeratosis, a type of localised variety is a rare entity with a distinct large size which requires regular surveillence for development of any malignancy. It has been described on buttocks, genitalia, hands and feet.[1,3] In this case report we describe a case of facial giant porokeratosis which has not been described yet without any other association.

CASE REPORT
A 21 year old male presented with a single, asymptomatic dark brown colored lesion with raised boder over face since 19 years. Initially, a small erythematous plaque of size 2×2cm^2 was first noticed at the age of 6 years over his left cheek which gradually increased to the present size of 17×10 cm^2 involving his left and right cheek, glabella, nose and jawlines. The lesion had central atrophy with a well- defined raised hyperkeratotic rim of size varying from 0.5×1cm (Figure 1). His family history was negative. No history of artificial or natural UV exposure or any immunosuppressive drug intake could be elicited. General physical and systemic examinations were also unremarkable. Routine laboratory investigations including complete blood count, hepatic and renal function tests, serum electrolytes were within normal range. ECG and chest X-ray results did not show any signs of abnormality. Histopathological examination from the raised margin of the lesion showed hyperkeratosis, parakeratosis and acanthosis (Figure 2a). The characteristic finding of a thickened column of keratin containing parakeratotic nuclei known as 'cornoid lamella' extending outward from notch in the malpighian layer was also present, confirming our diagnosis of porokeratosis (Figure 2b). Considering the size and site of the lesion being an issue of aesthetic value to the patient, we planned him on cryotherapy alongwith 5% 5-fluorouracil cream once daily application without occlusion for 2 weeks. Two sessions of cryotherapy were done. It was performed with a hand-held cryogen spray canister using timed spot freeze technique. The lesion was treated for 30 seconds which led to the formation of ice ball and then it was allowed to thaw. Similar procedure was performed once more after 2 weeks. Active borders and inflammatory part of the lesion responded well to the treatment but he did not get much relief in the already scarred area. We counselled him regarding the prognosis of the condition. He was explained about the chances of development of malignancy in the lesion and and the necessity of regular follow up to look for any grave signs in the lesion.

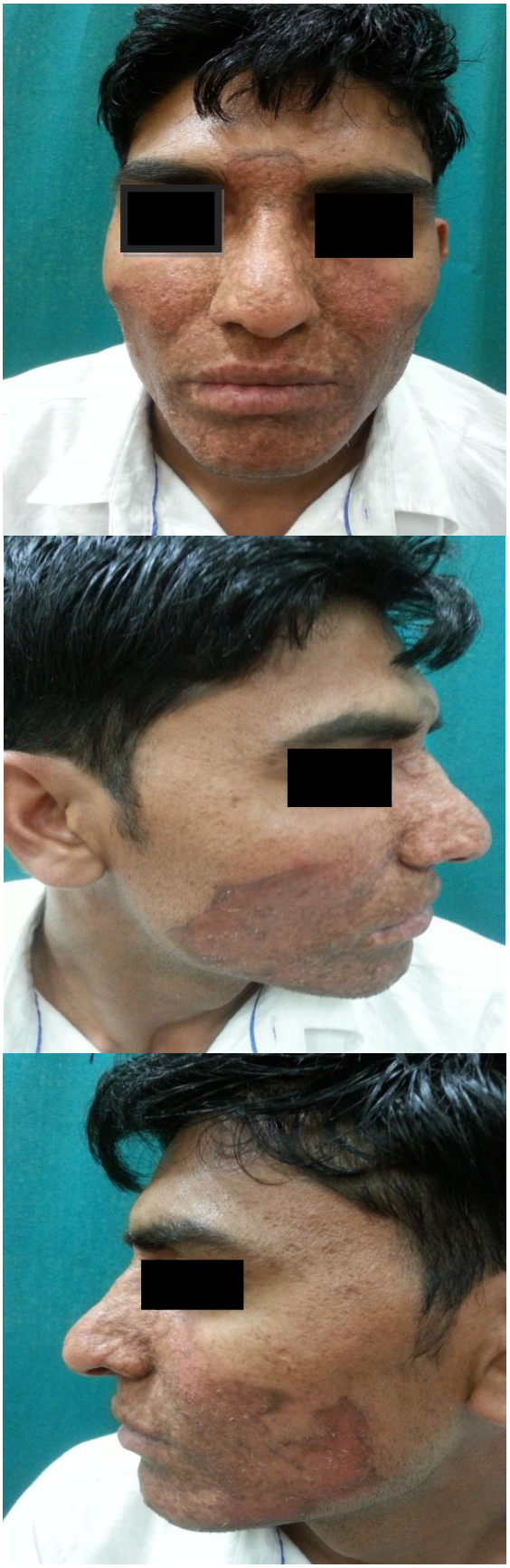
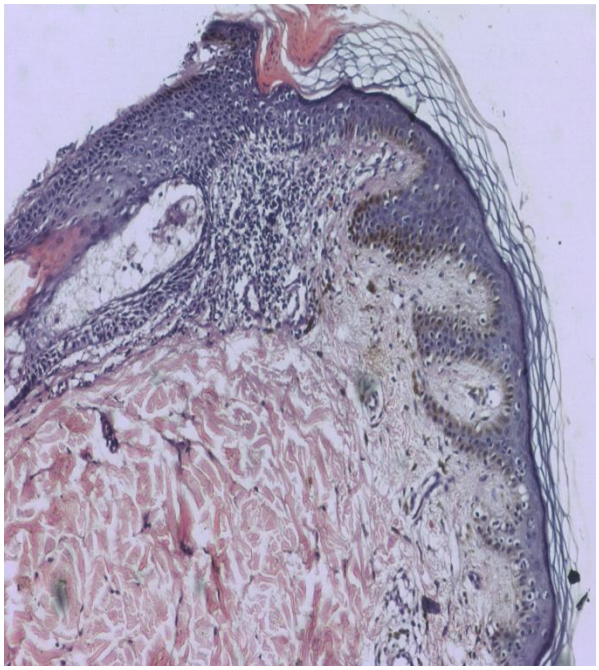
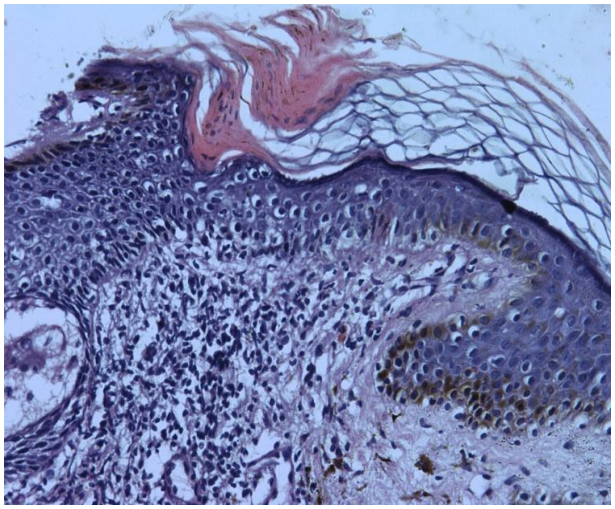


Figure 1: Giant porokeratotic plaque on face (a) front view (b) left view (c) right view.



Figuire 2: (a) Histopathologic photograph showing hyperkeratosis, parakeratosis and acanthosis (H&E,×10).



(b) A thickened column of keratin containing parakeratotic nuclei extending outward from notch in the malpighian layer (H&E,×40).

DISCUSSION

Porokeratosis is a heterogenous group of hereditary or acquired disorder of abnormal keratinisation. A number of localised and diffuse variety of porokeratosis are as follows- Porokeratosis of Mibelli, linear porokeratosis, punctate palmoplantar porokeratosis, genital porokeratosis, perianal porokeratosis. Disseminated forms include disseminated superficial actinic porokeratosis(DSAP), disseminated superficial porokeratosis, Systematized linear porokeratosis, disseminated palmoplantar porokeratosis.[4] Certain distinct rare variants are giant porokeratosis, verrucous porokeratosis, hypertrophic, reticulate porokeratosis.[5] Predisposing factors include drug-induced

immunosuppression in various diseases including organ transplantation, natural or therapeutic exposure to UV radiation are recognized trigger factors for DSAP and porokeratosis of Mibelli.[4]

Various treatment modalities like excision and grafting, cryotherapy, electrodessication, dermabrasion, carbon dioxide laser have all been tried depending on site, size, functional and aesthstic requirements.[6] Other newer modalities which have been used include 5% topical imiquimod, 5% 5-fluorouracil and systemic retinoids.[5,7] However the results are largely unsatisfactory tending to recurrence of lesion after stopping of treatment with a few modalities and moderate degree of response with others.

In our patient we tried cryotherapy alongwith 5% 5-fluorouracil but the response was unsatisfactory with mild improvement in the activity of the lesion but no response in the burnt out scarred area. The large size of the lesion in our patient is a matter of concern for development of malignancy such as squamous cell carcinoma. Therefore we advised our patient to come for regular check up and look for any signs pertaining to abnormal growth like sudden increase in size, surface irregularities, development of ulceration or hemorrhage. We have advised strict sunprotection with regular use of broad spectrum sunscreen(SPF 30) and regular monitoring for other disorders of immunosuppression is being undertaken.

A few case reports of giant porokeratosis present over sites other than face along with variants like DSAP, linear, mibelli have been reported. This case has been presented for its unusual presentation, as idiopathic occurrence of giant porokeratosis over face has not been reported so far and is also a matter of great aesthetic concern to the patient.

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