POROKERATOSIS

THESIS

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BY

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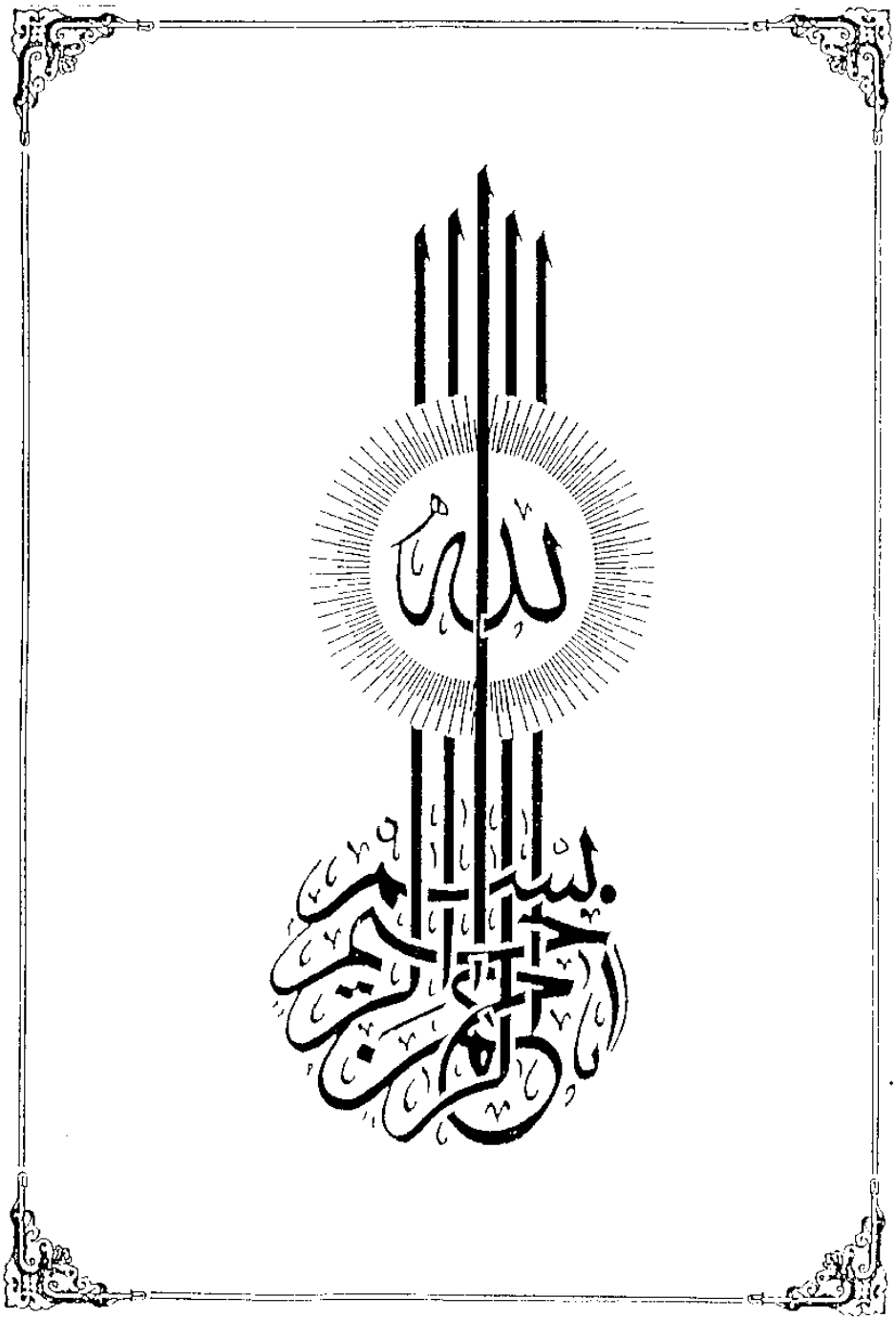

A C K N O W L E D G M E N T

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A. MOHSEN IBRAHIM.

TO MY MAJOR MOTIVATORS: MY

Wife And Our Son, Islam .

P O R O K E R A T O S I S

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I N T R O D U C T I O N

INTRODUCTION

Porokeratosis is a genodermatosis which consists of one or more keratotic plaques surrounded by a distinct raised border of epidermal proliferation with characteristic histological findings - the cornoid lamella - which consists of column of parakeratotic cells filling an epidermal invagination. It was thought that the initial lesion occurs over the orifices of sweat ducts as hyperkeratosis of their outlets, hence the name "porokeratosis".

But it is obvious now that porokeratosis is a misnomer because the characteristic changes do not necessarily initiate in, or remain confined to pores. It has been suggested that the lesion consists of abnormal clones of epidermal cells.

Mibelli and Respighi independently described classic porokeratosis in 1893. Reports of this uncommon skin disease have appeared sporadically since then. More recently a number of clinical variants largely based on distribution patterns have been recognized. The classic form consists of large annular keratotic plaque with a thread like furrowed border appearing on the extremities and face. In various forms, smaller more

numerous lesion develop either in a generalized distribution or in forms limited to sun - exposed skin, palms and soles or in a linear configuration.

The aim of this work is to review thoroughly the subject of porokeratosis as regards its epidemology, aetiology different clinical variants, histopathology and pathogenesis, differential diagnosis, complication, and the various lines of treatment which can be performed.

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REVIEW OF LITERATURE

HISTORICAL REVIEW

Although Majocchi in Italy was the first to recognize porokeratosis as early as I883, naming it "linear and hystrix ichthyosiform dermatosis", his successor Mibelli described the same condition in such detail in I893, that it bears his designation "porokeratosis". Independently Respighi in I893 described the same condition and named it "hyperkeratosis figurée centrifuge atrophicante". Again Respighi in the same year gave this disorder another name"hyperkeratosis excentrica". Miescher in I94I suggested the term of"Parakeratosis centrifuga atrophicans" as more appropriate than porokeratosis. Hutchins in I896 was the first to report a case of porokeratosis in America. The following cases from America were those of Gilchrist who in I897 reported a group of eleven cases, Since then, reports from all parts of the world began to appear in the literature. In I966 Chernosky recognized a more common type, now called disseminated superficial actinic porokeratosis(DSAP).

In I97I, Guss et al reported eight patients who represented a third type designated as porokeratosis plantaris palmaris et disseminata(PPPD).

EPIDEMOLOGY AND AETIOLOGY

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Epidemology:

The incidence of classic porokeratosis of Mibelli(PM) and PPPD is rare, while that of DSAP is not rare.

PM occurs at any age usually at childhood but DSAP occurs at the third to the fourth decade while PPPD occurs between 18 - 22 years in the reported cases(Chernosky,1979).

As regard the sex incidence it was found that PM is more in males than in females, the ratio is two to one to three to one. DSAP is equal in both sexes while that of PPPD is double in the males than females.

PM has been observed in all races and all parts of the world (Sanderson and Mackie,1979).

There are reports of the occurence of porokeratosis in negroes(El Nofty, 1956, Seif El Nasr et al, 1962) and Orientals(Hall, 1928). Chernosky in 1979 stated that all reported patients with PPPD were white.

DSAP is reported more in geographical areas where sun exposure is intense especially on lighter coloured skin.

Aetiology:

Inspite of many investigations, the causes of porokeratosis remains a mystery. Many theories were put forward for the origin of this disorder. It has been suggested that porokeratosis has a relation to thyroid dysfunction, vitamin A deficency, verrucae, infection, seasonal variation and actinic radiation. Others suggested that the condition to be of a naevoid origin or of having a familial and genetic background.

Thyroid Dysfunction:

In I927, Acton believed that porokeratosis was merely a type of localised hyperkeratosis with central atrophy associated with hypofunction of the thyroid gland. Acton reported complete cure in his three cases within two weeks after thyroid medication was instituted. This observation was not substantiated by most other workers.

Vitamin A Deficency:

Bloom and Abramowitz in I943 regrded porokeratosis as an hereditary dyskeratosis and suggested that vitamin A deficency might be of aetiological importance.

Haber and Porter(I95I) mention that there is a possibility of

a defect in the metabolism of vitamin A in porokeratosis and it is associated with a gene transmitting porokeratosis.

Relation To Verrucae:

The warty appearance of the porokeratotic lesions especially in their early stage, the absence of any nevus cells in the histological sections studied, the histological picture of hypertrophy of the epidermis and papillae and well marked hyperkeratosis, all these features favored the assumption of a probable relationship to the group of verrucae(Seif El Nasr et al, 1962).

Infective Theory:

Many attempts were done to reproduce porokeratosis by inoculation of affected tissue, both in human beings and animals. One of these attempts had been made by wende in 1898 who noticed, after many attempts a typical lesion developed at the site of inoculation on the unaffected hand of a patient suffering from porokeratosis. Ritchie and Becker in 1932 recorded development of porokeratosis through inoculation of the skin of a guinea-pig.

The centrifugal spread, followed by atrophy and recurrence of