SARCOIDOSIS

THESIS

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By

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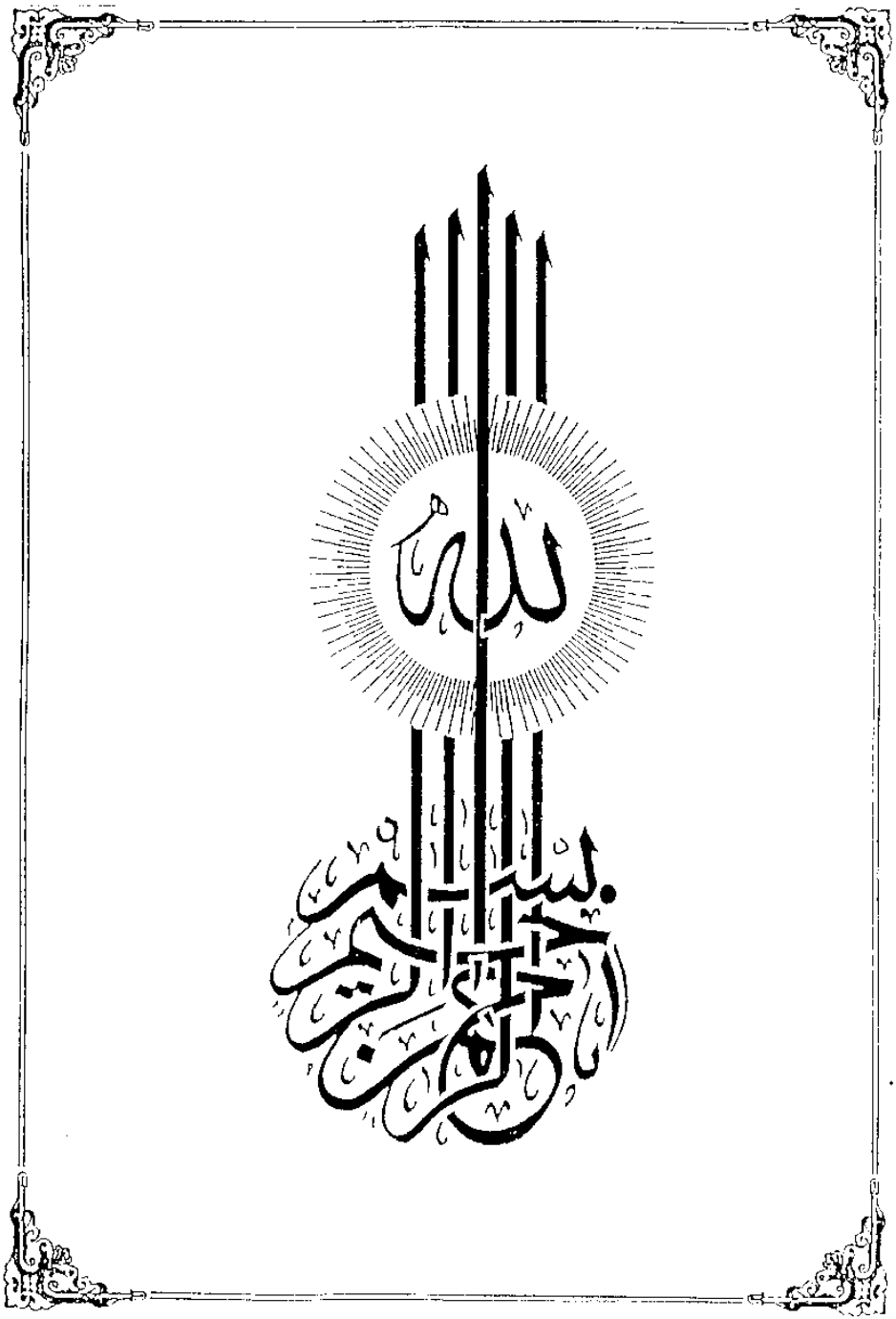

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INTRODUCTION AND AIM OF WORK

Introduction and aim of work

Sarcoidosis is a systemic disease of uncertain aetiology. The pathologic features of sarcoidosis were recognized shortly befor world war I. Sarcoidosis is a strange disease, especially intriguing to the internist, the radiologist and the pathologist. The dermatologist, the orthopedist, the ophthalmologist and recently even the epidemiologist and the immunologist have encountered this disease. To all it has been an enigma and challenge. Because there are no pathognomonic, clinical, laboratory,or histologic features.

The aim of this work is to give a short idea about the pathology and clinical picture of sarcoidosis and to show its different radiological manifestations.

PATHOLOGY

Epidemiology

Sarcoidosis is mainly a disease of young adults. Of newly diagnosid cases, 70 per cent are between 20 and 40 years of age (Hinshaw & Murray, 1979). The disease is rarely observed in children (Barnett and Einhorn, 1972). The incidence of sarcoidosis in the general population is not Known. Once regarded as rare disease. It has been difficult to establish the worldwide distribution of sarcoidosis for several reasons. Awareness of the disease by physicians varies among countries, and there are considerable international differences in the frequency of the use of screening chest roentgenograms, the principal means of discovering patient with subclinical sarcoidosis. A high incidence in some countries of diseases that share clinical and histologic features with sarcoidosis, such as leprosy and tuberculosis, may falsely lower the incidence of sarcoidosis because of diagnostic confusion; in these countries, sarcoidosis may be recognized only when the somewhat similar diseases are brought under control. Accordingly, it is conceivable that the incidence of sarcoidosis among American, Indians and Eskimos has been underestimated in the past because both groups have a relatively high incidence of tuberculosis. Certainly, early studies

underestimated the occurrence of sarcoidosis in African blacks. More and more patients with sarcoidosis are being discovered in Africa, and many of those with chronic skin manifestation have been found erroneously placed in leper colonies (Hinshaw & Murray, 1973).

Despite these difficulties, a few broad generalizations about the international distribution of sarcoidosis are permissible. Sarcoidosis is more common in cool or temperate climates than in warm or tropical ones. In Japan, for example, there is a geographic gradient of incidence, with higher concentrations of cases occurring in the cool northern sections and lower concentrations in the warmer southern areas. Sarcoidosis is relatively common in Sweden, Great Britain, and the United States, where estimated rates of prevalence range between 10 and 60 cases per 100,000 persons in the general population. In contrast, the disease is rare in Asia (Hinshaw & Murray 1979).

In Egypt sarcoidosis is a rare disease, yet it has not been statistically documented.

Aetiology

The prevailing theory about the aetiology of sarcoidosis assumes that the disease probably begins with the inhalation of an agent from the environment; however, the existence, let alone the identity, of such an agent has never been proved. (Hinshaw & Murray 1979).

In the late nineteenth and early twentieth centuries, Mycobacterium tuberculosis, perhaps in an altered, obscure form, was considered the most likely cause of sarcoidosis because patients with sarcoidosis often developed classic tuberculosis and because both conditions produce granulomas. However, the association between sarcoidosis and tuberculosis is now believed to be fortuitous because before being correctly diagnosed, patients with sarcoidosis often spent months in tuberculosis hospitals, where they acquired the disease. Furthermore, the following major differences between the two conditions make any aetiologic association unlikely:-

(1) Sarcoidosis characteristically involves the eyes, lacrimal and salivary glands, Skeletal muscle, and myocardium, sites in which tuberculosis is rare;

conversely, tuberculosis commonly affects the pleura, pericardium, peritoneum and adrenal glands, sites in which sarcoidosis is rare.

(2) In patients with sarcoidosis, hilar adenopathy is nearly always bilateral; in contrast, in patients with primary tuberculosis, enlargment of the hilar nodes is unilateral.

(3) Caseating granulomas are common in patients with tuberculosis and rare in patients with sarcoidosis, although eosinophilic necrosis of the collagen within sarcoid granulomas is at times indistinguishable from caseation.

(4) Anergy to cutaneous skin tests is the rule in sarcoidosis and the exception in tuberculosis.

(5) In countries such as Denmark, where careful sarcoid registries have been kept for several decades, the prevalence of sarcoidosis has remained constant despite a remarkable decrease in the rate of tuberculosis.

(6) In Great Britain, at least, sarcoidosis affects all socioeconomic levels of the population equally, whereas tuberculosis is more common in disadvantaged

socioeconomic groups.

(7) Corticosteroids often improve the status of patients with sarcoidosis, and antituberculous medications have no effect. In patients with tuberculosis, corticosteroids may cause clinical deterioration and antituberculous drugs are the definitive therapy (Hinshaw & Murray 1979).

Many theories have been advanced, including a causal relation with brucellosis, leprosy, Syphilis and infections due to viruses, fungi, protozoa and helminths, and the possibility that sarcoidosis may be a syndrome caused by a number of aetiologic agents. (Kendig 1961).

Bronchogenic carcinoma and other intrathoracic neoplasms may produce sarcoid-like changes in regional nodes and elsewhere, perhaps this is an auto-immune reaction (Hinshaw & Garland 1966).

Genetic predispostion to the development of sarcoidosis is suggested by a slightly higher incidence of intrafamilial cases than would be expected by chance alone, by more frequent occurrence of the disease in monozygotic than in dizygotic twins, and by the lack of the demonstrable increase in the occurrence among husbands and wives

(Hinshaw & Murray 1979).

Immunology

Sarcoidosis is accompanied by a confusing array of immunologic abnormalities, some suggesting overactivity of the immune system, such as polyarthralgias, erythema nodosum, and hyperglobulinemia, and others suggesting depression of immunity, such as anergy to delayed skin tests. In the last 10 years, research concerning sarcoidosis has focused on the immunologic abnormalities of the disorder. These studies have sought an inherent defect that predisposes to sarcoidosis and a specific in vitro diagnostic test. Neither has been found, but the studies have led to a better understanding of the humoral (B or bursa cell-mediated) and cellular (T or thymus cell-mediated) abnormalities associated with sarcoidosis. Approximately 50% of the patients have greater than 4gm/100 ml total serum protein and 20% have greater than 5gm/100 ml. indicating a disturbance of humoral activity (Hinshaw and Murray 1979).

Morphology

The distinctive, although not diagnostic, morphologic feature of sarcoidosis in all sites is the noncaseating granuloma. This is a " hard tubercle " of epithelioid cells, commonly containing giant cells of either the foreign body or langhans type. In 80 to 90 per cent of these granulomas, laminated concretions of calcium and proteins, known as Schaumann's bodies, can occasionally be found within giant cells. In additions stellate inclusions, termed asteroid bodies, are seen within giant cells in approximately 60 per cent of the granulomas. None of these changes, however, is pathognomonic. Similar hard tubercles are seen with berylliosis, the mycoses and syphilis. With long-standing sarcoidosis, the granulomas undergo progressive collagenous fibrosis and ultimately are totally replaced by scar tissue or are hyalinized. The most affected tissue are:- Lungs, lymph nodes, skin, occular, C.N.S. Cardiac, liver, spleen, renal, bone, alimentary system (Cappell & Anderson.1971, Crofton 1981).

(1) The pathology of pulmonary sarcoidosis

The basic lesion is the sarcoid, atubercle-like. The lesions may be miliary in size or they may fuse to form large masses and as the diseaseprogresses fibrosis becomes more and more a prominent feature (Boyd 1961).

(2) The pathology of lymph nodes

The lymph nodes are involved in sarcoidosis as part of the generalized disease. Characteristically, the lesions are granulomatous and noncaseous (Anderson 1971).

Often multiple groups of node are involved. Thoracic sarcoidosis is very frequently associated with involvement of the deep groups of nodes in the supraclavicular region, especially that group of nodes lying on the floor of the scalene muscles and under the sternocleidomastoid muscle especially on the right side. (Hinshaw & Garland 1966).

Albrecht et al, (1967) reported abnormal pelvic and retroperitoneal nodes in 55% of 20 patients in whom there was no other clinical evidence of node enlargement (Clouse, 1977).