Cameo

Giant porokeratosis

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Porokeratosis is a disorder of keratinization characterized by annular plaques with an atrophic center surrounded by a raised, keratotic wall. It has several clinical forms including a porokeratosis of Mibelli, giant porokeratosis, linear porokeratosis, disseminated superficial actinic porokeratosis, palmoplantar porokeratosis and punctate porokeratosis. We report a patient with the rare condition of giant porokeratosis. Several variants of porokeratosis coexist in our patient.

Case Report

A 41-year-old man presented with hyperkeratotic irregular plaques with central atrophy on his arms, legs, face and dorsal side of the left hand. There was no family history of similar lesions.

On dermatologic examination, he had an 11 × 8-cm lesion with central atrophy surrounded by a 1-cm raised hyperkeratotic wall on his left hand; 1–3-cm lesions with central atrophy surrounded by keratotic wall on his limbs, trunk, hands and feet (Fig. 1); and small annular lesions with a less distinctive border on his face and neck (Fig. 2). The histopathologic examination of the biopsy samples from the three different morphological lesions confirmed the diagnosis of porokeratosis (Fig. 3).

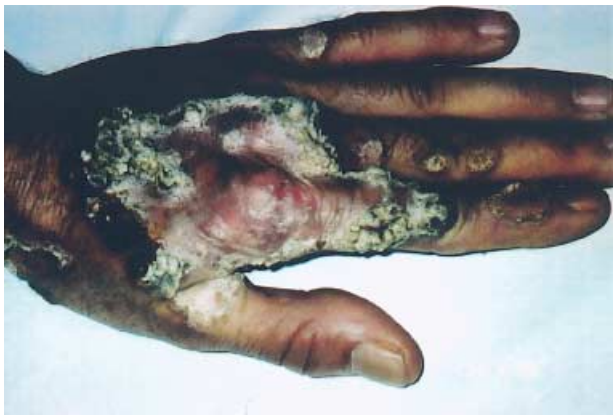

Figure 1 Giant porokeratosis lesion and Mibelli porokeratotic lesions on the left hand of the patient

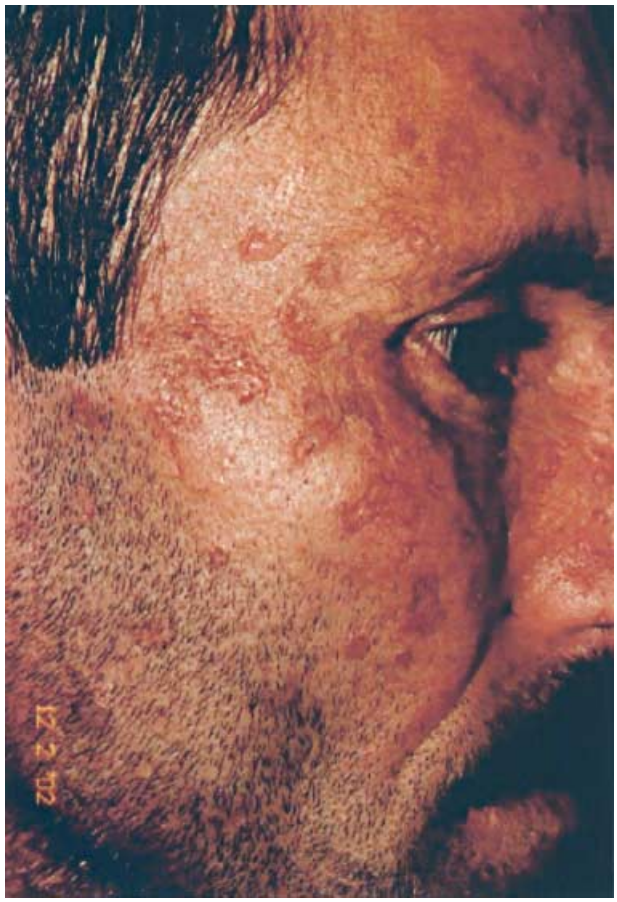

Figure 2 Disseminated superficial actinic porokeratosis lesions on the face of the patient

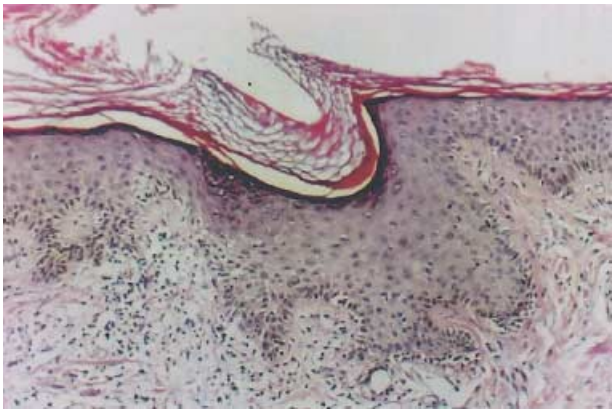

Figure 3 Granular layer is thin, and the epidermis invaginated and filled with keratin lamella (H&E ×110)

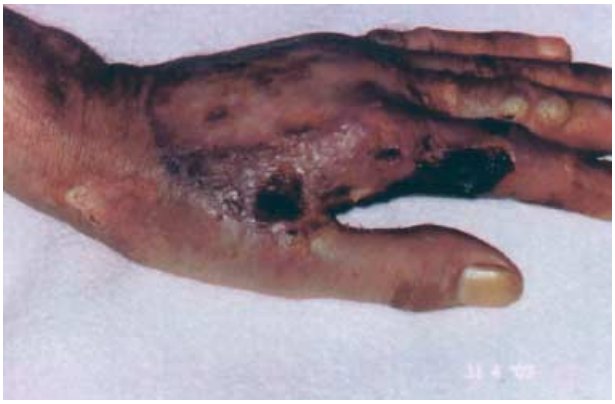

Figure 4 One month after the total excision and grafting of the giant porokeratotic lesion. The Mibelli lesion on the wrist healed with macular atrophy 1 month after cryotherapy. There were bullae formation on the 3rd and 5th fingers 1 week after cryotherapy

The giant porokeratosis lesion on the left hand was totally excised and grafted. Fifteen of the PM lesions were treated by cryotherapy. Lesions healed with macular atrophy (Fig. 4).

Discussion

Porokeratosis is a disorder of keratinization. Clinical forms include porokeratosis of Mibelli (PM), giant porokeratosis, linear porokeratosis, disseminated superficial actinic porokeratosis (DSAP), palmoplantar porokeratosis and punctate porokeratosis.[1]

The lesions of classic porokeratosis were described by Mibelli in 1893 as one or more localized, chronically progressive, hyperkeratotic, irregular plaques with central atrophy and prominent peripheral keratotic ridge. A more superficial, disseminated form was described at approximately the same time by Resphigi. In 1966 disseminated superficial actinic porokeratosis was described by Chernosky, and in 1971 Guss *et al.* added disseminated porokeratosis with palmar and plantar involvement (PPPD) to the spectrum.

Giant porokeratosis is a very rare form of porokeratosis. Several reports suggest it as a morphological variant of porokeratosis of Mibelli,[2,5,6] but others consider it as a different clinical form.[1,7]

The lesion may be 10–20 cm in diameter and the surrounding wall raised up to 1 cm.[1,2] Rarely, variants of porokeratosis may coexist in one patient.[2,8] Our patient has typical Mibelli lesions, 1–3 cm in size, with central atrophy surrounded by keratotic walls on his limbs, trunk, hands and feet, widely disseminated flat and small annular DSAP lesions on his face and neck as well as the giant porokeratotic lesion on his left hand. He was immunocompetent and had no other triggering factors. The histopathologic examination of the biopsy samples from the three different morphological lesions confirmed the diagnosis of porokeratosis. Giant and linear porokeratosis lesions may be seen with underlying bony anomaly and mutilation,[2,9] but none was present in our case.

The etiology of porokeratosis is unknown. An autosomal dominant mode of inheritance has been established for PM, PPPD, disseminated superficial porokeratosis (DSP) and DSAP. The similarities of clinical appearance and histopathology as well as the coexistence of different variants of porokeratosis in one patient or in several members of an affected family make us consider them as different phenotypic expressions of a common genetic aberration.[2]

Reed and Leone have proposed a mutant clone of epidermal cells expands peripherally, leading to formation of a cornoid lamella at the boundary between the clonal population and normal keratinocytes in porokeratosis. The tendency for abnormal clones to develop is probably inherited, and additional triggering factors such as ultraviolet light, photochemotherapy and loss of immunocompetence (HIV infection, organ transplantation) lead to clinical manifestation.[2,3] An alternative hypothesis is that an inflammatory mononuclear infiltrate composed of helper T cells, suppressor T cells and Langerhans cells beneath the cornoid lamella may provide a mitotic stimulus for overlying keratinocytes.[4]

The optimal treatment procedure must be selected depending on lesion size and localization, and functional and aesthetic requirements. Excision and grafting, cryotherapy, electrodesiccation, dermabrasion and CO_2 laser are all options.[2] Topical 5-fluorouracil is effective in PM, linear porokeratosis and in DSP and DSAP. Oral retinoids have conflicting results. Excellent results were obtained with oral retinoids in some patients with DSAP, widespread PM, PPPD, and linear porokeratosis. With retinoid therapy cytologic atypia has disappeared. Retinoids may have an inhibitory effect on cutaneous carcinogenesis in porokeratotic lesions.[2,10] One must be aware that relapses usually follow several weeks or months after discontinuation of retinoid therapy.[1,2]

The giant porokeratosis lesion on the left hand of our patient was totally excised and grafted. Fifteen of the porokeratosis Mibelli lesions were treated by cryotherapy, and these lesions healed with macular atrophy. Malignancy was not detected by histopathological examination in the excised giant porokeratosis lesion.

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Cameo

Systemic sarcoidosis presenting with alopecia of the scalp

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A 39-year-old African-American woman was referred to the department of dermatology, Mount Sinal Hospital, New York for evaluation of a skin lesion on her scalp which had been present for 3 years and had recently changed in appearance. On examination, she was found to have erythematous plaques with hair loss extending from the frontal hairline on the right side in a "C-shaped" distribution, terminating behind the left ear. Two years later the lesion had rapidly extended to the vertex of the scalp and left temporal area (Fig. 1). There was a 10 cm × 12 cm area of alopecia at the vertex with slight erythema, but there was no evidence of scarring, inflammation or nodularity of the scalp (Fig. 2). A skin biopsy taken from the forehead revealed granulomatous dermatitis showing noncaseating granuloma with negative acid fast bacilli (AFBC) and ammonical-silverstain for fungus (GMS) (Fig. 3).

She complained of chronic nasal congestion for 8 months. Nasal cavity examination and laryngeal endoscopy showed multiple nodules on the nasal septum and vocal cord. At that time, a presumptive diagnosis of sarcoidal nodule of the nasal septum and vocal cord was made and corticosteroids were administered orally.

A chest X-ray demonstrated prominence of the right peritracheal and perihilar regions consistent with sarcoid. A 67 Gallium scan disclosed increased uptake in the lung and perihilar lymph nodes.

A skull X-ray showed soft tissue density of the skull, probably indicating a subcutaneous sarcoidal nodule. Computed tomography (CT) scanning of her brain and skull base showed a soft tissue mass along the outer table of the calvarium within the frontal region.

Six years ago, she experienced visual changes: blurred vision, tearing, floaters on both eyes and a 1 cm × 1 cm subcutaneous, painful, firm mass on the lateral side of her left upper eyelid. A skin biopsy was performed in another hospital and revealed noncaseating granuloma.

A diagnosis of sarcoidosis involving the central nervous system, lacrimal gland, nasal septum, vocal cord, lung and scalp was made, and the patient was treated with 20 mg of methylprednisone on alternate days with intralesional triamcinolone injection for skin lesions. During the follow-up period, nasal, laryngeal, pulmonary, ocular and cutaneous lesions were slightly improved and magnetic resonance imaging (MRI) scanning will be carried out for further evaluation of the brain lesion.

Discussion

Sarcoidosis is a systemic disease with unknown etiology that is characterized by the accumulation of mononuclear phagocytes with the formation of noncaseating granulomas.[1] Multiorgan involvement, including the skin, lungs, lymphatic system, liver, spleen, eyes, parotid glands, central nervous system, bones, and joints, is possible.[2,3] Sarcoidosis occurs worldwide, affecting persons of all races, both sexes, and all ages.[4] African-American women between the ages of 30 and 39 years were found to have the highest annual incidence.[5] Sarcoidosis affects African-Americans more acutely with more extensive cutaneous involvement.[4] The lesions are morphologically extremely variable, are frequently atypical, and often demonstrate fibrinoid necrosis on histology.[6]

Figure 1 Erythematous plaque with hair loss on the left temporal area

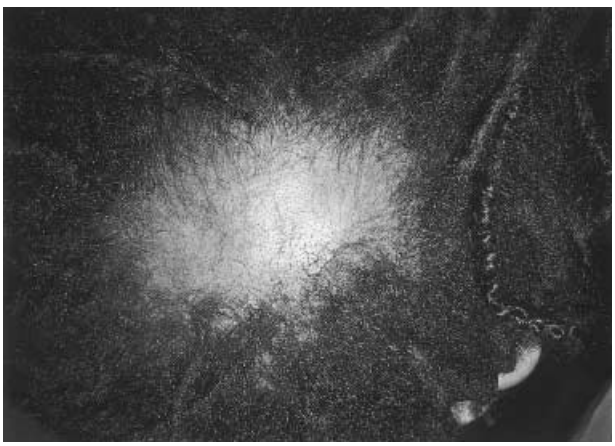

Figure 2 Nonscarring alopecia at the vertex with slight erythema

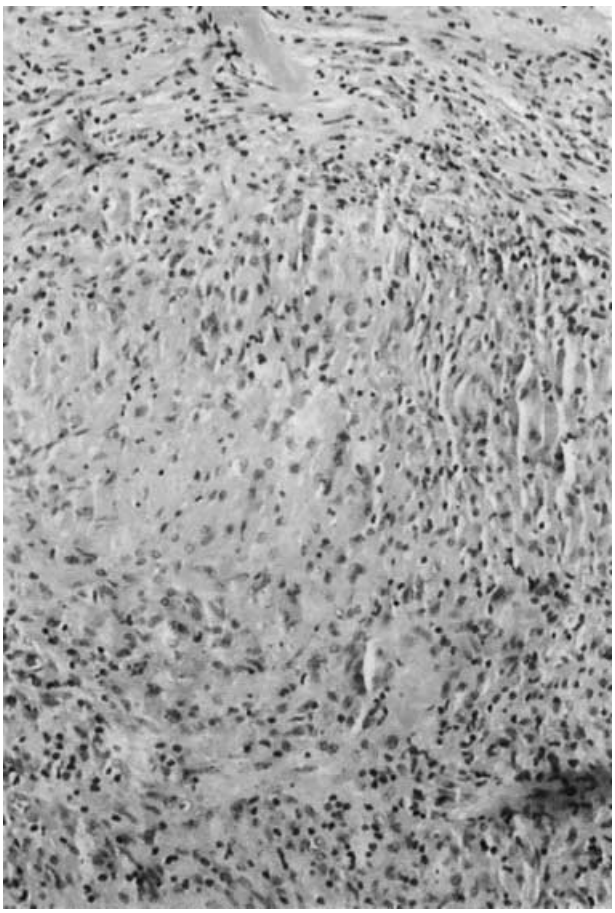

Figure 3 Noncaseating granuloma consisting of lymphocytes, histiocytes, plasma cells and multinucleated giant cells (hematoxylin and eosin staining; magnification ×100)

A diagnosis of sarcoidosis is generally made on the basis of a biopsy finding of noncaseating granulomas without other explanation.[4]

On average, 25% of sarcoidosis cases have cutaneous involvement that can occur at any stage; however, most often

cutaneous involvement occurs at the onset of the disease.[4,7] Scalp lesions, eyelid papules, andichthyotic lesions are also associated with severe systemic disease.

Although cutaneous involvement in sarcoidosis is relatively common, sarcoidosis of the scalp is rare. There are few reported cases of cicatricial alopecia due to sarcoidosis, which predominantly affects black women with evidence of intrathoracic sarcoidosis and cutaneous involvement at other sites.[8–11] Cases of nonscarring alopecia have also been reported, either with localized involvement or with total alopecia.[9] The high incidence of systemic involvement in reported cases indicates that a systemic work-up is needed when scalp sarcoidosis is diagnosed. This entity is usually resistant to treatment.[8]

There are no definitive diagnostic tests. A biopsy (skin, preitracheal nodes, or salivary glands) should be performed to obtain histologic confirmation of noncaseating granulomas in addition to pulmonary function tests, electrocardiography, slit-lamp eye examination, and tuberculin/anergy testing.[4,5]

Because sarcoidosis of the scalp may exhibit markedly variable morphologies, we emphasize that sarcoidosis should be considered in the differential diagnosis of plaques or nodules of the scalp as well as both cicatricial and noncicatricial alopecia.

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Cameo

N95 acne

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Two women, aged 27 and 45 years, presented to the Dermatology Outpatient Clinic with acne vulgaris. Both had nodular acne in a similar distribution over the cheeks, chin, and perioral areas (Fig. 1). Each had a history of acne vulgaris as a teenager. Both were healthcare assistants working in the Singapore General Hospital throughout the severe acute respiratory syndrome (SARS) crisis, had worn N95 masks continuously for about 3 months whilst on the wards, and had suffered an outbreak of acne of the skin occluded by the mask. They were treated with topical retinoid and systemic antimicrobials, and both responded well.

Discussion

Singapore was one of several countries affected by severe acute respiratory syndrome (SARS). The outbreak in Singapore started in March 2003. The Centers for Disease Control (CDC) advisory against travel to Singapore was lifted on May 4th, 2003 and the alert was terminated on June 4th, 2003.[1] There were 205 recovered cases, 33 deaths, and 238 probable cases in Singapore between March and May.[2] A number of these were healthcare workers infected with the

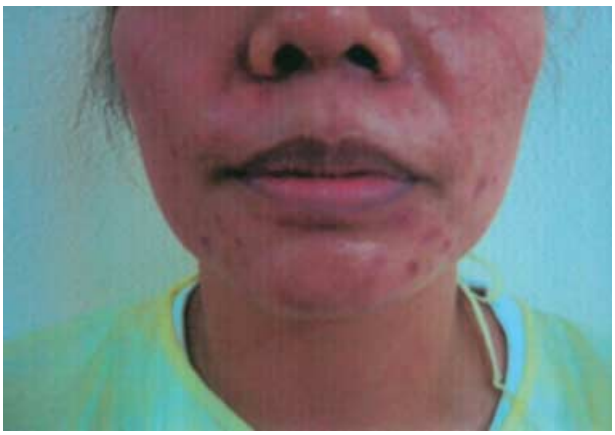

Figure 1 Twenty-seven-year-old woman with acne papules and nodules on the face over the area occluded by the N95 mask

SARS-associated coronavirus. Apart from SARS infection, healthcare workers were also affected in other ways. Skin disorders were seen as a consequence of the various measures instituted to curb the transmission of SARS. The two patients with acne vulgaris reported here are representative of several seen during and after the SARS period.

Acne vulgaris is a self-limiting, multifactorial disorder affecting the sebaceous glands and pilosebaceous follicles. It is a common problem in Singapore. There are several clinical subgroups of acne,[3] including tropical acne. This is a well-known entity which occurs in hot and humid climates. It is particularly common in soldiers[4] and affects mainly the trunk and buttocks.

In the two patients reported here, there was localized exacerbation of acne on the part of the face covered by the N95 mask. During the SARS period, N95 masks were recommended by the CDC and World Health Organization (WHO) for use when healthcare workers came into contact with confirmed or suspected SARS patients. They provided at least 95% filtration against oil-free particles and needed to be worn tightly against the face to be effective. It was not surprising to see acne occurring in the regions of the face covered by the masks. Donning of these masks over prolonged periods of time creates a humid "tropical" skin microclimate conducive to a flare-up of acne. Alternatively, the flare-up could have been a consequence of simple pilosebaceous duct occlusion due to local pressure on the skin from the close-fitting masks.

Apart from the risk of contracting SARS, the disease also affected healthcare workers in other physical and psychologic ways.[5] Acne vulgaris resulting from the wearing of tight-fitting masks over prolonged periods of time was one of the physical hazards. We will no doubt continue to see this condition as long as we need to take precautions to prevent another SARS epidemic.

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