ATROPHODERMA OF PASINI AND PIERINI

THESIS

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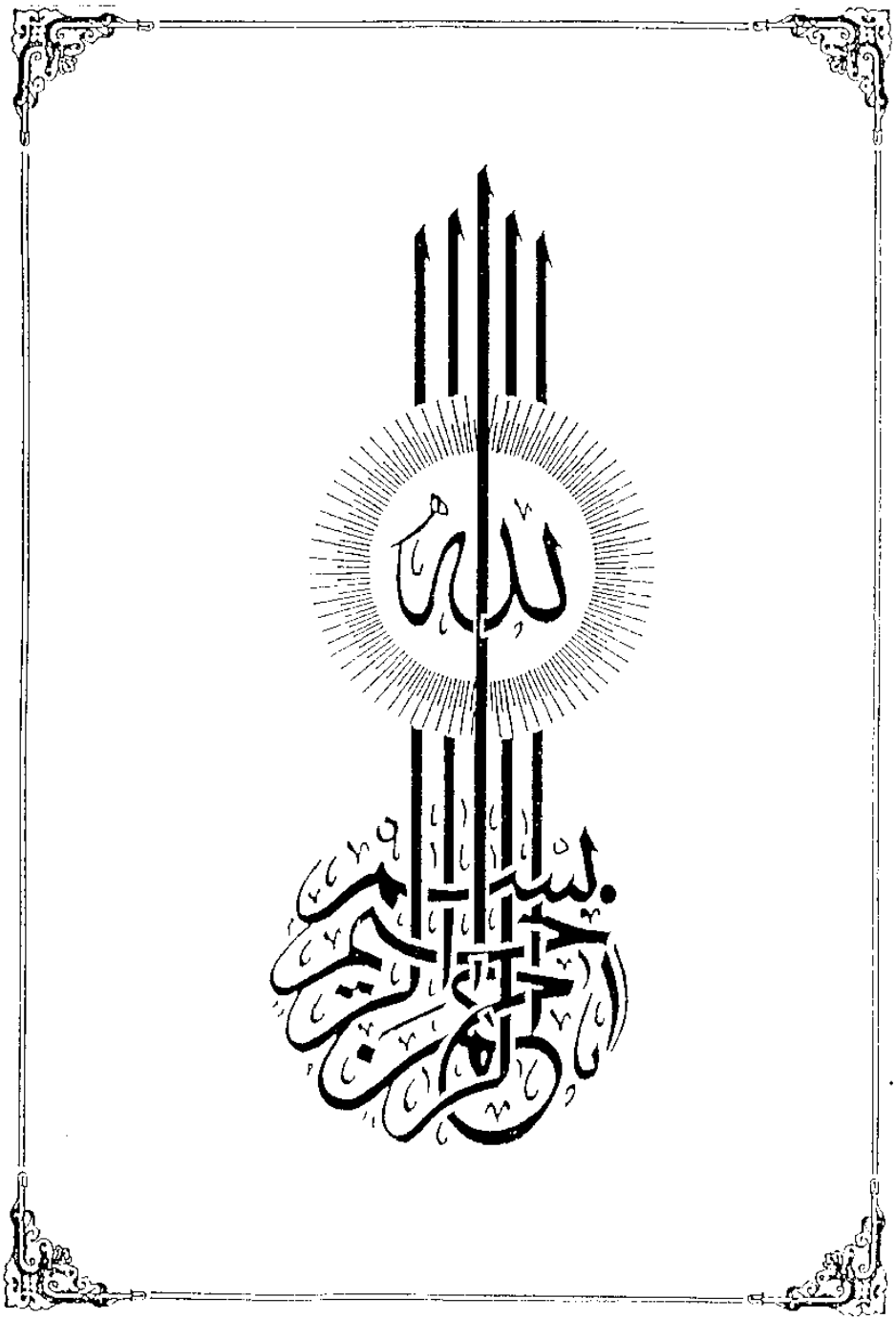

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INTRODUCTION

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The study of the cutaneous atrophies is one of the many obscure chapters in dermatology.

In this thesis I shall discuss a peculiar cutaneous atrophy described by Pasini in 1923 under the title of progressive idiopathic atrophoderma, which has not received enough attention. This fact made the disease more or less obscure. Not only the etiology and treatment are problems, but also to put the disease into the proper classification (nosological status) is not easy. It is still not clear whether atrophoderma of Pasini and Pierini is a distinct disease entity or a condition comprising various types of skin atrophies and atrophic varieties of scleroderma.

Generally speaking, there is no agreement as to either the nature of atrophoderma of Pasini and Pierini or its pathogenetical connection with scleroderma on the one hand, and other idiopathic skin atrophies on the other.

In a trial to throw light on this disease the gathered knowledge from literature has been presented in this work to make a review for such disease.

This work includes review of the cases reported under the name of idiopathic atrophoderma of Pasini and

Pierini in the form of the historical review of the disease, the etiology, clinical features, differential diagnosis, treatment and the nosological status of the disease.

HISTORICAL

REVIEW

HISTORICAL REVIEW

Idiopathic atrophoderma had been known earlier under various names, as superficial primary atrophic scleroderma, atrophic scleroderma "d'emblee", dyschromic and atrophic type of scleroderma and morphea plana atrophica (Brocq and Civatte, 1902; Jeanseleme, 1903).

As early as 1908, Nobl., under the name of "association of idiopathic atrophy of the skin and circumscribed scleroderma", reported a girl, aged 14 years, with two patches on her back. One was soft, blue violaceous and thin. The other was hard, yellowish, and shiny. He concluded that the same cause may lead in one person to lesions of scleroderma and idiopathic atrophy.

The earlier report on the disease and its clinical features was done by Pasini (1923). Under the title of "Progressive idiopathic atrophoderma", he reported a case of a young woman, aged 21 years, with a peculiar cutaneous atrophy involving the trunk. His description for the dermatosis consisted of large plaques of cutaneous atrophy, essentially characterized by thinning and depression of the skin. A sharp depression of the skin separated the affected area from the surrounding normal skin with absolute absence of erythema or other inflammatory changes. The patches were depressed 1 to 2 mm. and were smooth and

flat, apparently with normal keratinization, and showed no evidence of sclerosis. There was an apparent decrease of the follicular openings. The elasticity and consistency of the skin was well preserved. His description revealed a peculiar form of atrophoderma that was clinically and histopathologically different from the atrophic stages of localized scleroderma or any other known atrophy.

Pierini and Vivoli (1936) reported a case of a woman, aged 19 years, with lesions of three years duration on the right side of the chest and back, extending in a zosteriform distribution. Two years later new lesions appeared on the abdomen and thighs, mostly on the right side. The lesions were nonindurated bluish violaceous patches typical of idiopathic atrophoderma.

Pierini and Bocq (1941) reported the third case of idiopathic atrophoderma. The patient was a 28-year-old man with a lesion of nine years duration on the right side of the back. It was a single, depressed buish plaque, 5 cm. long, without superficial atrophy, erythema, or telangiectasia.

Pierini and Sanchez Basso (1944) reported a woman, 34-years-old with large lesions on the back, flanks, and upper aspect of the thighs, probably of several years duration. All patches showed the typical bluish-slate colour and were noninfiltrated.

Cordero and Noussitou (1946) reported a case of a 23-year-old man with many depressed bluish patches on his back. Two of these, after four years, developed whitish areas in the center. The others remained unchanged.

Pierini and Pierini (1952) reported a boy, aged 14 years, with lesions of three years duration on the right forearm and trunk. All lesions were depressed, slate-coloured patches, typical of idiopathic atrophoderma.

Borda (1952) reported a man, aged 56 years, with two patches on the anterior aspect of the thighs and back which were clinically and histologically suggestive of idiopathic atrophoderma.

Grinspan et al. (1953) reported two patients with lesions resembling the clinical features of idiopathic atrophoderma, secondary to localized scleroderma. The first one was a boy, 10 years old, with bilateral and symmetrical eruption of four years duration, localized to the anterior aspect of the abdomen. One of the lesions was palm-sized and presented at its upper part the characteristic clinical and histological appearance of idiopathic atrophoderma, while at its lower part, it presented the clinical induration and ivory color of a morphea. The second case was seen before by Dr. Pierini and Dr. Borda, and observed by them for a period of 12 years. The diagno-

sis of morphea and idiopathic atrophoderma had been considered.

Grinspan and Zurita (1953) reported two cases of idiopathic atrophoderma. One was a youth 20 years old who presented with lesions of six years duration on the right side of the back. The other, a youth aged 17 years, presented a violaceous patch with telangiectasia, which followed by induration. Histologically, the findings of both cases suggested a scleroderma in its atrophic phase.

Pomposiello (1953) reported two cases of idiopathic atrophoderma confirmed by microscopic examination. In one case there were lesions on the back, dorsal aspect of the hands, and extensor aspect of the forearms. The other patient presented with well-defined depressed bluish patches of four years duration on the back. The same author, in 1954, reported a case with two slate-coloured, depressed, atrophic patches. One lesion presented a whitish indurated center which, histologically showed the appearance of scleroderma.

Borda and Abulafia (1956) reported an interesting case that showed lesions of idiopathic atrophoderma, scleroderma "en goutte" and lichen sclerosus et atrophicus. The patient was a 38-years old man, with lesions of 15 years duration. Some of the lesions were so superficial that they clinically resembled vitiligo. The authors suggested

that there was a close relationship between the idiopathic atrophoderma, scleroderma "en goutte", and lichen sclerosus et atrophicus.

Canizares et al. (1958) were the first to call the disease "idiopathic atrophoderma of Pasini and Pierini". In their article, they pointed out the importance of the clinical picture presented by Pasini (1923) and they considered the contribution done by Pierini (1936) to the knowledge of this condition to be so valuable that, it should be called after their names. So the diasease was called "idiopathic atrophoderma of Pasini and Pierini", and known by that name until now, after them. In their article they described five cases of idiopathic atrophoderma of Pasini and Pierini, two male and three female patients. The onset of the lesions was in the teens or early twenties. One patient showed an incipient diabetes. The lesions were on the trunk, always affecting the back. They were asymptomatic in four cases; one complained of occasional tingling or sensation of warmth. The course was extremely slow and progressive and ranged from 4 to 15 years. There was an apparent spontaneous arrest in two cases, but no complete involution of the lesions in any case. If sclerodermatous changes appear, they developed years later within the patches of atrophy or simultaneously in separate lesions.

Weiner and Gant (1959) presented two brothers with atrophoderma of Pasini and Pierini. Their ages were 17 and 19 years respectively. They complained of brown patches on the trunk. The clinical and histological pictures confirmed the diagnosis. They suggested the possibility of hereditary transmission of the disease.

Piraino (1959) reported a 39-years-old white male complaining of discoloured patches on the back of two years duration. Examination revealed round and oval, slightly depressed, bluish-violaceous macules, especially located on the back, chest, and inguinal areas. The lesions varied considerably in size, and there was no real atrophy. The deeper vessels seemed to be visible. The diagnosis was confirmed by the histological examination.

Kee et al. (1960) reported a case with idiopathic atrophoderma of Pasini and Pierini with coexistent morphea. The case was a white man aged 22-years-old, complaining of asymtopmatic dark depressed areas of the skin on his back. The author discussed whether this condition is an entity or just a variation of scleroderma.

Barker (1960) reported a woman, aged 32 years, with two brownish coloured patches on the back. The picture was clinically and histologically suggestive of idiopathic atrophoderma of Pasini and Pierini.