IMMUNOLOGIC STUDY OF CUTANEOUS VASCULITIS

Thesis
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By
Maha Hassan Anwar Aboul Magd
M.B.,B.Ch., M.Sc. (Dermatology and Venereology)

SUPERVISED BY

Professor Dr. Hafez Monamed Khafagy
Professor of Dermatology and Venereology
Faculty of Medicine - Ain Shams University

Professor Dr. Mona El Okby
Professor of Dermatology and Venereology
Faculty of Medicine - Ain Shams University

Professor Dr. Adly Farid Ghaly
Professor of Pathology
Faculty of Medicine - Ain Shams University

Dr. Samia Ammar
Assistant Professor of Pathology
Faculty of Medicine - Ain Shams University

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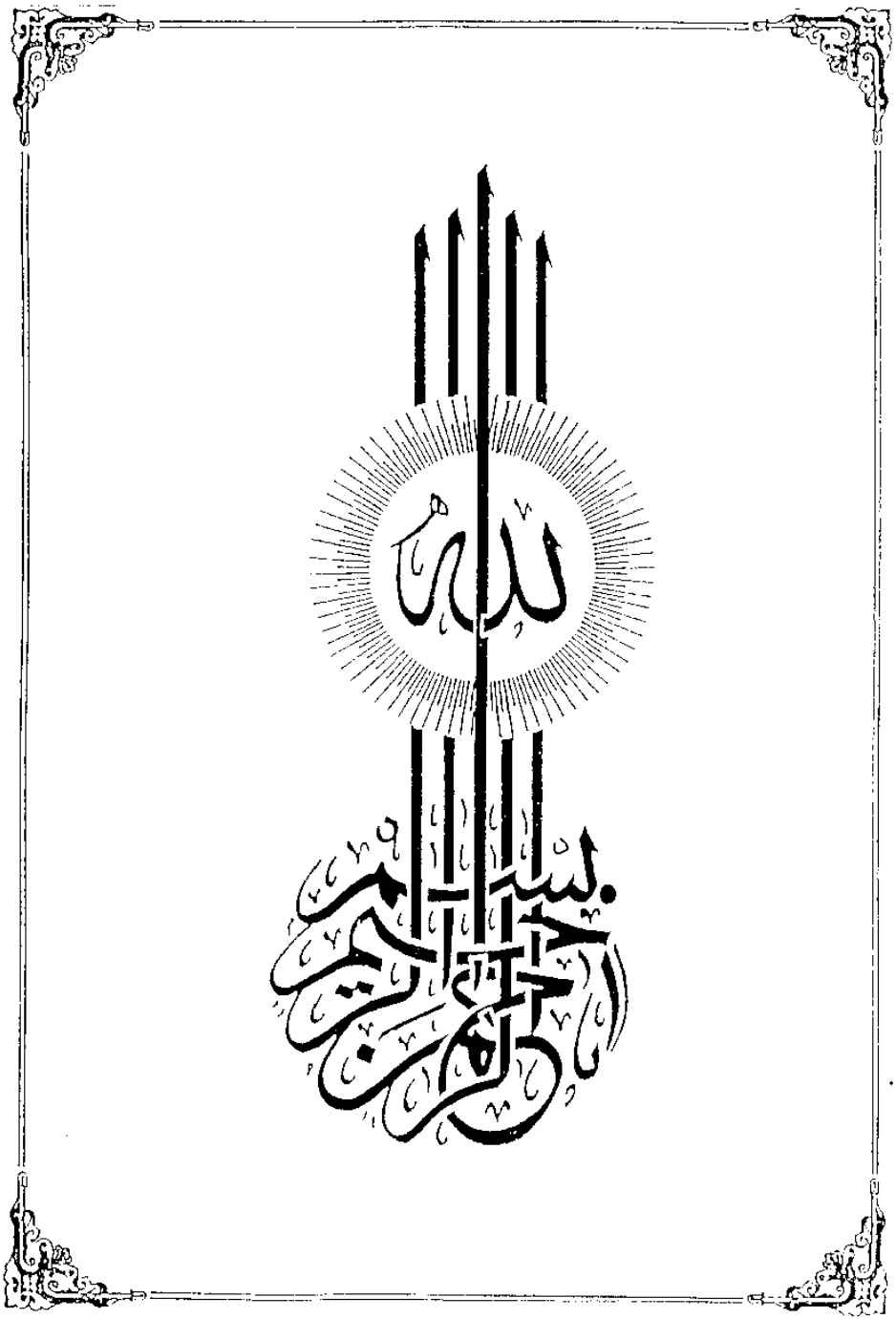
بسم الله الرحمن الرحيم

سُبْحَانَكَ لَا عِلْمَ لَنَا إِلَّا مَا عَلَّمْتَنَا إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ .

صدق الله العظيم



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AIM OF WORK

AIM OF THE WORK

The subject of vasculitis is a detailed and broad one.

Despite intensified efforts, research has yet to reveal the aetiology of vasculitis. The experimental findings are still in a state of disarray. Data however are accumulating, indicating that the immune system may be important in the pathogenesis of this multifactorial disease.

This work has been performed to study some parameters of the immune system (humoral and cell mediated) of patients with cutaneous vasculitis. A direct immunofluorescence study was done to search for immunoglobulins, complement and fibrinogen deposits in the skin lesions of vasculitis. It is hoped that this may bring us one step closer to our eventual understanding of the role of some immunological factors in this puzzling disease.

Review of Literature

DEFINITION AND NOMENCLATURE

The subject of vasculitis is a broad and detailed one including many varied aetiologies, clinical syndromes, classification systems and pathological descriptions.

The varying syndromes which have often been separated clinically are most likely manifestations of the same pathogenetic process immune-complex disease.

The literature on vasculitis during the period from 1886-1952 as reviewed by Zeek used the terms vasculitis, periarteritis nodosa nd arteritis synonymously with periarteritis nodosa being the most commonly used term (Callen et al., 1978). They also revealed that by the early 1950's, the term periarteritis nodosa was a broad one including necrotizing or hypersensitivity angiitis, polyarteritis, temporal arteritis, Wegener's granulomatosis, the allergic vasculitis of Churg and Strauss and others.

In 1970, Copeman and Ryan defined angiitis as an inflammatory change primarly in and around the vessel

wall but not secondary to embolus or inflammation in neighbouring tissues although adjacent tissues will be affected as a secondary manifestation as in the panniculitis of "nodular vasculitis".

Vasculitis or angiitis refers to the primary inflammatory process involving small and medium-sized blood vessels resulting in organic damage, namely necrosis, fibrinoid degeneration, hyalinization and granulomatous reaction (Koenigk, 1975). A more or less similar definition was revealed by Ashgar and Cormane (1974). They were of the opinion that the term vasculitis generally refers to conditions of which the primary pathological lesion is an alteration in and around the vessel wall. These alterations include vascular changes of thickening, endothelial swelling and infiltration of blood vessel wall by inflammatory cells such as eosinophils, neutrophils and extravasated red blood corpuscles.

The angiitis group of disorders has acquired a great number of synonyms and eponyms derived from their varied clinical appearance or on the basis of a supposed pathogenesis. In general, the term vasculitis is unsatisfactory and has no exact definition. It is an ill defined entity and if used alone includes all inflammatory reactions

around and within all blood vessels (Roenigk 1975).

Alarcon - Segovia (1977) described vasculitis as an inflammatory disease of small and/or medium-sized vessels with fibrinoid necrosis of the vascular wall and surrounding inflammatory infiltrate. It can occur in a variety of diseases or be a disease per se. He also revealed that for many years all necrotizing vasculitides were called under the term polyarteritis nodosa until it became clear that, while this seems to be a relatively distinct entity, diseases leading to necrotizing vasculitis are many.

Fauci et al. (1978) revealed that vasculitis is a clinicopathologic process characterized by inflammation and necrosis of blood vessels. Certain disorders have vasculitis as the predominant and most obvious manifestation, whereas others have various degrees of vasculitis in association with other primary disorders. They also revealed that necrotizing vasculitis was first described more than 100 years ago in a 27 year old man by Kussmaul and Maier The patient had what is now called classic polyarteritis nodosa.

With this early description all vasculitides were originally thought to be polyarteritis nodosa. However, it soon became clear that the dissiminated vasculitides comprised a broad spectrum of disorders involving vessels of different types, sizes and locations characterized by various clinical manifestations with or without identifiable precipitating factors.

Ryan and Wilkinson (1986) stated that dermatologists use the term (vasculitis) for a group of conditions characterized by histological changes involving the small and medium sized blood vessels of dermis (and adjacent tissue) with leukocytoclasis and fibrinoid degeneration being the principal histological features.

Fauci (1979) was of the opinion that the vasculitic syndromes encompass a broad spectrum of diseases with the common dominator of an inflammatory and often necrotic process of the blood vessels themselves. Vessels of various types, (arteries, veins and capillaries) sizes and locations may be involved within the same syndromes, or a particular pattern may be expressed almost exclusively in a given disease. In addition, the vasculitic process may be the predominant or even sole manifestation of

disease (systemic necrotizing vasculitis or the polyarteritis nodosa type) or it may be a part of another underlying primary disease (vasculitis associated with certain connective tissue diseases). Whatever the clinical setting, the prevailing clinically and pathologically relevant manifestation of the vasculitic process is the resulting tissue ischaemia.

Monroe et al. (1981) believed that the vasculitis is not a specific disease per se but it represents a reaction pattern in the skin caused by a wide variety of substances involving different pathogenic mechanisms including deposition of immune complexes.Cutaneous vasculitis, which can be seen as part of a heterogenous group of clinical syndromes or as an isolated disease entity is characterized by inflammation of small vessels of the skin (venules, capillaries and arterioles). A number of terms including hypersensitivity vasculitis as well as others have in common inflammation of the small vessels of the skin (Cupps and Fauci, 1981).

Rothschild (1984) also added that it should be apparent from the different presentations and multisystemic involvement that there is no formula that defines whether or not a given disease-state represents a form of vasculitis. The diagnosis is made perhaps on the basis of the clinical and laboratory findings and in light of one's understanding of the pathophysiology

and certain characteristics of vasculitis.

Ryan (1985) reported that for more than two centuries any clinician faced with a palpable purpura had equated this with inflammation of small vessels. More recently such lesions have become recognized as part of a disease spectrum ranging from urticaria to infarction and terms such as vasculitis, angiitis or the vasculitides have been used to describe such cases. The physical signs of urticaria, purpura, vesiculation, nodules, necrosis, as will as scars, pigmentation or patterns such as target lesions and reticulate markings of the skin have been variously named. Consequently vasculitis has accumulated a most varied and complex literature.