EOSINOPHILIC GRANULOMA

ESSAY.

SUBMITTED IN PARTIAL FULFILMENT

FOR

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SURGERY

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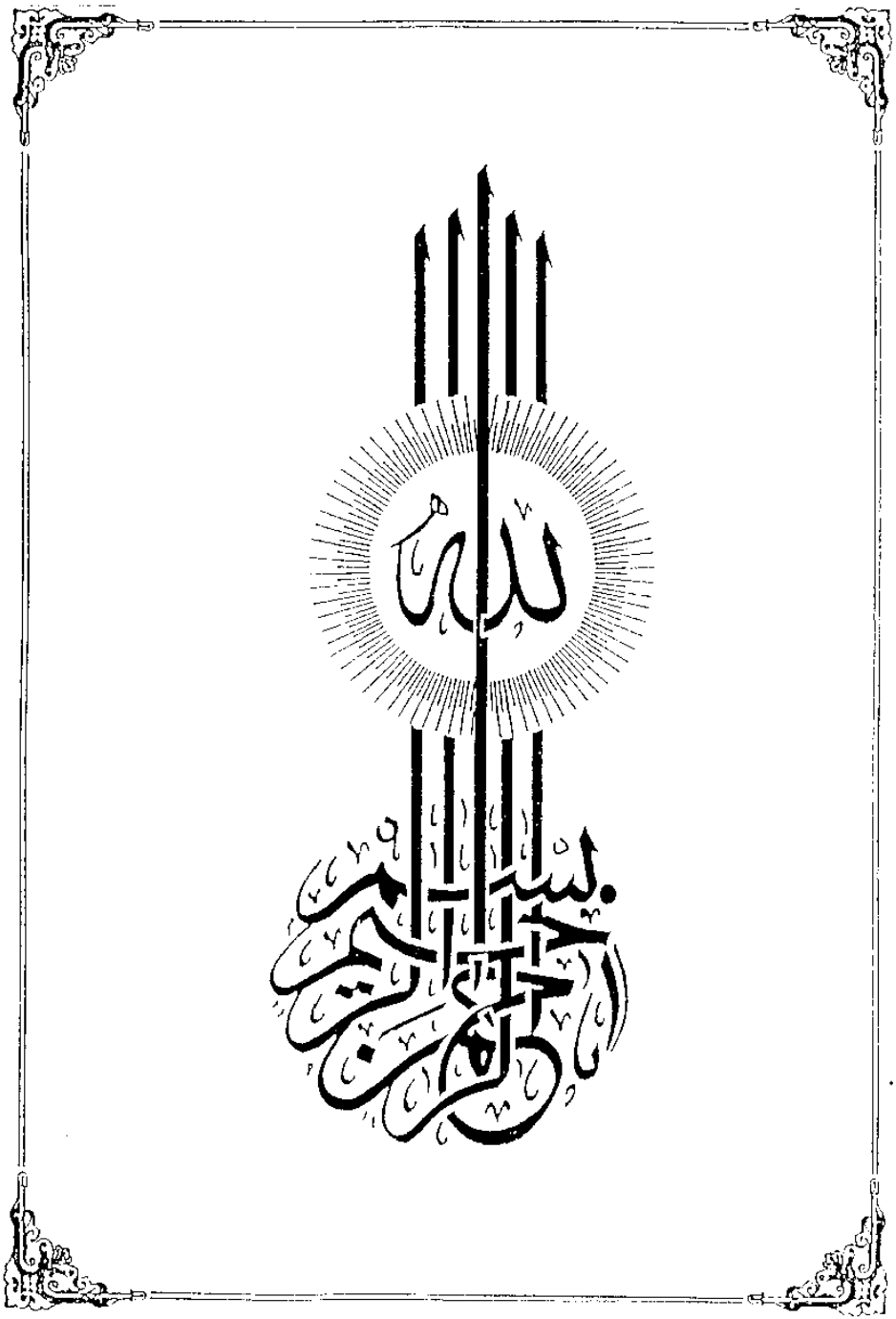

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INTRODUCTION AND HISTORICAL REVIEW.

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Definition:-

A non-neoplastic lesion of unkown etiology characterized by an intense proliferation of reticulohistiocytic elements with varying number of eosinophilic leucocytes, neutrophilic leucocytes,, Plasma cells and multi-nucleated giant cells (Schajowicz 1981).

Historical review

Prior to 1940, there were only isolated reports describing granulomatous lesions of bone with eosinophilia Otani and Ehrlich(1940) described7 cases of "Solitary granuloma of bone"in which the histologic picture was that of granulomatous lesion composed predmoninately of histiocytes and eosinophilis.

During the same year lichtenstein and Jaffe described a similar lesion which they labeled "eosinophilic granuloma of bone" and suggested that it may have a viral etiology in view of negative bacterial cultures.

Otain and Ehrlich suggested trauma as a cause,

and pointed out the similarity of one of their cases to Hand-Schuller Christian disease.

Farber beleived that some patients demonstrated a transitional form of the disease intermediate between eosinophilic granuloma and Hand-Schuller-Christian disease,which would imply a relation between the 2.

Green and Farber(1942) described 5 cases of eosinophilic granuloma of bone in children from 11 months to 11 years of age and pointed out that:-

i - The predominance for males

ii- The predilection for skull, pelvis, ribs, vertebrae, femur and humerus.

iii- The pathology in the early stage was one of bone destruction with infilration by eosinophils and large phagocytes.

iv- The lesion of eosinophilic granuloma healed by various stages of transformation in which the large mononuclear cells becomes lipophages and then developed the appearance of foam cells.

Gradually, the cells would be replaced by fibrous connective tissue and ultimately bone would replace this connectiv tissue

Juffe and Lichtenstein(1944) supported the view that eosinophilic granuloma was clinical maifestation of

a basic disorder which included, Hand-Schuller-Christian disease and Letterer-Siwe disease.

These authors pointed out that:-

Eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease may be considered as different clinical and anatomic expressions of the same basic disorder

It was concluded that :-

i - Eosinophilic granuloma of bone represent the mildest and most localised form of the disorder

ii - Letterer-siwe disease represents the gravest and most generalized expression of the disorder

iii- Hand-Schuller-Christian disease representing its most chronic and protean manifestation

Ponseti(1948)

i - Stressed the close similarity between eosinophilic-granuloma of bone, Hand-Schuller-Christian disease and Letterer-Siwe-disease and also emphasized that they were probably different manifestations of the same basic disorder.

ii - He made a clear distinction between the solitary and the multiple forms of eosinophilic granuloma, stating that in the solitary lesion the prognosis is better and that surgical curettage is usually curative.

iii- It was his opinion that multiple form of eosinophilic granuloma as a group was more resistance to therapy.

Lichtenstein 1953 proposed the term "histiocytosis X" and defined further the pathologic interrelationships of the 3 processes

- He deseribed:-

i - Localized histiocytosis-X as being equivalent to eosinophilic granuloma of bone.

ii - The acute disseminate histiocytosis-X as being equivalent Letterer-Siwe disease

iii- The chronic disseminated histiocytosis X. as being equivalent to Hand-Schuller-Christian disease.

(Sbarbaro.J.L and Francis.K.C; 1961)

Histiocytosis-x

Definition

Is a granulomatous hyperplasia of the reticuloendothelial system involving usually the bones, skin, lungs, nervous system and frequently other organ as well .

(Altman.J and Winkelmann.R.K ; 1963)

Synonyms:

Hand-Schuller-Christian disease, eosinophilic granuloma, Letterer-Siwe disease, xanthomatosis, reticuloendotheliosis, reticuloses, reticuloendothelial granuloma, reticulo-granuloma, granulomatosis, histiocytic granulomatosis, eosinophilic xanthomatous granuloma,, idiopathic histiocytosis, and progressive diffrential Histiocytosis.

(Doede.K.G. and Rappaport.H , 1967)

Nomenclature :-

- The term "histiocytosis X" was introduced by Lichtenstein in 1953 and is now also frequently used by others as a comprehensive designation for the three conditions (eosinophilic granuloma of Bone, Letterer-Siwe disease and Schuller-Christian disease).
- "An appreciable number of such names have already been proposed, although non of theses is altogether satisfactory in my opinion" (Louis Lichtenstem;1953)
  In particular, the designations employing "reticuloendotheliosis" as their central theme are objectionable for-two counts

  First, the proliferating cells in question, though apparently derived from reticuloendothelial cells are already clearly differentiating as histiocytes dysplaying phagocytic activity.

- Reticuloendotheliosis is a generic term covering proliferation of reticuloendothelial cells due to whatever cause and, as such, may have reference to neoplasia and response to abnormal lipid storage, as well as inflammatory hyperplasia of many types.
- The designations employing "reticulogranuloma and reticuloendothelial granuloma" (or granulomatosis), sometimes qualified as systemic or primary, are somewhate better from the fact that they at least convey the impression of inflammatoy response, although they also lack specificity in that . they could be applied with a much justification to tuberculosis, typhoid or brucellosis
- The designation employing "histiocytic granulomatosis" the same dissent mentioned in regard reticulogranuloma and reticulo-endothelial granuloma may be expressed
- The designation of "eosinophilic xanthomatous granuloma" seek to overcome this objection by referring to specific features of the pathologic reaction
- The designation of "idiopathic histiocytosis" suggested itself, But this name again lacks specificity in that it might apply as well to lesions of sarcoidosis for example

(Lichtenstein;1953)

Classification Of Histiocytosis X.

1. Histiocytosis X, localized to bone(**eosinophilic granuloma**, solitary or multiple)
2. Histiocytosis X, disseminated, acute or subacute (L-S syndrome)
   a) With destructive skeletal lesions (E.G)
   b) With transition to chronic phase (S-C syndrome)
3. Histiocytosis X, disseminated, chronic (S C syndrome)
   a) With destructive skeletal lesions (E.G)
   b) With early extra-skeletal lesions(indicate site resembling E.G)
   c) With acute or subacute exacerbation (L-S)
   d) With involvement predominantly of bones, lungs, pituitary and/or brain, skin, mucous membranes(oral,anal, genital),liver or lymph nodes, etc.

(Lichtenstein;1953)

It should be **reralized** that there are limits to the usefulness of classification in this disorder, since transitional cases of histiocytosis X combining features of 2 or perhaps all 3 types the disease are common. This cases are best referred to simply as histiocytosis X. without qualifying terms that may imply prognostic significance.

(Altman,J. and Winkelman R.K;1963)

Mallory has classified this group of diseases according to the most frequent age of incidence.

- Letterer-siwe disease occur-in infancy or early childhood. It is oftent rapidly fatal and the lesion are usually widely distributed through the soft tissues occasionally the bones are involved .

- Hand-Schuller-Christian disease occur in childern or adults in chronic form; the skeletal lesion frequently heals and there is somewhat less involvement of the viscera than in Letterer-Siwe disease. Healing occurs by replacement of reticuloendothelial cells with fibrous tissue and cholesteral depostis.
- Eosinophilic granuloma of bone in children and young adults involves the bone primarily, with littel or-no involvement of viscera.

(Hodgson, J .Rand Kenndy 1.J. 1951)

- Williams and Dorfman(1979) argue convincingly that these various syndromes should be grouped together under the name Histiocytosis-x- as the proliferation of histiocyte occur in these various syndromes

(Martin and David, 1973)

Martin and David stated that unifocal and multifocal eosinophilic granuloma of bone and Hand-Schuller