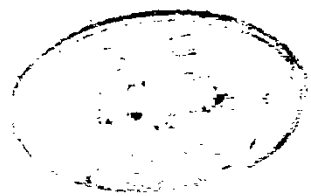
SARCOIDOSIS OF THE EYE

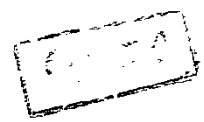
THESIS

SUBMITTED FOR PARTIAL FULFILMENT

OF THE MASTER DEGREE IN

(OPHTHALMOLOGY)





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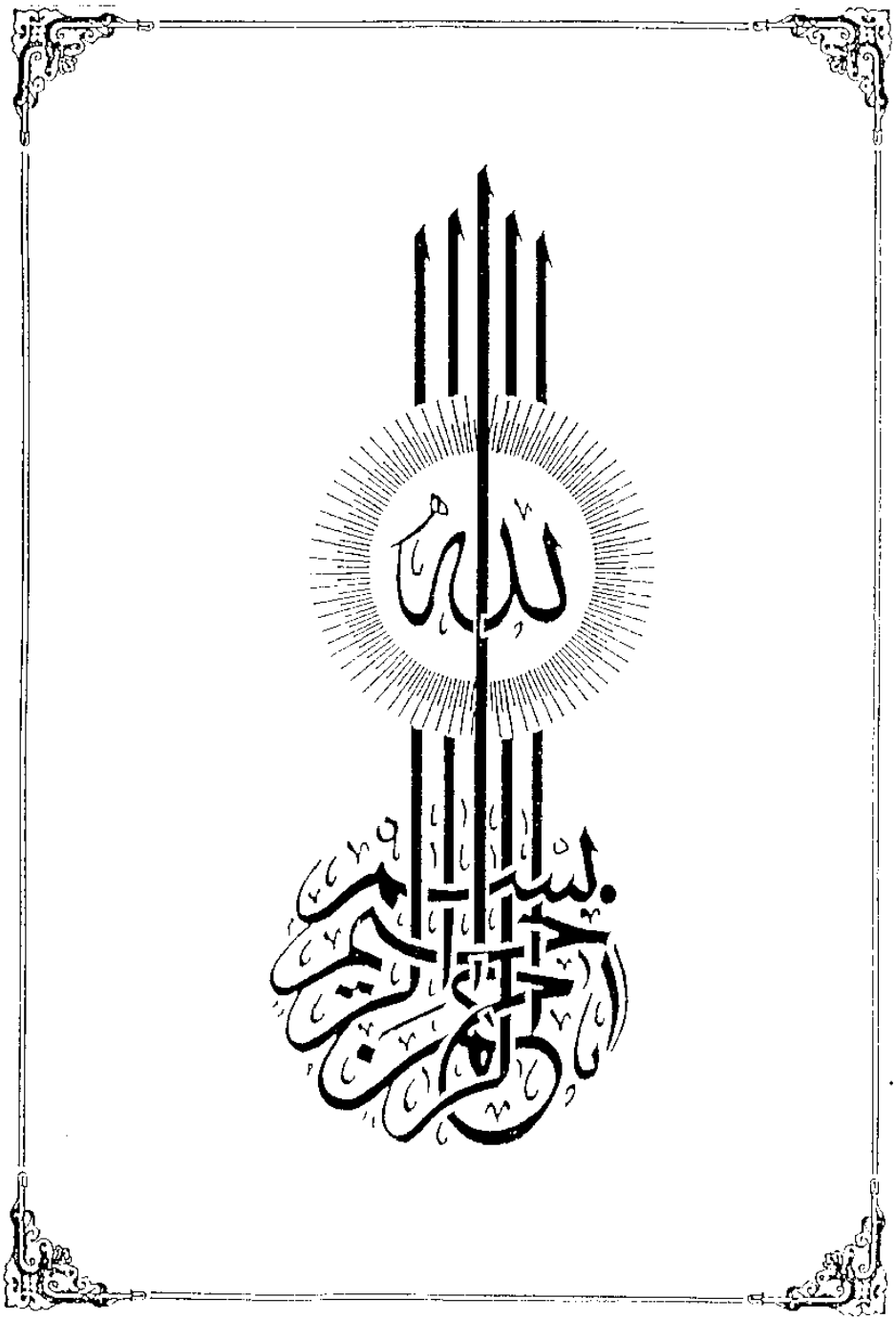

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TO MY MOTHER .....

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INTRODUCTION

Sarcoidosis, first recognised as a dermatological rarity by Caesar P.M. Boeck (1899) "Boeck's Sarcoid" and later related by Schaumann (1921-36) to a widely disseminated involvement of the reticulo-endothelial system, is an affection or perhaps a group of affections of unknown aetiology which may involve almost every organ in the body (Duke-Elder 1972).

It is considered now as a multisystem granulomatous disorder of unknown etiology most commonly affecting young adults and presenting most frequently with bilateral hilar lymphadenopathy, Pulmonary infiltration, Skin or eye lesions (James et.at. 1976).

The uveoparotid fever described by Heerfordt in 1909 was later found to be a manifestation of sarcoidosis (Bruins Slot 1936),and since then numerous reports on the various ophthalmic changes in sarcoidosis have been published (Longcope & Freiman 1952, Crick et.al., 1961, James et. al., 1964).

The frequency of ocular manifestations varies a great deal, depending on how the materials have

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been selected and how thoroughly the eyes have been examined (Longcope & Freiman 1952).

The varying part played by Sarcoidosis in the etielogy of uveitis, the complications of which may lead to deterioration of the vision or even to blindness, has begun to attract increasing interest (Perkins 1968, James et.al., 1976a).

Other manifestations of sarcoidosis in the eye and its adnexa has received less attention. These include sarcoidosis of the lacrimal apparatus, band Keratopathy and conjunctival changes. Conjunctival granulomatosis however might will be assumed to be the most common of the ophthalmic manifestations of sarcoidosis, Since the conjunctiva contains lymphatic tissue readily affected by the disorder.

The ophthalmologist is in a unique position when examining patients with sarcoidosis for the eye is the only part of the body where granulomas can readily be recognised and their clinical behaviour observed.

Histologically, the lesion of Sarcoidosis consists of a non-caseating tubercle comprised of accum-

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ulations of epithelioid cells with giant cells. Clinically it may be symptomless but usually it tends to pursue an indolent course for many years, often with exacerbations and remissions, sometimes to show spontaneous recovery but occasionally is progressive with a fatal temination.

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REVIEW OF LITERATURE

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HISTORY OF THE DISEASE

The first attempt to throw light on ocular sarcoidosis was made in 1939 by osterberg who collected 27 cases of iritis from among 500 patients with sarcoidosis, mostly reported in dermatological literature and concluded that iritis of Boeck is a rare disease. Two years after osterberg, however, among 100 patients with sarcoidosis reported in the literature, Levitt (1941) found 43 cases with ophthalmic spmptoms. On the basis of these cases, he came to the conclusion that sarcoidosis is relatively liable to attack the eye, and that any part of the eye or its adnexa may be affected.

Up to the 1950s. Sarcoid iritis was generally called iritis of Boeck (Blegrad 1938, Woods 1949) and the term is still used in the German literature (Wegner & Wurm 1957). In 1963. Mayock with his coworkers reported on a series of 145 patients with sarcoidosis and compared it with nine materials collected from the literature, comprising a total of 1254 cases.

The frequency of ophthalmic manifestations in

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these 10 materials overaged 21% with a range from the 8.7% of the retrospective study by Ricker & Clark (1949) to the 64% of the study by Longcope & Freiman (1952). In the latter study, special attention had been paid to ophthalmic symptoms. In both these studies, the vast majority of the patients were negroes.

The work of Longcope & Freiman in the USA was followed by several large, Histologically confirmed series with carefully recorded manifestations of sarcoidosis, published by the British Crick (1955), James (1956), Ainslie & James (1956) Crick et.al., (1961), James et. al., (1964, 1976b) and by the Japanese Yamada et al.,(1971). In these reports, the frequency of ophthalmic changes ranged from 25% to 63%.

The thoroughness of the examination and selection of the material undoubtedly affect the frequency figures for the ophthalmic manifestations in sarcoidosis. Without biomicroscopy it would not have been possible to record the ophthalmic manifestations in more than an average of 7%.

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Aetiology and Epidemiology:

The aetiology of sarcoidosis is unknown and wether it is one disease of a single cause or a type of response to multiple causes is undetermined. The body presumably produces granulomas when antigen is present that cannot be easily removed by the polymorphonuclear leucocytes and macrophages that occur in simple inflammation.

The tissue reaction bears strong resemblances to certain types of infection such as tuberculosis,- leprosy, histoplasmosis while it is indistinguishable from berylliosis (Caused by exposure to fumes or dust of beryllium) or the response to certain chemicals or foreign bodies.

Some of the difficulties and confusion have arisen from the definition of the disease, And Scadding (1967), indeed suggested that sarcoidosis should be considered a syndrome the principal defining feature of which is essentially histological and that if there were sufficient evidence of any particular aetiology this should be added to the descriptive definition as for example. "tuberculous sarcoidosis" or "beryllium sarcoidosis".

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Most controversy has centered around the aetiological role of tuberculosis, and the older writers assumed that sarcoidosis was an attenuated form of this infecton.

Jackson (1975) and other authors have reported cases of typical sarcoidosis which developed after BCC-Vaccination, again suggesting that in some cases tuberculosis may initiate the sarcoid response. there are however- many findings against this view: the consistent failure to find tubercle bacilli either on culture or on injection into guinea-pigs, the anergy to tuberculin, the massive involvement of the lymph nodes in sarcoidosis and the general location of evidences of this disease such as the peppering of the lungs without the common involvement of such organs as the liver and spleen which habitually occurs in miliary tuberculosis.

Again it has been claimed that other organismal infections may be causative, the patient acting as a host with a high degree of resistance and a low allergy.

Among these as would be expected a virus has been inculpated such as that of mumps, a view perhaps supported by the finding of Greene (1957) that macera-

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ted sarcoid tissue with embryonic thymus from the mouse could produce a sarcoid reaction in the inoculated brain of an adult mouse.

On the other hand, the syndrome may result from exposure to berylluim or Zirconium or other non-organismal factors such as a foreign body, while its geographical concentration in heavily forested regions suggests a relationship with pine pollen.

Indeed, Coleman (1961) succeeded in producing *sarcoid-like granulomata in the iris of the guinea-pig* by injecting pine pollen combined with Freund's adjuvant into the anterior chamber of animals previously sensitized to this antigen.

It is therefore possible that the reaction may be a peculiar cellular response to a non-specific stimulus. (Duke_elder 1972).

As a matter of fact the cause of sarcoidosis remains unknown but most authorities agree that it is a specific Disease. Whether there is a single inciting infectious or other exogenous agents is unclear. Impaired regulation of the cell functions of thymus-derived lymphocytes (T cells) and bone marrow derived lymphocytes

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