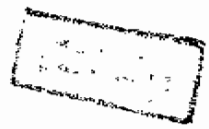
CUTANEOUS VASCULITIS



Thesis

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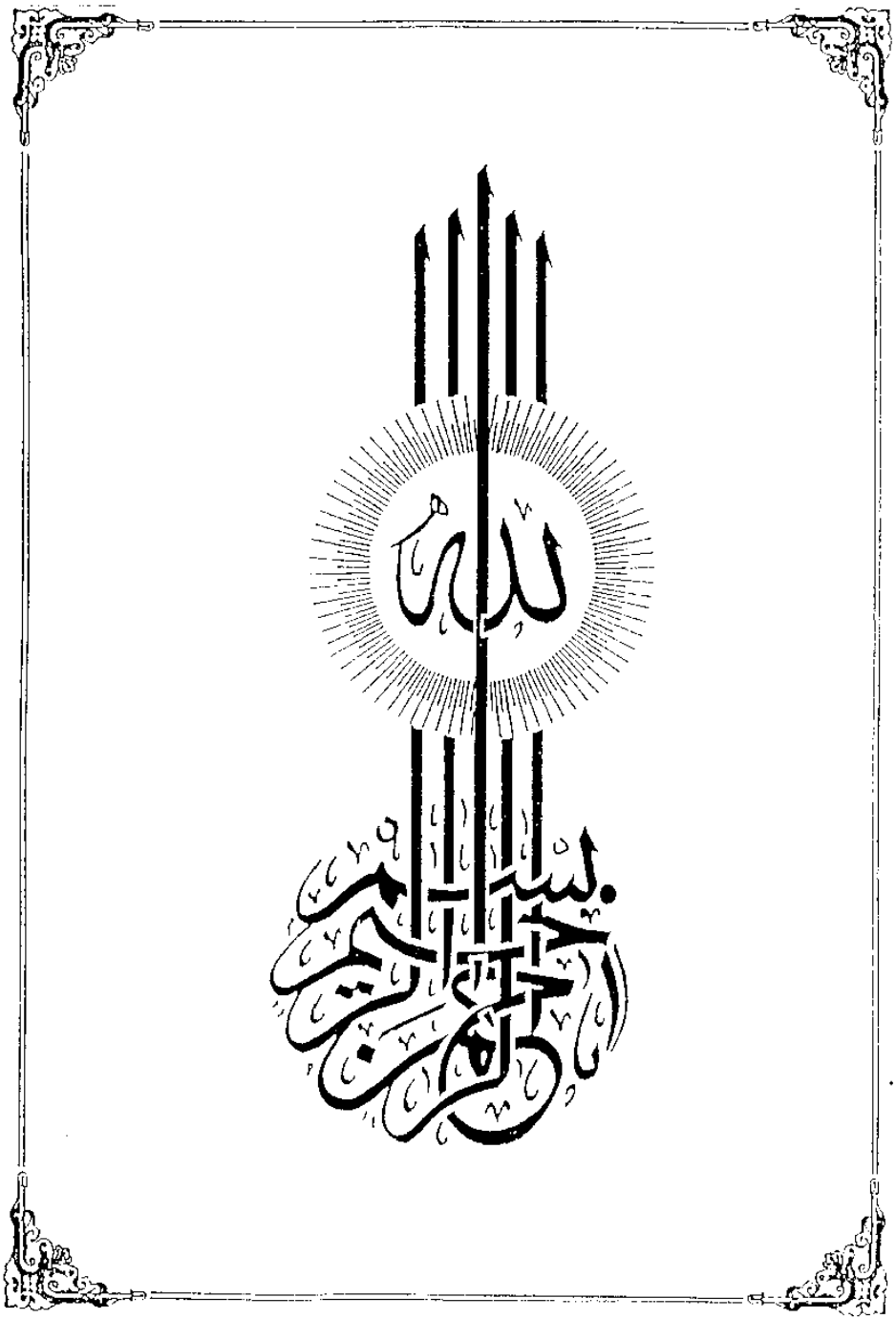

List of Abbreviations of Medical Terms

Ag	Antigen
Ab	Antibody
C.I.C.	Circulating immune complex
ECF	Eosinopnil chemotactic factor.
I C	Immune complex.
L.C.C.V.	Leucocytoclastic vasculitis.
N C f.	Neutrophil chemotactic factor.
P A F	Platelet activating factor.
P A N	Poly arteritis nodosa.
P M N	Polymorphonuclear.

Contents

INTRODUCTION

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Vasculitis means inflammation of blood vessel walls. The concept of this disease represent the efforts of a number of authors [Winkelmann and Ditto, 1964].

In 1866, Kussmaul and Maier described a disease of unknwon etiology and called it "periarteritis nodosa". It was a necrotizing arteritis of muscular arteries whose caliber was that of coronary and hepatic vessels, with this early description all vasculitides were originally thought to be polyarteritis nodosa (P.A.N.)

In 1925, Gruber suggested that polyarteritis might represent a systemic hyperallergic reaction to various infections and toxic agents to which blood vessel walls had been exposed previously.

In 1943, Rich and Gregory noted necrotizing vascular lesions which they called polyarteritis nodosa (P.A.N.) in serum sickness and in cases with sulfonamide hypersensitivity. In the subsequent years, many cases of angiitis were classed as polyarteritis nodosa, though their clinical features didn't resemble the original description of that disease.

In 1952, Zeek reviewed the problem of P.A.N. and stated that much confusion has been created by including under that term a conglomerate of syndromes many of which were probably separate entities. The diagnosis of P.A.N. was also being applied to necrotizing inflammation involving arterioles, venules, capillaries, the symptoms and signs of this small vessel arteritis were extremely variable, so, she preferred the term "necrotizing angiitis" to encompass all the entities.

In 1953, Zeek also stated that the term necrotizing angiitis is non committal in regard to etiology and was applicable to lesions in either arteries or veins of any caliber and in any location in the body. She had separated a group characterized by involvement of small vessels with little or no involvement of medium sized vessels. Clinically, all patients in this group had represented evidence of hypersensitivity during the last months of life and a fatal termination, she called this disorder "hypersensitivity angiitis".As a result of several studies, she has proved that P.A.N. and hypersensitivity vasculitis represent two distinct diseases conditions and could be differentiated clinically.

While Zeek was clarifying the problem of P.A.N. Several syndromes were being defined as allergic granulomatosis of Churg and Strauss and Wegener's granulomatosis. Other forms of necrotizing vasculitis had reported under the terms: Arteriolitis allergica of Ruiter, dermatitis nodularis necrotica, nodular dermal allergid, anaphylactoid purpura, acute parapsoriasis, allergic microbid, extracellualr cholesterésis and pyoderma gangrenosum (Winkelmann and Ditto, 1964).

These numerous syndromes were beleived related to each other by the common denominator «the microscopic picture»: a marked cellular infiltrate of P.M.N. leucocytes and numerous eosinophils, leucocytes became fragmented and nuclear debris was seen "leucocytoclasis"; extravasation of erythrocytes; necrosis take place in walls of blood vessels and hyalinization in and around the vessels. With the passage of time, a histiocytic response to amorphous necrotic material might occur leading to granuloma formation [Soter et al., 1976; Braverman, 1981].

The term angiitis was referred to necrotizing inflammatory changes primarily in and around the vessel

wall but not secondary to embolus or inflammation in neighboring tissues although adjacent tissues will be affected as a secondary manifestation as the panniculitis of nodular vasculitis. The varied manifestations of syndromes grouped as cutaneous vasculitis were not discrete entities and were not confined to the skin but they were often manifestations of one of several systemic syndromes [Copeman and Ryan, 1970]. There was vasculitis whose effects were limited to the skin or to skin and internal organs whereas some might not involve the skin at all [Stewart et al., 1978].

It was preferred to refer the small vessel angiitis by its histopathologic descriptive term "leucocytoclastic vasculitis" rather than by an etiologic or pathogenic term" hypersensitivity angiitis", and, since the cutaneous post capillary venule was the major site of involvement in small vessel angiitis, the term cutaneous necrotizing venulitis was the more appropriate term [Braverman, 1981; Jones and Eady, 1984].

The cutaneous vasculitis is a disease with diverse clinical manifestations and often involves organ systems other than the skin and frequently the skin lesions are the initial features that first calls attention

to the disease process [Ekenstam and Callen, 1984]. It is an immune complex disorder with pleomorphic lesions, the most distinctive features are: palpable purpura, persistence of urticarial lesions longer than 24 hours and most often on dependent parts of body, ischemic necrosis and infarction. Exudation of serum or blood cells occurs and damage to vessel wall range from minimal alteration of the shape of endothelial cells or its complete loss to the complete destruction of wall itself. Granulomatous changes are sometimes seen. Leucocytoclasis and fibrinoid are the principal histological features of vasculitis. The classification of cutaneous vasculitis was and still a subject to debate because the essential etiological and pathogenic factors which help to formulate a beneficial skeleton of a classification are missing. Many authors have proposed differents classifications based upon either clinical, pathological or upon the mechanism by which vasculitis might probably develop [Monroe, 1980; Steven et al., 1985].

In this thesis we wish to review the current knowledge on the subject of cutaneous vasculitis which include its clinical varieties, classification, pathogenesis, histology and treatment.

CIRCULATION

CUTANEOUS CIRCULATION

Embryology:

The epidermis contains no blood vessels. Endothelial and pericapillary cells arise from the mesoderm developing recognizable simple channels by the 10th week of intrauterine life which form arterioles and venules after blood flow. By birth, the skin shows a disorderly capillary network, most prominent in skin creases. Superficial capillary loops appear in selected areas above the general cutaneous vasculature and grow into the developing ridges of the papillary dermis to the dermoepidermal junction. A distinct horizontal vessel orientation and uniform looping papillary vessels are present in all areas of skin by the 14th to 17th week after birth. [Perera et al., 1970].

Anatomy:

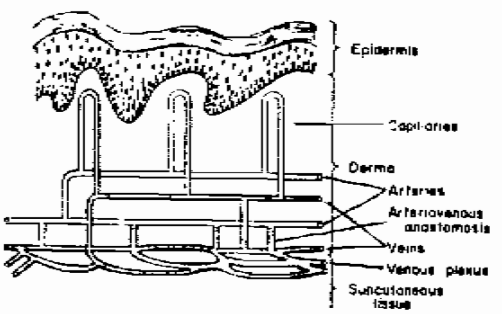

Figure 29–7. Diagrammatic representation of the skin circulation.

Guyton, (1976)

Circulatory apparatus consists of:

1- Nutritive arteries, capillaries and veins.

2- Vascular structures concerned with heating the skin: (a) extensive subcutaneous venous plexus. (b) in some skin areas, arteriovenous anastomosis (large vascular communications directly between arteries and venous plexus).

In the skin, the arterioles upon approaching the bases of the papillae, turn horizontally and give rise to metaterioles from which originate, in turn "the capillary loops". The proximal or arterial limb of the capillary loops ascend in the papilla and then turns upon itself to form the venous limb. The latter on reaching the base of the papilla joins with the venous limbs of neighboring loops to form a collecting venule. The collecting venules anastomose with one another to form a rich plexus "the subpapillary venous plexus" which run horizontally beneath the bases of the papillae and drain into the deeper veins [Guyton, 1976; Smith and Kampine, 1984].

Functions of Cutaneous Circulation:

1- nutrition of skin and removal of metabolites