Sarcoidosis in Relation to Ear, Nose, and Throat

An Essay
Submitted in partial fulfillment
for Master Degree in Otorhinolaryngology

By

George Israel Yassa
M.B. B.Ch.

Supervisors

Prof. Dr. Mohamed Nassar
Professor of Otorhinolaryngology
Faculty of Medicine
Ain-Shams University

Dr. Ali Gamal El-Din
Lecturer of Otorhinolaryngology
Faculty of Medicine
Ain-Shams University

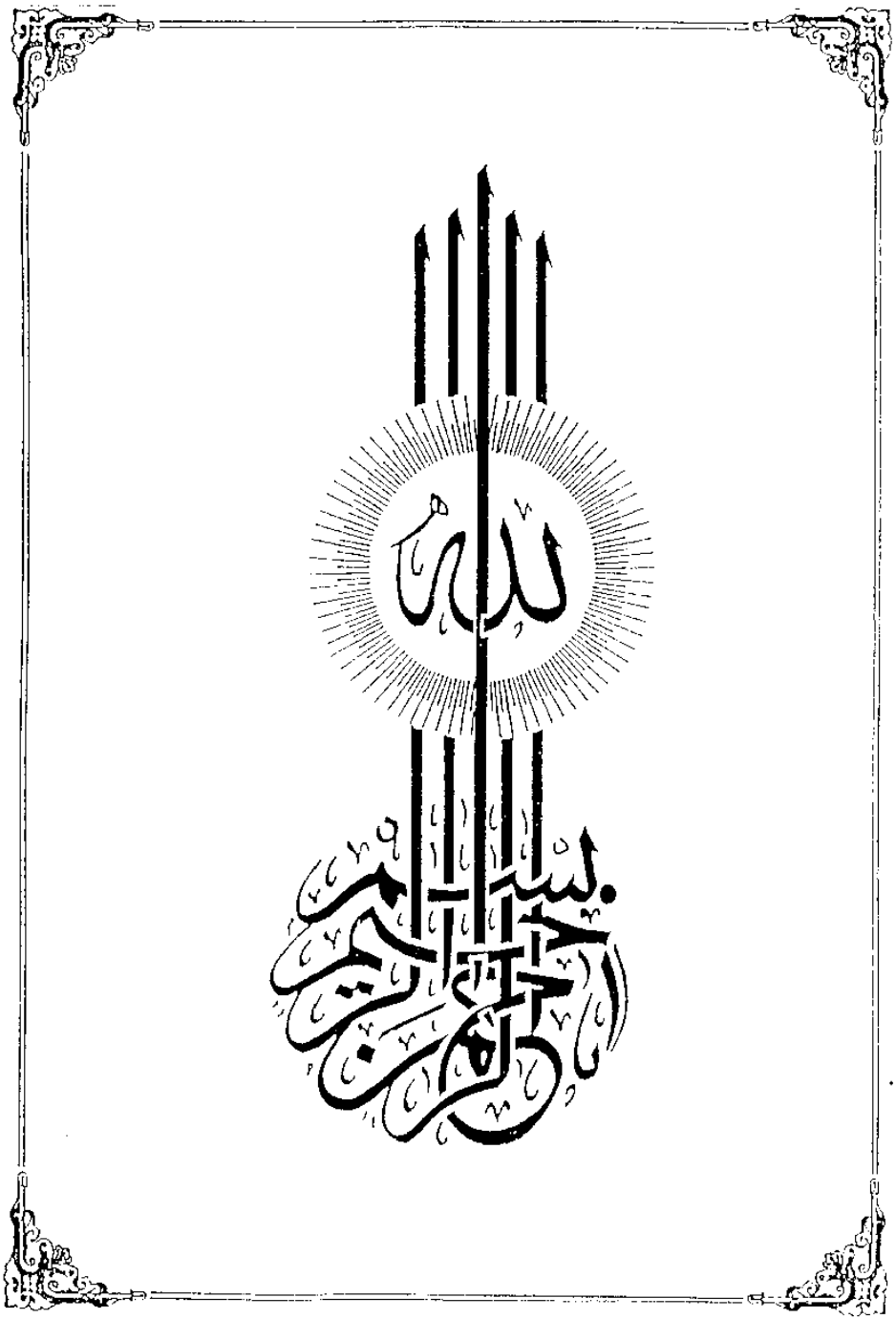
1992

Acknowledgement

I wish to acknowledge my deepest gratitude and thanks to Prof. Dr. Mohammed Nassar for his generous care and wise guidance and because he supported me with his thoughts and whenever he felt my need.

I am deeply indebted to Dr. Ali Gamal El-Din, for without him, this work would have never been completed.

I owe a particular debt also to my wife who encouraged, helped, and tolerated me during the development of this work.



CONTENTS

	Page
Chapter 1: Introduction	1
Chapter 2: Epidemiology	8
Chapter 3: Etiology	12
Chapter 4: Immunology	18
Chapter 5: Pathogenesis	30
Chapter 6: Pathology	36
Chapter 7: Course & Prognosis	45
Chapter 8: Clinical Presentation	52
Chapter 9: Diagnosis	98
Chapter 10: Differential Diagnosis	114
Chapter 11: Complications	120
Chapter 12: Treatment	123
Chapter 13: Summary	131
Chapter 14: References	136
Chapter 15: Arabic Summary	

List of Figures

Page

Fig. (1): Illustration of Mrs. Mortimer in J-Hutchinson's parer 3

Fig. (2): Illustration of C. Boeck's first patient in his paper "Multiple Benign Sarcoid of the skin" 3

Fig. (3): Pathogenesis of sarcoidosis 31

Fig. (4): Typical histologic appearance of granuloma of sarcoidosis with accompanying schematic representation 37

Fig. (5): Sarcoid granuloma in parotid gland (Histopathology) 38

Fig. (6): Segmental demyelinization within nerve 39

Fig. (7): Focal axonal degeneration 39

Fig. (8): Skin plaques on face and right nostril 53

Fig. (9): Involvement of the tip of the nasal bones associated with soft tissue swelling 54

Fig. (10): Saddle nose deformity 56

Fig. (11): A- Beak-Shaped, erythematous and shiny nose.

B- Nose of good contour covered by a split-skin graft 60

Page

Fig. (12): A- The nodular type of cutaneous sarcoidosis.
B- After a full-thickness skin graft 61

Fig. (13): Sarcoidosis of the parotid gland 64

Fig. (14): Extensive granulomatous lesions in the soft palate and oropharynx 67

Fig. (15): Sarcoid nodules at the site of earlobe piercing 69

Fig. (16): Anatomic localization of mucosal abnormalities in patients with laryngeal sarcoidosis 74

Fig. (17): Organ abnormalities associated with sarcoidosis 80

Fig. (18): Postero-anterior chest X-ray 81

Fig. (19): Air contrast CT scan of CPA 87

Fig. (20): Erythema nodosum 88

Fig. (21): Lupus pernio of the nose 90

Fig. (22): CT scan of the chest 102

Fig. (23): A- Extensive lupus pernio.
B- Same patient after corticosteroid treatment 126

INTRODUCTION

INTRODUCTION

Sarcoidosis was first recognized clinically by ***"Jonathan Hutchinson"*** in 1869, however 30 years passed before ***"Caeser Boeck"*** described the histological appearance of the cutaneous lesions. He coined the term ***"sarcoidosis"*** because of its superficial resemblance to sarcoma, the terminology stemming from the Greek, sarco-***flesh*** and eidos-***form [Miglets et al., 1977].***

The Historical Aspects of Sarcoidosis:

The first important observations on the independent character of this disease were made at the end of the last century by ***"Jonathan Hutchinson"*** in England, by ***"Ernest-Besnier"*** in France and ***"Caeser Boeck"*** in Norway, but many years had to pass before it became evident that the clinical pictures painted by the above- mentioned pioneers in sarcoidosis research were expressions of one and the same disease.

As early as 1869 Jonathan Hutchinson observed a case which most likely must have been one of sarcoidosis, but Hutchinson's most important study of this disease was in 1898

when he described ***(Mortimer Malady)*** named after the first patient whom he had observed it.

The clinical picture was characterized by an eruption of multiple, raised, dusty-red patches which have no tendency to inflame or ulcerate. They were very persistent and extended slowly. They occurred in groups and were usually on both sides and almost symmetrical (Fig. 1).

In 1889 the Frenchman, ***Ernest Besnier***, described a condition which he called lupus pernio and which presented a clinical picture differing greatly from the case observed by Hutchinson. Several decades had passed before it was admitted that this clinical picture was an expression of sarcoidosis.

In 1897, the Norwegian ***Caeser Boeck*** called this disease ***multiple benign skin sarcoid***. Boeck made a reservation: that his case was possibly of the same character as Hutchinson's Mortimer's malady (Fig. 2).

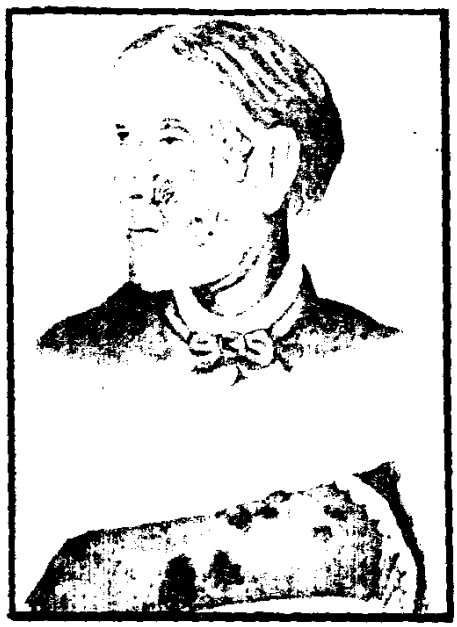


Fig. (1): Illustration of Mrs.Mortimer in J-Hutchinson's paper-in Arch of Surgery, London, 1898.

Fig. (2): Illustration of C. Boeck's first patient in his paper "Multiple Benign Sarcoid of the skin" J Cutan Dis, 1899.

Up to 1916 ***Caeser Boeck*** published 23 cases of this disease, noting its localization in the skin, the superficial lymph glands, the nasal mucosa, conjunctiva and lungs. He insisted repeatedly that he was dealing with a general disease. He came gradually to believe that the disease was a variety of tuberculosis.

Sarcoidosis research at the beginning of this century is characterized by a series of important isolated observations which have done much to the outlining of the picture we now have of this systemic disease.

In 1904, ***Darier and Roussy*** described the so called ***'subcutaneous sarcoids'*** presenting the histological picture of a granulation tissue whose appearance resembled that described by Boeck in his first case of ***'multiple skin sarcoid'***.

As early as 1909 ***Boeck*** had become convinced that ***'lupus pernio'*** and ***'multiple sarcoid'*** or ***'benign miliary lupoid'*** as he called the disease later on, presented the same character in spite of the greatest differences of the clinical picture. ***Caeser Boeck*** undoubtedly stands out as the man who carried out the most comprehensive studies of this disease. Therefore in the American literature his name is often linked to the disease as

"Boeck's Sarcoidosis".

Also, in 1909 the Danish physician, ***C.F. Heerfordt,*** described the syndrome ***'febris uveoparotidea subchronica'***. Which later on came to be identified as a manifestation of sarcoidosis.

Although earlier investigators had repeatedly insisted that sarcoidosis is a disease which can also involve internal organs, it remained for ***Kuznitzky*** and ***Bittorf*** to present in 1915 convincing evidence that the disease can give rise to a characteristic lung disease.

The Sweden, Jorgen Schumann in 1914 gave an instructive account of the localization of the disease in the lung. He has also shown that the bony changes are due to proliferation of sarcoid tissue in the Haversian canals of the bones of the hands. He showed how very frequently ***'Sarcoid'*** tissue occurs in the tonsils of patients suffering from this disease and how the liver and spleen are involved.

As regard to the pathology of sarcoidosis, an important observation was made by the Americans, ***Williams and Nickerson*** in 1935 and, independently of them, by the Norwegian, ***A. Kviem***, in 1941. These investigators were able to show that an

intracutaneous injection of a heat-sterilized emulsion of sarcoid tissue could provoke a slowly growing nodule in the skin. The histological structure of this nodule presented the same appearance as that of spontaneous sarcoid nodules in the skin. This skin reaction has been shown to be specific to a high degree and it is of great interest, not only to the diagnosis, but also to the discussion of the pathogenesis of sarcoidosis.

In 1953 ***Lofgreen*** has described the syndrome ***'bilateral hilar lymphoma'*** combined with erythema nodosum [Danbolt, 1958].

There is an apparent increase in sarcoidosis over the last 40 years due partly to better detection, especially by mass radiography ***[Savin, 1992].***

Definition

In seeking a definition of sarcoidosis, we must first decide what are its defining characteristics ***[Scadding, 1970].*** Because no agent or factor that uniquely causes sarcoidosis has yet been discovered, it is obvious that sarcoidosis cannot be defined etiologically ***[Mitchell et al., 1977].***

Consideration both of the history of the development of this

concept and of current practice shows that these observations are of 2 sorts:

1- A histologic pattern consisting of a non caseating epithelioid-cells granulomatosis, not only in clinically affected organs and tissues, but also in most instances in organs showing no clinical evidence of involvement and proceeding either to resolution or to hyaline fibrosis.

2- The concurrence in individual patients of a great variety of affected organs and tissues.

Lastly the formal definition of sarcoidosis is ***a disease characterized by the presence in all of several affected organs and tissues of non caseating epithelioid-cell granulomas, proceeding either to resolution or to conversion into featureless hyaline connective tissue.[Mitchell and Scadding, 1974].***

The study of sarcoidosis is a meeting point for specialists of different disciplines, for the lesions caused by this diffuse granulomatous process or its fibrotic sequelae are wide spread.

The aim of the work is to review in a fully detailed manner the documented literature concerning the etiology, presentation, diagnosis, and management of sarcoidosis in relation to ear, nose, and throat.

EPIDEMIOLOGY