Dr. Baskar Ambady Oration 1979

POROKERATOSIS AND DISSEMINATED ACTINIC POROKERATOSIS

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Permit me to thank you for the honour you have bestowed upon me by giving me the opportunity to deliver Dr. Baskar Ambady Memorial Oration. Dermatologists everywhere and particularly members of the Indian Association of Dermatology Venereology and Leprology have been saddened by the death of Dr. Baskar Ambady.

Dr. Ambady is remembered for his dynamism as a dermatologist and administrator. After qualifying for M.B B.S. from the Madras Medical College and after having served the army and undergoing training in various international institutions, he was the pioneer in starting dermatology and venereology departments of the Medical College, Trivandrum. He was the founder member of the Indian Association of Dermatology, Venereology and also the president of the association. No one can forget the Indian Association of Dermatology, Venereology and Leprology meeting held at Trivandrum. It was characteristic of Dr. Ambady that he disliked all pomp. He had an international outlook and we remember him for his generosity of spirit. He emphasized the value of fostering personal relationship with dermatologists everywhere. He has published seven papers of which some have been reviewed in foreign journals. His papers were on psoriasis, pemphigus, syphilitic peripheral vascular disease, yaws in Trivandrum, herpes zoster and chickenpox, penicillin resistant gonorrhoea and non gonococcal urethritis. He was vice president of the Foot ball Association, president of the Weight lifters Association and Wrestling Association.

When I was the District Governor of Lions International, District 304 South I had the unique privilege of knowing Lion Dr. Ambady as the president of the Lions Club of Trivandrum in the year 1963-64. He was a great man.

"Nasti Tesham Yasah - Kaye
Jara, Maranajambhayam."

Great men who dedicated their lives for service to society have no fear of old age or death. They become immortal and emancipated.

Dr. Ambady showed a keen interest in the study of porokeratosis. Hence I have chosen this subject for Dr. Ambady's Oration.

In this communication five cases of porokeratosis Mibelli and cases of D.S.A.P. are reported. Their clinical features and histopathology are discussed and review of literature is presented.

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Definition

Porokeratosis Mibelli is a well recognised chronic persistent genodermatosis with hyperkeratosis, excrescence with

festooned border on the margin and an atrophic centre.

Males are more commonly affected than females. Several pedigrees show simple dominant mode of inheritance.

Epidemiology

The disease may occur at any age but usually appears in childhood and persists indefinitely.

Synonyms

Porokeratosis is also known as Hyperkeratosis Excentrica (Resphigi) and Keratoderma Excentrica.

Etiology and pathogenesis

This condition occurs as an autosomal dominant trait. Mibelli[1] believed that the lesions develop due to a disorder of keratinisation in acrosyringial portion of the eccrine ducts and hence gave the name porokeratosis, which is probably a misnomer. Lever[2] states that the cornoid lamella seen in histological sections may be in relation to hair follicles or independent of any cutaneous appendages. Furthermore, porokeratosis may occur on the mucus membrane, glans penis and other areas devoid of eccrine sweat glands. Saunders[3] showed a histochemical similarity between granules of cornoid lamella and eccrine sweat glands but Chernosky & Freeman[4] found the granules to contain D. N. A., R. N. A. and P. A. S. positive diastase resistant materials identical to those of parakeratotic nuclei.

Clinical manifestation

Lesions can occur on the dorsa of feet, hands, face, neck, trunk and genitalia. The primary lesion is a keratotic papule with central depression or horny plug (Fig. 1). The papule slowly enlarges centrifugally in an irregular circinate fashion. The periphery of the plaque is a distinct hyperkeratotic ridge usually 1 to 10 mm in height, grey or brown and contains a shallow longitudinal groove and numerous keratotic plugs. Hairs are usually absent. Although the dry centres are atrophic, they sometimes contain areas of hyperkeratotic thickening.

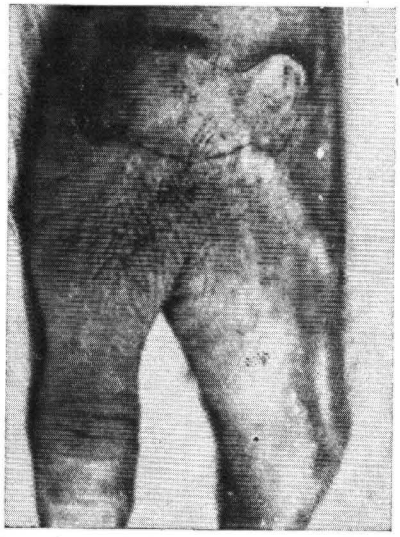

Fig. 1 Keratotic lesion with central atrophy

Histopathology

The histological feature essential for diagnosis of porokeratosis is the cornoid lamella which corresponds to the sharply defined margin of the lesion grossly and forms annular ring at the periphery of the lesion. Histologically, it is a column of lighter staining stratum corneum containing parakeratotic cells beginning in the malpighian layer and extending upward through the granular and keratin layer. The malpighian layer at this point may be either atrophic or slightly acanthotic. The granular layer is absent beneath the cornoid lamella. A few isolated dyskeratotic cells are usually seen in the malpighian layer beneath the cornoid lamella. In addition some of the malpighian cells at the base of keratin layer are vacuolated. A focus of lymphocytes and histiocytes are present beneath the cornoid lamella in the dermis. Within the centre of the ring

formed by cornoid lamella most lesions present atrophic thin malpighian layer with effacement of rete ridges and varying amount of atrophy of the dermis and chronic inflammatory infiltration (Fig. 2).

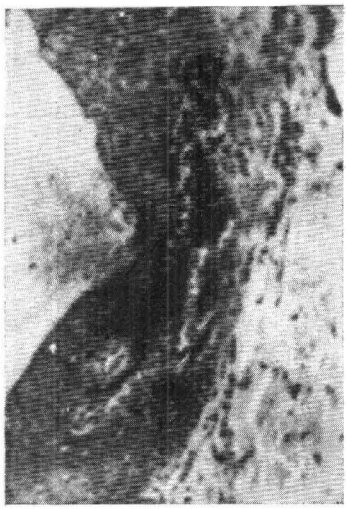

Fig. 2 Shows effacement of rete ridges

Case Reports

Five patients belonging to one family having porokeratosis since their 18th year of life were reported from our Dermatology Centre by Venkatesan[5] in the year 1965.

Three were brothers, their ages being 51, 48 and 42. The fourth case aged 22 was son of number 1 and 5th case aged 18 was son of number 2. (Fig. 3, 4, 5). In all these cases skin biopsy was done and the diagnosis of porokeratosis Mibelli was confirmed.

Fig. 3 Shows lesion in 51 year old man on dorsum of hand.

Treatment

In 2 cases lesions were surgically excised. In 3 cases they were destroyed by electrodessication.

Nine cases of disseminated superficial actinic porokeratosis were seen during the period 1966-1974 in our Dermatology Centre, Madurai. Their clinical features are summarised in Table 3.

Among the nine cases only one was female. Family history was available in three cases of which one gave history of a similar condition in maternal aunt. Four patients complained of pruritis.

TABLE 1

Showing age of patients, number of lesions and distribution

Case	Age	Approximate No of lesions	Extremities Upper	Extremities Lower	Trunk	Face	Neck	Bilateral
1	51	2	×	—	—	—	—	—
2	48	1	—	×	—	—	—	—
3	42	2	×	—	—	—	—	×
4	22	2	—	—	×	—	—	—
5	18	1	×	—	—	—	—	—

TABLE 2
Comparison Chart

	Porokeratosis Mibelli	D. S. A. P.
Age of onset	Any age, but usually childhood	Not in childhood. More common in 3rd decade.
Pruritis	Usually none	Usually in one third of cases
Number of lesions	One or very few	Large numbers
Distribution	Variable including areas not exposed to sunlight	Sun exposed areas
Size of lesion	Large	Small
Heigh of keratotic border	Usually 1 to 10 mm	Usually less than 1 mm
Presence of furrow in the keratotic border	Frequent	Not observed
Histopathology	Cornoid lamella the prominent feature	Cornoid lamella not a prominent feature

All patients gave history of exacerbation after exposure to sun. In all cases skin biopsy showed the presence of cornoid lamella.

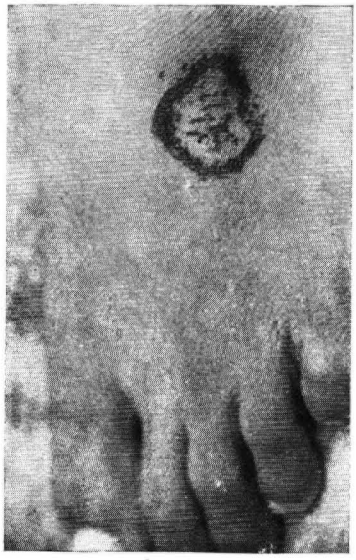

Fig. 4 Shows lesion in 48 year old man on dorsum of foot

Several authors have described porokeratosis in the mouth, genitalia, conjunctiva and anus. Butterworth[8] claims that lesions occur in the mucus membranes in one of the cases. None of our cases showed mucus membrane lesions.

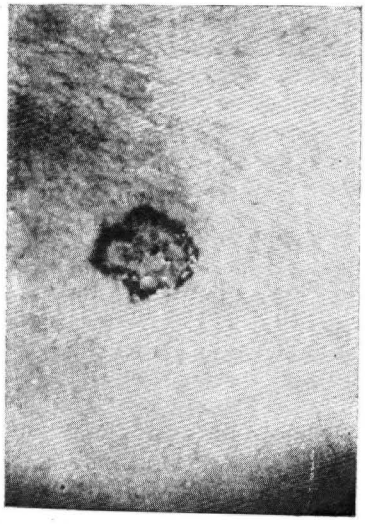

Fig. 5 Shows lesion in 22 year old man on back of trunk

D. S. A. P. is not a rare condition It is a genodermatosis with an autosomal dominant mode of inheritance[6] Exacerbation of the lesions of D.S.A.P. occurs following natural or experimental U. V. R. light exposure. Lesions are frequently asymptomatic but sometimes produce significant cosmetic problem. About one third of patients complain of itching. 50% of patients show exacerbation during summer. The early or primary lesions are tiny

conically shaped papules 1 to 3 mm in diameter usually follicular lesion topped by a small keratotic plug (Fig. 6).

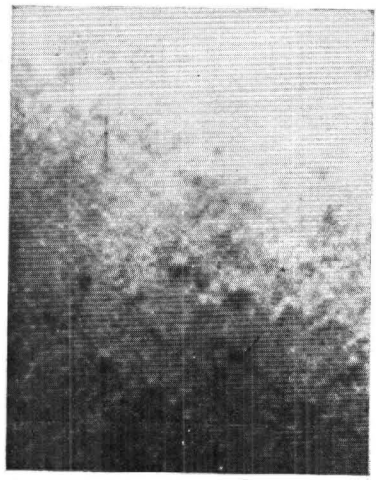

Fig. 6 Early follicular lesion

This plug sometimes falls out leaving a minute central depression. The papule then enlarges centrifugally leaving a somewhat depressed and atrophic area surrounded by a slightly raised hyperkeratotic sharply defined ridge. The central area remains pigmented or erythematous and is frequently separated from the border by a hypopigmented zone. The primary papules and enlarging keratotic plaques resemble lesions of porokeratosis of Mibelli except for their minute sizes and large numbers that develop bilaterally on sun exposed areas particularly on extensor surfaces of the extremities (Fig. 7). Keratotic papules with delling are present in some, giving an appearance similar to lichen sclerosis et atrophicus. Longitudinal furrow has not been observed.

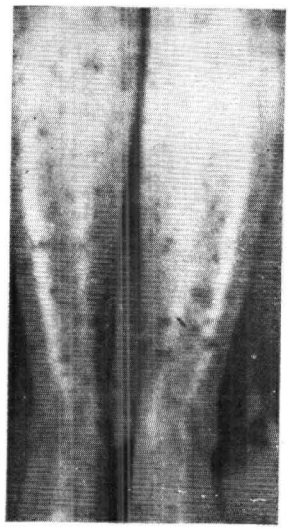

Fig. 7 Lesions on exposed areas of legs

Differential Diagnosis

1. Actinic Keratosis : This is seen commonly in the third decade in predisposed persons who get excessive sun

TABLE 3

Showing clinical features of 9 cases of DSAP

Case No.	Approximate number of lesions	Duration	Age	Sex	Extremities Upper	Extremities Lower	Trunk	Face	Family history	Pruritis
1	26	11 Yrs.	36	M	×	×	—	—	—	×
2	31	14 Yrs.	44	M	×	×	×	×	One maternal aunt	×
3	14	21 Yrs.	51	M	×	×	×	—	Daughter Case No. 4	×
4	7	2 Yrs.	15	F	×	×	—	—	Father Case No. 3	×
5	16	8 Yrs.	37	M	—	—	×	—	—	—
6	27	4 Yrs.	31	M	×	—	—	—	—	—
7	13	3 Yrs.	28	M	—	—	×	—	—	—
8	6	1 Month	19	M	×	—	—	—	—	—
9	12	10 Yrs.	40	M	×	—	×	—	—	—

exposure. It is usually associated with freckled pigmentation and yellow furrowed skin representing actinic elastosis. The keratosis may vary from skin colour to yellow brown or black depending upon the amount of adherent horny material, which is difficult to remove.

Histologically the epidermal changes are characterised by acanthosis, dyskeratosis and a downward proliferation of rete cells arranged in a disorderly manner. The histological picture may at times resemble Bowen's disease.

2. Seborrhoeic keratosis : These are multiple slightly raised light brown to black, sharply defined lesions that are located mostly on the seborrhoeic areas namely face, neck and upper part of chest. The nummular warty lesions are covered with greasy crust which is loosely attached. When the crust is removed a raw pulpy base is revealed. Histologically seborrhoeic keratosis shows keratin cysts coalescing to form crypts.

3. Xerosis.

4. Psoriasis : This shows typical clinical features and also histological pictures of micro pustules in the epidermis, elongation of rete ridges, acanthosis, reduced or absent granular layer and para keratosis.

5. Lentigenosis : This shows uniform brown to black lesions anywhere on the body and mucosa. Histologically there is an increase in the number of normal melanocytes just above the basement membrane.

6. Lichen sclerosis et atrophicus can occur on the skin or mucus membrane as white plaques or papules with cigarette paper wrinkling on the surface and telangiectasia.

Biopsy shows hyperkeratosis, oedema and loss of bundle structure in collagen and disappearance of elastic fibres. The affected zone shows dilated capillaries. Below the affected zone there are perivascular infiltrate of lymphocytes or plasma cells.

7. Lichen planus : In this there are typical flat topped papules with lilac colour. The hypertrophic variety are verrucous lesions. Histologically there is thickening of the granular layer, liquefaction degeneration of the basement membrane and basal cells and band like lymphocytic and histiocytic infiltration in the upper dermis.

8. Keratosis pilaris : Keratotic papules distributed diffusely are seen. The lesions are most common on the lateral aspect of the upper arms, thighs and buttocks. The histological picture consists of a follicular keratinous plug and a paucity of inflammation in the dermis.

9. Acrokeratosis verruciformis : The lesions consist of verrucous and lichenoid papules 1 to several mm. in size and occur predominantly on dorsa of hands and feet. Histopathologically the papule shows an increase in thickness of the statum corneum and granular layer, acanthosis and slight papillomatosis.

In all these above mentioned conditions, the keratotic wall and cornoid lamella are absent.

Histopathology

The hstological features of D.S.A.P. are essentially the same as those of porokeratosis Mibelli. The cornoid lamella however is insignificant. Recent histological studies by Reeds and Leone[7] point out that the advancing abnormal dyskeratotic and oedematous epidermal cells beneath cornoid lamella curve below adjacent normal epidermis causing it to buckle. These abnormal cells are referred as a mutant clonal keratosis of the epidermis.

Treatment

The method of management in our centre is as follows :

(a) Intralesional injection of triamcinalone 10 mgm, once in 15 days for six sittings for lesions less than 15. If number of lesions are more, number of sittings are increased proportionately upto about 10 patients. Lesions persisted one year after treatment, it was considered a failure.

Out of nine cases, four failed to respond. Of the four cases two cases were tried on 5 FU and showed partial clearance. Some workers have advocated large dose of Vitamin A, retinoic acid externally, thyroid extract and Cryo therapy (Solid carbondioxide dipped in acetone). We have not used any of these in our centre.

Comments

1. The conditions are of genetic disorder.
2. Both Porokeratosis Mibelli and D.S.A.P. are rare.
3. Porokeratosis is a keratotic papule with a prominent raised border with longitudinal groove at the border.
4. D. S. A. P. resemble porokeratosis but there is no longitudinal groove.
5. Biopsy in all cases show the histological picture of cornoid lamella.
6. Management – No 100% success. Intralesional Triamcinalone 10 mgm. 50% success.

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