

## Curvaceous Cornoid Lamella- Porokeratosis

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**Received:** June 12, 2019; **Published:** July 12, 2019

### Preface

Porokeratosis was initially described by Isidor Neuman in 1875. Vittorio Mibelli designated the condition as "Porokeratosis" in 1893 on account of incriminated eccrine ostia. An infrequent disorder of obscure aetiology, deranged epidermal keratinisation and an unpredictable outcome defines classic porokeratosis.

Genital porokeratosis was initially described in 1985 by Helfman and Poulos. Chronic, persistent reticulated dermatosis incriminating the groin, genitals and thighs is cogitated which is refractory to treatment. Multiple cornoid lamellae are detected on histology which are characteristic of porokeratosis [1].

Genetically susceptible individuals enunciate expansive mutant clones of keratinocytes which classically induce porokeratosis. Porokeratosis is also triggered by specific factors such as exposure to ultra-violet light or immune suppression [1,2].

### Disease characteristics

Apart from the classical plaque type porokeratosis of Mibelli, additional variants incorporate disseminated actinic and non actinic superficial porokeratosis, porokeratosis in a linear arrangement, palmoplantar porokeratosis and punctuate porokeratosis. Exceptional categories include facial porokeratosis, giant porokeratosis, reticulate porokeratosis and eruptive pruritic papular porokeratosis. Infrequently, adjunctive clinical variants can arise within a singular subject.

However, classic porokeratosis of Mibelli can coexist with variants such as disseminated superficial porokeratosis or linear and hypertrophic verrucous porokeratosis. Adjunctive variants of porokeratosis display a familial concurrence [2,3].

Distinctive variants of porokeratosis incorporate:

- Classic porokeratosis of Mibelli,
- Disseminated superficial actinic porokeratosis (DSAP),
- Linear Porokeratosis,
- Porokeratosis Palmaris et Plantaris Disseminata (PPPD),
- Punctate porokeratosis.

The classic, autosomal recessive CDAGS syndrome manifests aspects such as craniosynostosis and clavicular hypoplasia, enlarged cranial fontanelle with delayed closure, defective hearing with auditory loss, anomalies of the anal canal, genitourinary malformations and cutaneous eruptions with porokeratosis which manifests as erythematous plaques on the skin surface. The syndromic condition is devoid of an associated molecular deformity [2,3].

### Clinical elucidation

Generalized porokeratosis with incriminating additional sites manifest lesions in the genital and adjacent regions such as buttocks, perineum, groin and proximal thighs. However, porokeratosis confined solely to the genital or gluteal region is rare. Keratotic papules or annular plaques with a miniature, centrifugally enhancing, elevated perimeter are denominated.

Genital porokeratosis is infrequent in females and is devoid of familial associations. Friction with clothing and scratching is implicated in the pathogenesis of genital lesions. Pruritus is common. Diagnosis of the exceptional condition can be delayed.

Squamous cells carcinoma can appear in comprehensive variants of porokeratosis, except punctuate porokeratosis. Elderly subjects, long established lesions and linear variants of porokeratosis depict enhanced proportion of malignant degeneration [3,4].

### **Histological elucidation**

Characteristic attribute of porokeratosis is the presence of cornoid lamella which is composed of a column of curvilinear parakeratosis with an inward inclination towards the lesion's centric zone accompanied by an absence of basal granular layer. Dyskeratotic keratinocytes emerge beneath the upper spinous zone and are cogitated as a diagnostic feature.

Cornoid lamella exemplifies an attenuated column of compressed parakeratotic cells extending from an epidermal invagination to invade the adjacent stratum corneum. Granular layer is absent or markedly attenuated. Additionally, dyskeratosis and pyknotic keratinocytes are enunciated with the demonstration of perinuclear oedema confined to the spinous layer [4,5].

Classic variant of Mibelli typically demonstrates minimal quantities of cornoid lamella at either side of the lesion. Multiple cornoid lamella can be discerned in penoscrotal porokeratosis or porokeratosis ptychotropica.

Cornoid lamella can also arise from adnexal structures, follicular infundibulum and eccrine ostia, as exemplified in porokeratotic adnexal ostial nevi. Cornoid lamella can emerge without characteristic clinical aspects such as annular papules or plaques.

Aforesaid manifestations are secondary to an epidermal reaction pattern of extended duration and respond to steroid administration [5,6].

### **Disease variants**

Porokeratosis confined to male and female genitalia depict lesions in scrotal, penoscrotal, penile, vulval and genito- cruro-anal sites. Lesions displayed on the buttocks are described as "gluteal" porokeratosis. Principal site of genital involvement is designated as "genito-gluteal porokeratosis.

Instances of genito-gluteal porokeratosis are denominated as

- 1) Classical porokeratosis where lesions are restricted to the genital region.
- 2) Ptychotropic porokeratosis.
- 3) Penoscrotal porokeratosis.

Aforesaid groups depict a common attribute of the presence of cornoid lamella which is diagnostic of classical porokeratosis. However, morphologic appearance of the lesions, incriminated sites and age of presentation is variable [4,6].

Classical porokeratosis of Mibelli can exceptionally be confined to the genital region, particularly in Asians and African Americans. Of uncertain pathogenesis, genetic and environmental factors are implicated in disease evolution. Middle aged males are commonly incriminated and detection of lesions is delayed to beyond one year duration. Lesions are usually discerned betwixt 22 years to 61 years of age with a mean age of initial diagnosis at 39 years. Lesions are frequently cogitated in the scrotum, buttocks, adjacent thighs, penis, glans penis and external urethral meatus. Majority of the lesions display features of classic porokeratosis of Mibelli with enunciation of annular plaques, centroidal atrophy, elevated margins and the typical ridge.

Linear configuration of the lesions can be observed or multiple erythematous linear atrophic lesions appear along dorsal penis and prepuce. Classic cornoid lamellae can be discerned [5,7].

Disseminated superficial actinic porokeratosis (DSAP) is the most frequent subtype of porokeratosis and commonly appears on the lower limbs of adult women. Multiple, attenuated papules are enunciated at focal sites. Associated genetic mutations exemplify SART3, SSH1 and ARPC3 genes. The variant displays a minimal possibility of malignant conversion.

Chronic immune suppressive conditions treated with extended narrow band ultraviolet B (NB-UVB) phototherapy can activate clone specific proliferation of aberrant keratinocytes.

Autosomal dominant disseminated superficial actinic porokeratosis (DSAP) demonstrates numerous erythematous plaques confined to the sun exposed skin. Palms and soles are exempt from the lesions.

Extensive and cumulative sun exposure, immune suppression, infections such as hepatitis and possibly phototherapy are implicated in the pathogenesis of DSAP. Narrow band - ultraviolet B (NB-UVB) radiation augments the release of keratinocytic growth factors with the concomitant suppression of immune system. Consequently, inflammation is reduced accompanied by modulation of cytokines, irradiation of T lymphocytes and promotion of apoptosis [2,3].

Lesions of disseminated superficial actinic porokeratosis (DSAP) are frequent on the face and abdomen and depict miniature, spherical hyper-pigmented macules with a keratotic, attenuated margin. A shallow, keratin filled invagination, cogitated as cornoid lamella, is enunciated on histology, accompanied by an irregular configuration of keratinocytes within the spinous layer [6,7].

Follicular porokeratosis of Mibelli restricted to the genitalia can occur. Brownish papules of one millimetre to five millimetres magnitude appear along the gluteal folds or natal cleft. Histologically, cornoid lamella arising from the hair follicles are discerned.

Tissue specimens display minimal, diagnostic cornoid lamella. Multiple cornoid lamella are discerned in ptychotropic porokeratosis and penoscrotal porokeratosis. Evaluation of surgical specimen is mandated in plaques of long duration, particularly genital plaques unresponsive to treatment [7,8].

Ptychotropic porokeratosis as a terminology was designed by Lucker in 1995 and is contingent to the flexural location of lesions. A distinct clinical presentation of porokeratosis is cogitated which exemplifies gradual, enhancing, dense, pruritic, verrucous plaques within the natal cleft and buttocks with a butterfly like configuration. Lesions of the particular variant are exhibited in the penis, scrotum, vulva, genito-gluteal region or buttocks with adjoining extremities. The classic, clinical ridge and moat appearance is absent.

Lesions can be enunciated as verrucous and hypertrophic porokeratosis. Although primarily confined to the genito- gluteal region, ptychotropic porokeratosis can appear at varying locales.

Histologically, multiple cornoid lamella are cogitated which differentiate the variant from classical porokeratosis of Mibelli. Additionally, amyloid deposits can be delineated in the upper dermis [8,9].

The condition is frequent in males with an age range betwixt 27 years to 84 years. Of obscure aetiology, friction with clothing is implicated in the pathogenesis of ptychotropic porokeratosis [2,3].

Penoscrotal porokeratosis is a distinct entity limited to an age group of male subjects within the second or third decade. Penile shaft or anterior scrotum is incriminated with the emergence of severe burning and itching.

Lesions can appear on account of frictional trauma or as a contact reaction. Pruritic, ill defined plaques appear at the incriminated sites. Multiple, contiguous cornoid lamella can be detected on histology, a proportion emerging from eccrine and follicular articulations. Clinically and histologically, lesions do not coordinate with typical plaque porokeratosis. However, lesions may concur as porokeratotic epidermal reaction with multiple cornoid lamella.

Typically, penoscrotal porokeratosis emerges in young males within the third decade. Plaques and diffuse patches with a rough, granular external surface are enunciated, accompanied with severe pruritus and burning sensation within scrotum and penile shaft [8,9].

### Differential diagnosis

Lesions of porokeratosis necessitate a distinction from conditions such as psoriasis, lichen simplex chronicus, hypertrophic lichen planus, tuberculosis of the skin and Bowen's disease.

Long established pruritic, verrucous plaques of ptychotropic porokeratosis require a distinction from psoriasis, chronic eczema, lichen simplex chronicus, dermatophytosis and candidiasis [8,10].

**Investigative profile**

Dermatoscopic analysis of penoscrotal porokeratosis exhibits characteristic aspects such as central brown pigmentation with numerous blue grey dots, a circumscribing, singular hypo-pigmented band and a peripheral “white track”.

Dermatoscopic evaluation of ptychotropic porokeratosis depicts well demarcated, annular lesions with a dense, brown perimeter confining an erythematous, non-atrophic centric zone with regular dotted vessels. Aforesaid attributes differentiate the variant from adjunctive inflammatory conditions such as psoriasis, dermatophytosis and lichen simplex chronicus [10,11].

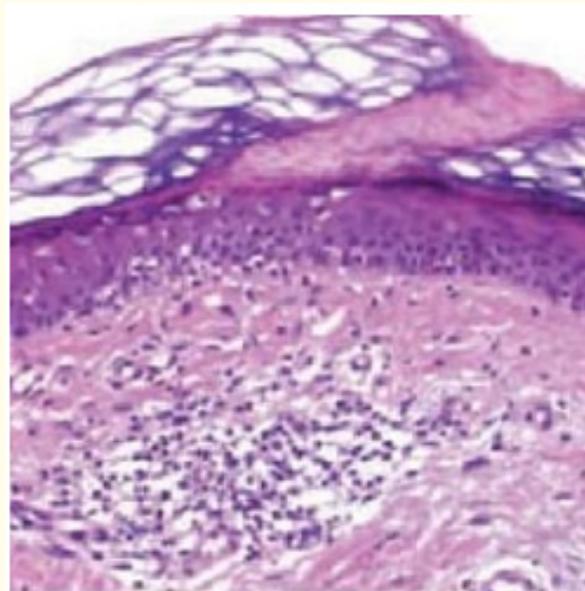
**Therapeutic options**

Lesions are treated with liquid nitrogen. The condition is devoid of reoccurrence. Therapeutic variations include the application of topical steroids, retinoids or 3% topical diclofenac cream. Options such as cryotherapy, carbon dioxide laser, oral retinoids, calcipotriol, 5- fluorouracil or imiquimod can be adopted although a long term benefit can be lacking.

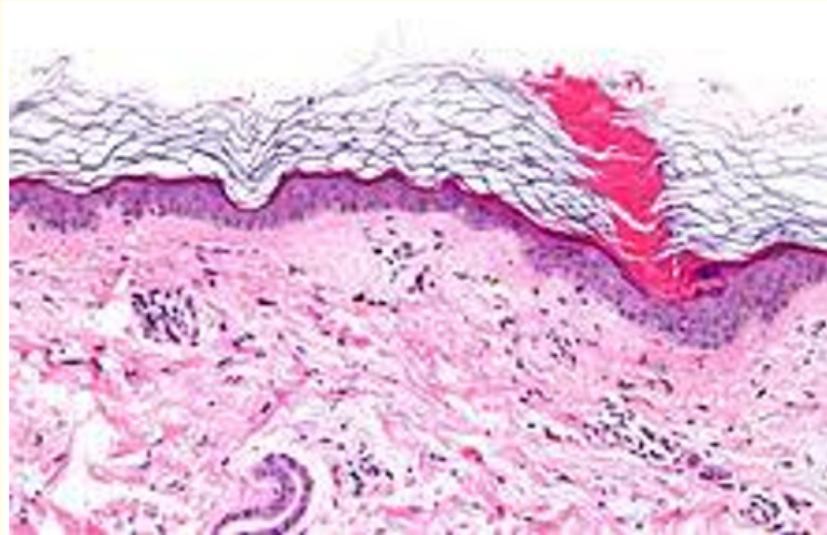
Cogent therapies offered for ptychotropic porokeratosis are cryotherapy with liquid nitrogen, 5 fluorouracil, imiquimod, calcipotriol, topical steroids, antifungal agents and lasers [12,13].

Dermatome can be employed to shave off the superficial tissue. The lesions treated with dermatome can reoccur and therapeutic response is usually inadequate.

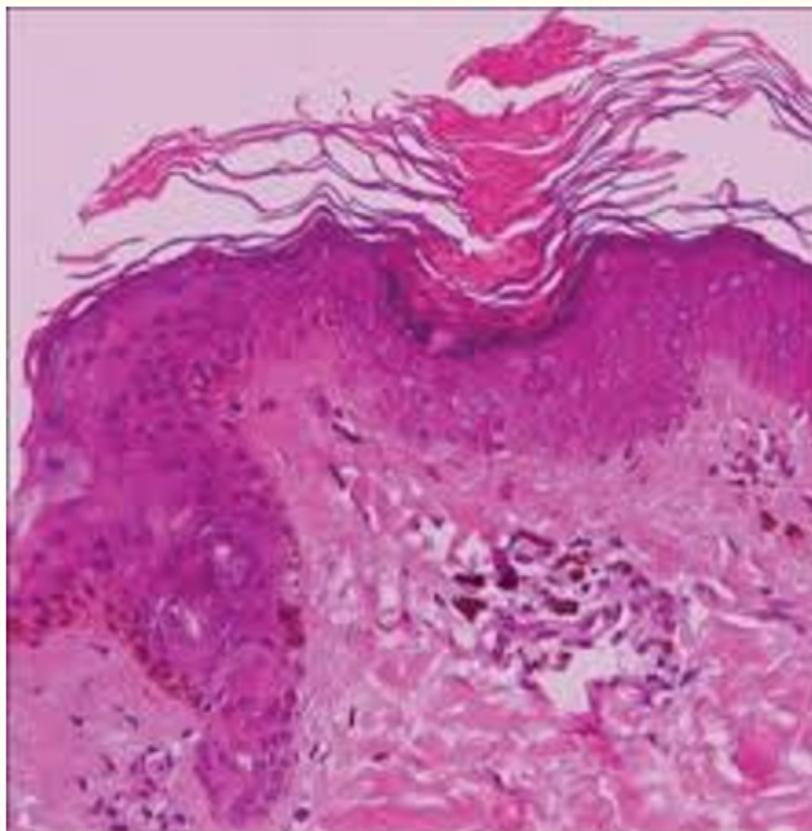
Penoscrotal porokeratosis is managed with antifungal agents and/or topical steroids or oral anti-histaminic agents which are satisfactory to a limited extent. Oral isotretinoin depicts partially attenuated lesions with decline in pruritus. Reoccurrence can occur on discontinuation of therapy. Topical and systemic therapy of penoscrotal porokeratosis is unsatisfactory [12,13].



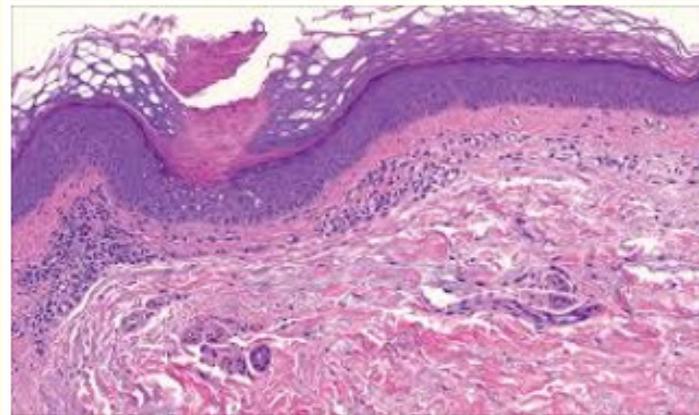
**Figure 1:** Acanthosis, parakeratosis, cornoid lamella and dyskeratotic keratinocytes in porokeratosis [14].



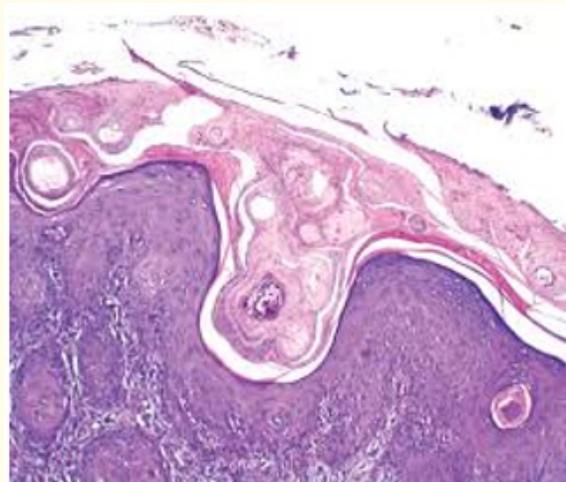
**Figure 2:** Hyperkeratosis and dyskeratotic keratinocytes in porokeratosis [15].



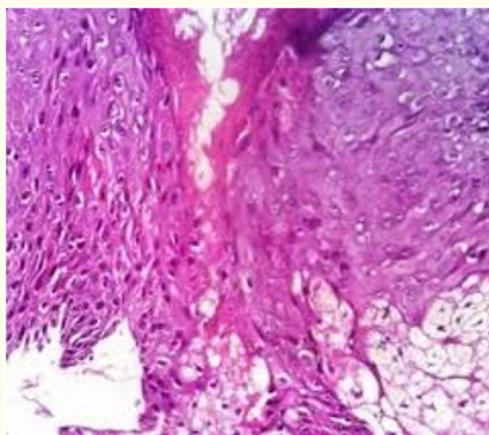
**Figure 3:** Acanthosis, hyperkeratosis, cornoid lamella and pyknotic keratinocytes in porokeratosis [16].



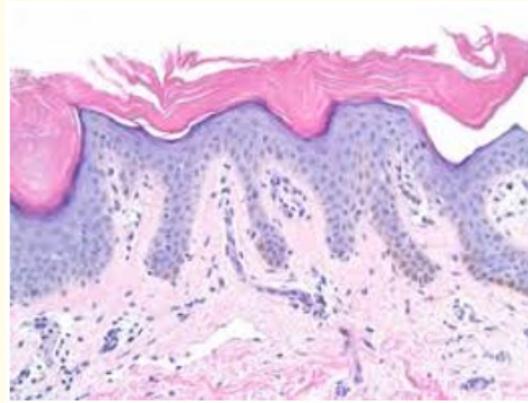
**Figure 4:** Linear configuration of porokeratosis with hyperkeratosis and dyskeratotic keratinocytes [17].



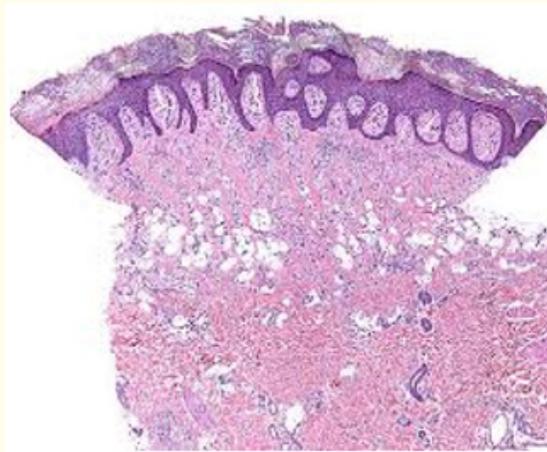
**Figure 5:** Solitary porokeratosis with cornoid lamellae, perinuclear oedema and impacted keratin [18].



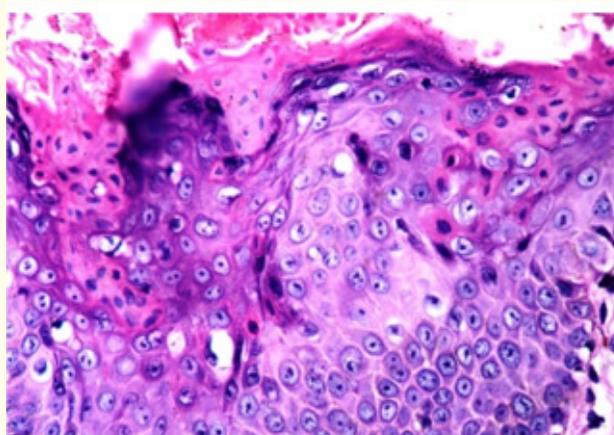
**Figure 6:** Porokeratosis with keratin impaction, cornoid lamella and dyskeratotic keratinocytes [19].



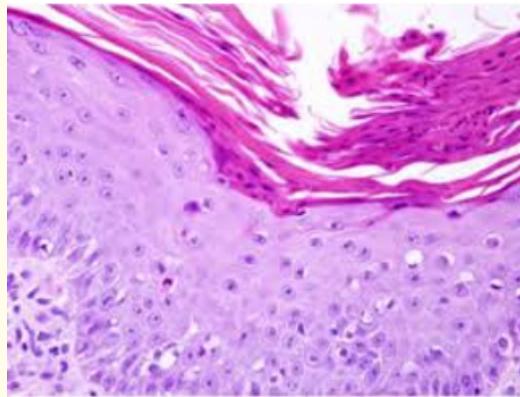
**Figure 7:** Acanthosis, papillomatosis, parakeratosis and keratinocyte dyskeratosis in porokeratosis [20].



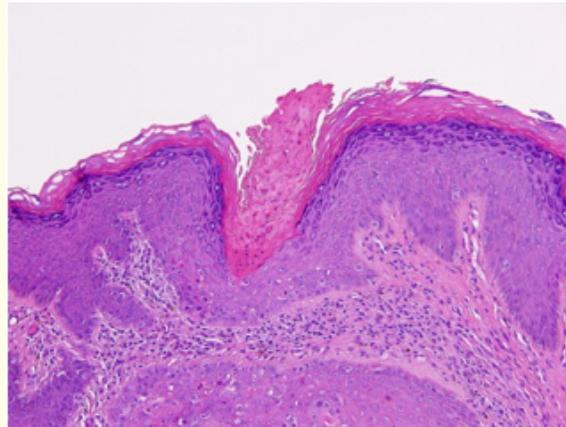
**Figure 8:** Ptychotropic porokeratosis with multiple cornoid lamella and reticulated dermatosis [21].



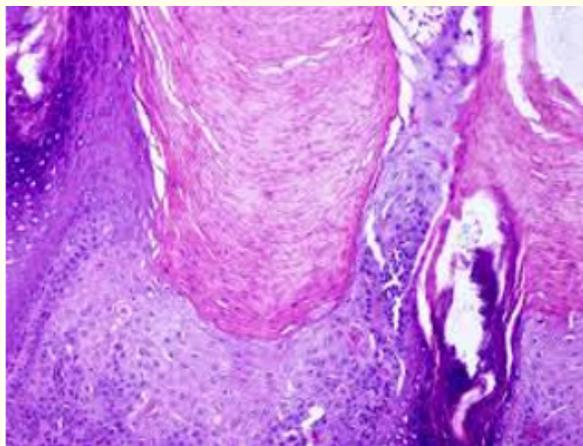
**Figure 9:** Cornoid lamella with dyskeratotic keratinocytes in porokeratosis [22].



**Figure 10:** Dyskeratotic keratinocytes and hyperkeratosis in porokeratosis [23].



**Figure 11:** Genital porokeratosis with acanthosis, cornoid lamella and dyskeratotic keratinocytes [24].



**Figure 12:** Cornoid lamella and dyskeratotic keratinocytes in porokeratosis [25].

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14. Image 1 Courtesy: Semantic scholar.
15. Image 2 Courtesy: Wikipedia.
16. Image 3 Courtesy: Indian Dermatology Online Journal.
17. Image 4 Courtesy: MDedge.com.
18. Image 5 Courtesy: Our dermatology online.
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**Volume 2 Issue 6 August 2019**  
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