SKIN MANIFESTATIONS OF RHEUMATIC DISEASES

THESIS

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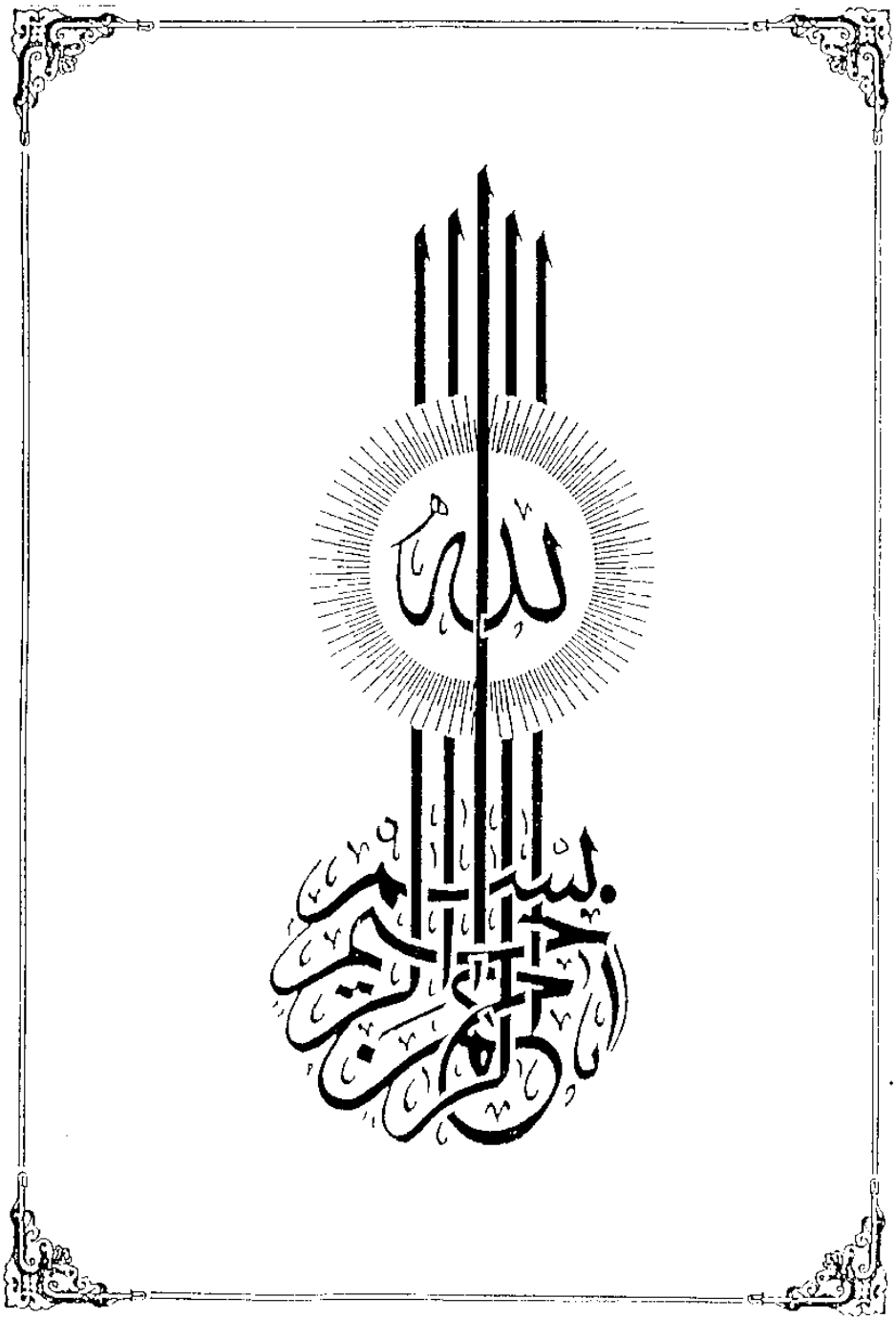

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INTRODUCTION AND AIM OF WORK

Introduction:

Careful examination of the skin can often be extremely helpful in the differential diagnosis when a patient presents with arthritis. Conversely, certain cutaneous disorders may alert the physician to the possibility of joint disorders presenting at a later date.

Pathologic process in many of the rheumatic diseases is rather systemic involving different organs in the body; some of which produce fatal complications. Indeed, it would be mostly beneficial to predict these complications by examining the skin. Hence, a control of the whole pathologic process can be accomplished.

While some skin lesions are specific, many are non specific and occur in several rheumatic diseases. Dermatologist as well as orthopedicians should be aware of these cutaneous manifestations and their individual significance.

Some types of arthritis are characterized by certain pathologic and clinical features which make their differentiation relatively easy. In other forms of joint diseases, the clinical pattern varies considerably, and identification may depend on detailed studies including examination of the skin.

Aim of the Work:

The aim of this work is to review the literature concerning the cutaneous manifestations of rheumatic diseases and other allied disorders. This study will lay some light on clinical data of these skin lesions in order to expand our experience and spot some important items that would help in the differential diagnosis.

Among the enormous scope of skin lesions associated with arthropathies, concentration will be conducted on the more important conditions in which skin lesions and joint diseases are associated.

Review of Literature

RHEUMATIC DISEASES

Hollander (1979) defined rheumatic diseases as those conditions in which pain and stiffness of some portion of the musculoskeletal system are prominent. These include diseases of connective tissue. The author also considered "arthritis" as a general term used when the joints themselves are the major seat of the rheumatic disease.

Until the etiology of all types of arthritis is known no grouping of joint diseases can be accurate. As additional knowledge is gained in the field of rheumatology, changes in terminology and classification will be made accordingly (Hollander, 1979).

Hench (1948) designed five major groups comprising most cases of arthritis. These groups are:

1. The frankly infectious cases caused by a specific micro-organism.
2. Cases that are possibly infectious but of unproved etiology.
3. Cases representing degenerative forms of joint disease, which are sometimes termed "arthroses".
4. Cases in which the arthritis results from direct trauma to the joint.
5. Cases of metabolic arthritis.

Many other classifications of diseases of joints and related structures have been suggested. All have certain disadvantages. For the sake of simplification. clarity,

and unification of terminology. Blumberg et al. (1964) cited the classification that was tentatively approved by the American Rheumatism Association in 1963. Although this classification included the whole list of arthritis, the following citation will only present the major headings and some examples of diseases that are known have skin lesions. Cutaneous manifestations of these diseases will be discussed later.

+ Citation of "Nomenclature and Classification of Arthritis and Rheumatism" *

I. Polyarthritis of unknown etiology

e.g. A. Rheumatoid Arthritis

B. Juvenile Rheumatoid Arthritis (Still's disease)

C. Psoriatic Arthritis

D. Reiter's syndrome

II. "Connective tissue" disorders

e.g. A. Systemic Lupus Erythematosus

B. Scleroderma

C. Others e.g. Mixed Connective Tissue Diseases

III. Rheumatic Fever

IV. Degenerative Joint Disease

V. Non-articular Rheumatism

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+ Examples written in each group in this citation are by no means a complete listing of the original classification that was presented by the author. Rheumatic diseases that are known to be associated with skin lesions are only mentioned.

* Blumberg et al. (1964)

VI. Diseases with which arthritis is frequently associated

VII. Associated with known infectious agents

VIII. Traumatic and/or neurogenic disorders

IX. Associated with known biochemical or endocrine abnormalities

e.g. A. Gout

B. Ochronosis

X. Tumour and tumour-like conditions

XI. Allergy and drug reactions

XII. Inherited and congenital disorders.

XIII. Miscellaneous disorders

e.g. Behcet's syndrome

RHEUMATIC FEVER

The criteria for guidance in the diagnosis of acute rheumatic fever were initially proposed by Jones (1944). Clinical and laboratory criteria were divided into major and minor categories according to the diagnostic importance of a particular finding. The presence of two major criteria, or of one major and two minor criteria, indicates a high probability of the presence of acute rheumatic fever. Among the major criteria are the two, well documented, skin manifestations of the disease; subcutaneous nodules and erythema marginatum (American Heart Association, 1984).

In addition to the two previously stated skin lesions, Burns et al. (1964) recorded the rare occurrence of cutaneous rheumatic papules with rheumatic fever.

1. Subcutaneous Rheumatic Nodules

The occurrence of subcutaneous nodules in patients undoubtedly suffering from rheumatic fever was recorded for the first time by Wells (1812).

Merritt (1928) noted that the nodules were most often found in patients with chronic forms of rheumatic fever with recurrent infections and severely damaged hearts. They usually appear several weeks or even months after the onset of the attack and are most common in patients in whom signs of activity of the rheumatic process continue for several weeks or months (Baldwin et al., 1960). As stated by Lowney and Simons (1963) nodules may also appear as a sole manifestation of rheumatic activity in patients who

previously have had definite attacks of rheumatic fever. It seems reasonable that occasionally a subcutaneous nodule may be the first sign of rheumatic fever appearing months or even years before other manifestations of the disease. However, nodules disappear by the time other evidence of the disease appears.

Altman and Caffrey (1964) stated that subcutaneous nodules can occur in patients without other evidence of rheumatic disease. They reported a 3-year-old case of isolated rheumatic nodules with no manifestations of any systemic disease. Thus, the occurrence of these nodules does not necessarily carry the serious diagnostic and prognostic implications as was formerly thought (Lynch, 1981).

Pathogenesis:

The morphologic observations, as stated by Bennett et al., (1940) do not permit conclusions concerning the nature of the agent or agents causing these nodules. They recorded that microorganisms were never observed in any of the appropriately stained sections. Cultural methods and animal inoculations likewise failed to disclose infection. Nevertheless, the observed pathologic changes could be the result of the activity of an infectious agent which to date has not been isolated. That such lesions might be caused by a chemical agent has also been suggested.

According to Keil (1938) the subcutaneous nodules arise from, and are attached to, the deeper structures,

such as tendon or tendon sheath, periarticular ligaments, superficial aponeurosis, and external layer of periosteum. They arise chiefly due to injury of small blood vesseles with subsequent exudation of plasma and blood cellular constituents into the connective tissues (Bennett et al., 1940). This latter observation was also recorded by Braverman (1970) and Lynch (1981).

Age Incidence:

Wallace (1924) stated that nodules were essentially manifested in children although they had been noted not infrequently in adults.

Baldwin et al. (1960) analysed a series of 101 separate rheumatic episodes of carditis accompanied by subcutaneous nodules which occurred in 96 patients. The nodules occurred in patients ranging in age from 3 to 13 years, with the highest incidence between age 7 and 9.

This age incidence is in accordance with what was recorded by Barnert et al. (1975) and Gilkes (1978). The latter author stated that the nodules were seldom seen in children under four years old and the peak age is six years.

Clinical Data:

Although pain, tenderness, itching and burning have been occasionally noted in connection with rheumatic nodules, they are, as a rule, totally innocent of producing any troublesome symptoms (Wallace, 1924; Canizares, 1957; and Persellin, 1978).