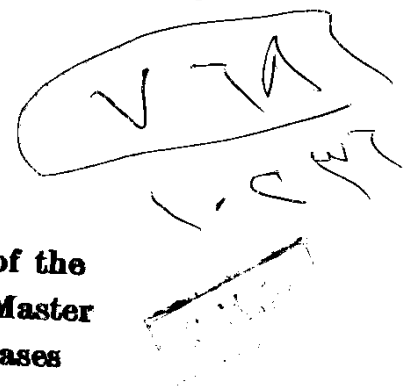
EPIDERMOLYSIS BULLOSA
HISTOPATHOLOGY AND ULTRASTRUCTURE



THESIS

Submitted in Partial Fulfilment of the
Requirements for the Degree of Master
Dermatology and Venereal Diseases

By
Samia Abdel Moniem Abdalla Shrief
M. B. Bh. B.

Under Supervision of

Prof. Dr. ZENAB EL GOTHAMY
Professor of Dermatology and Venereology
Ain Shams University

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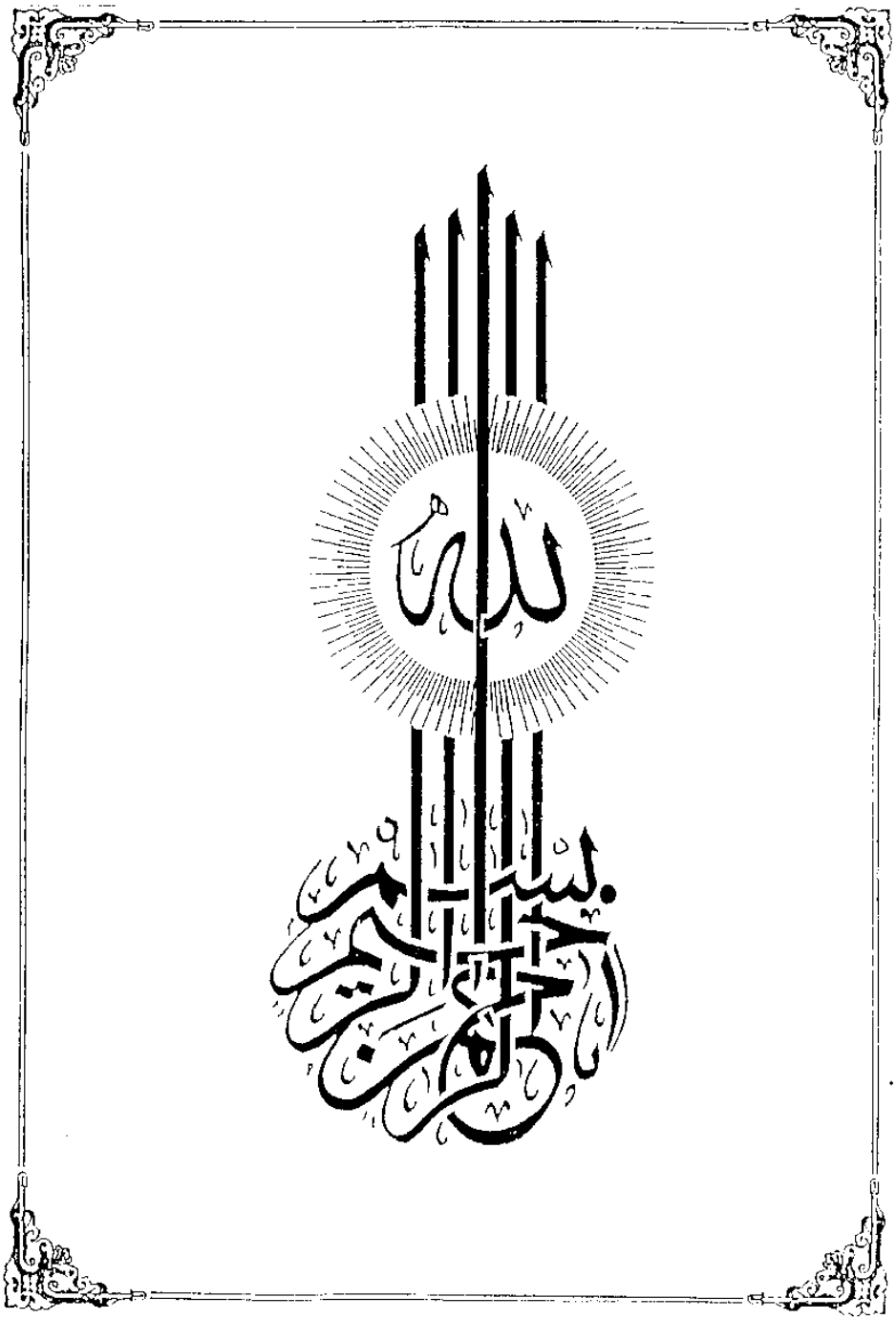

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SAMIA ABDEL MONIEM ABDALLA SHRIEF

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INTRODUCTION

INTRODUCTION

Epidermolysis Bullosa (E.B.) is a term applied to a heterogeneous group of genetically determined mechanobullous diseases, whose , common feature is blistering, after trivial trauma.

The electron microscopy has made possible the more reliable differentiation of the different syndromes and has led to a reassessement of the older, largely clinical classifications (Bauer et al., 1977).

Morecover, there are several antigenic sites known in the dermo-epidermal junction zone, where cleavage usually occurs in E.B. It might be possible to predict the cleavage sites by correlating the level of blister formation with the distribution of these antigenic determinants .

The aim of this study is to discuss in details, the histology and ultrastructural studies of different types of E.B.

REVIEW OF LITERATURE

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Epidermolysis bullosa (E.B.) , comprises group of genetically determined disorders characterized by blistering of the skin and mucous membranes.

The blisters may result from minor trauma or arise spontaneously. Although the syndromes have this feature in common and are often described as forms or variants of a single disease, they are genetically distinct diseases mediated by different mechanisms.

Various types of classification have been made by different authors. Some are based on the clinical aspects and others on the genetic aspects of the disease.

Cockayne (1933) , classified E.B. into five types.

1. Epidermolysis bullosa simplex.
2. Epidermolysis bullosa dystrophica-dominant.
3. Epidermolysis bullosa dystrophica-recessive.
4. Epidermolysis bullosa macular type (Mends da costa syndrome (MCS) , sex-linked recessive.

5. Epidermolysis bullosa (Heinrichbauer).

In this classification, the author had not mentioned epidermolysis bullosa letalis type, which has been described later on by Herlitz (1935) . Also he had considered that MCS as a variety of E.B., but now it is a separate entity.

Cockayne in 1938, added the recurrent bullous eruption of hands and feet as a localized form of epidermolysis bullosa simplex.

Touraine in 1942 , classified E.B. into :

1. Epidermolysis bullosa hereditaria simplex.
2. Epidermolysis bullosa hereditaria hyperplastica, variant: Albopapuloid form of pasini.
3. Recessive polydysplastic form which included :
   a. Proliferative (vegetative) type.
   b. Ulcero-proliferative.
   c. Mends da costa syndrome (MCS) .
   d. Malignant (fatal form letale).
   e. Heinrichbauer .

This classification has included many sub-groups of separate entities as MCS, as a variant of the polydysplastic E.B.

However, Silver (1957) classified E.P. according to the classification of Cockayne, Touraine and Herlitz, into four groups :

1. Simple non-scarring group. It appeared in infancy or childhood. It was dominantly inherited, manifested by superficial bullae over extremities mainly as a result of trauma or friction. Bullae healed without scarring or milia formation. Nails and mucous membranes were free.

2. Mendelian dominant dystrophic type . It appeared at birth or soon afterwards in children of normal physical and mental development. Lesions were severe on the hands and feet and over areas liable to trauma. They healed with their atrophic scars. Nails and mucous membranes were involved.

3. Mendelian recessive dystrophic type. It appeared shortly after birth and the affected children, were physically and mentaly poor. Bullae were large and filled with blood-stained fluid, they healed with keloid scars,

contractures and pigmentation might occur. Nails and mucous membranes were involved.

4. Lethal Hertilz type. It was a recessively inherited disease, appeared at birth, characterized by superficial bullae not related to trauma. There was marked deformity of fingers and toe-nails and some nails might be missed.lesions healed without scarring. Mucous membranes were involved.

Lapier in 1966, divided the disease into :-

1. Congenital bullous epidermolysis.
   a. Epidermolysis bullosa simplex .
   b. Epidermolysis bullosa hereditaria letalis (Herlitz disease).
2. Congenital bullous dermolysis.
   a. Hyperplastic type and albopapuloid type .
   b. Dysplastic type which was subdivided into :-
      i. Macular type (Mends da costa).
      ii. Ulcero-vegetative type.
      iii. Touraine's lethal form.

This classification correlated with the histopathological features of the disease, because he induced the term epidermolysis to the non-scarring group and induced dermolysis to the scarring, dystrophic group of the disease. However, the author represented MCS as a variety of the dysplastic type, while it is a separate disease entity.

Schnyder (1967) classified the disease into two main groups :

1. Non-scarring group including :
   a. Epidermolysis bullosa simplex.
   b. Recurrent bullous eruption of hands and feet.
2. Scarring group including ;
   a. Epidermolysis bullosa hereditaris letalis(Herlitz disease).
   b. Epidermolysis bullosa dystrophica - dominant
   c. Epidermolysis bullosa dystrophica - recessive.

In this classification Schnyder, classified the epidermolysis bullosa letalis as one of the scarring group, although the characterestic lesions of this entity heal

without scarring or milia formation, which is a great conflict.

On the oher hand, Gedde-Dahl (1971) divided E.B. according to the genetic background into :

I. Autosomal dominant group including :

1. Epidermolysis bullosa simplex (Koebner). Generalized blistering, no scarring, nails and mucous membranes are free.
2. Epidermolysis bullosa simplex (Weber - Cockayne) localized blistering of hands and feet, lesions healed without scarring, nails and mucous membranes are free.
3. Epidermolysis bullosa simplex (Ogna) characterized by skin bruising and blistering, lesions treated without scarring, but dystrophic nails might be present.
4. Epidermolysis bullosa albo-papuloidea. Characterized by skin blistering and pasini papules, which are ivory-white , perifollicular papules or plaques,