DEMYELINATING DISEASES AND THE MIND

Assay
Submitted for partial fulfilment of
Master degree in
Neuropsychaitry

Presented by
Ayman Mohamed Ahmed Nassef
(M.B.,B.Ch)

Supervised by

Prof. Dr. Adel Sadek Ameer
Prof. of Psychiatry
Dept. of Neuropsychiatry
Faculty of Medicine-Ain Shams University.

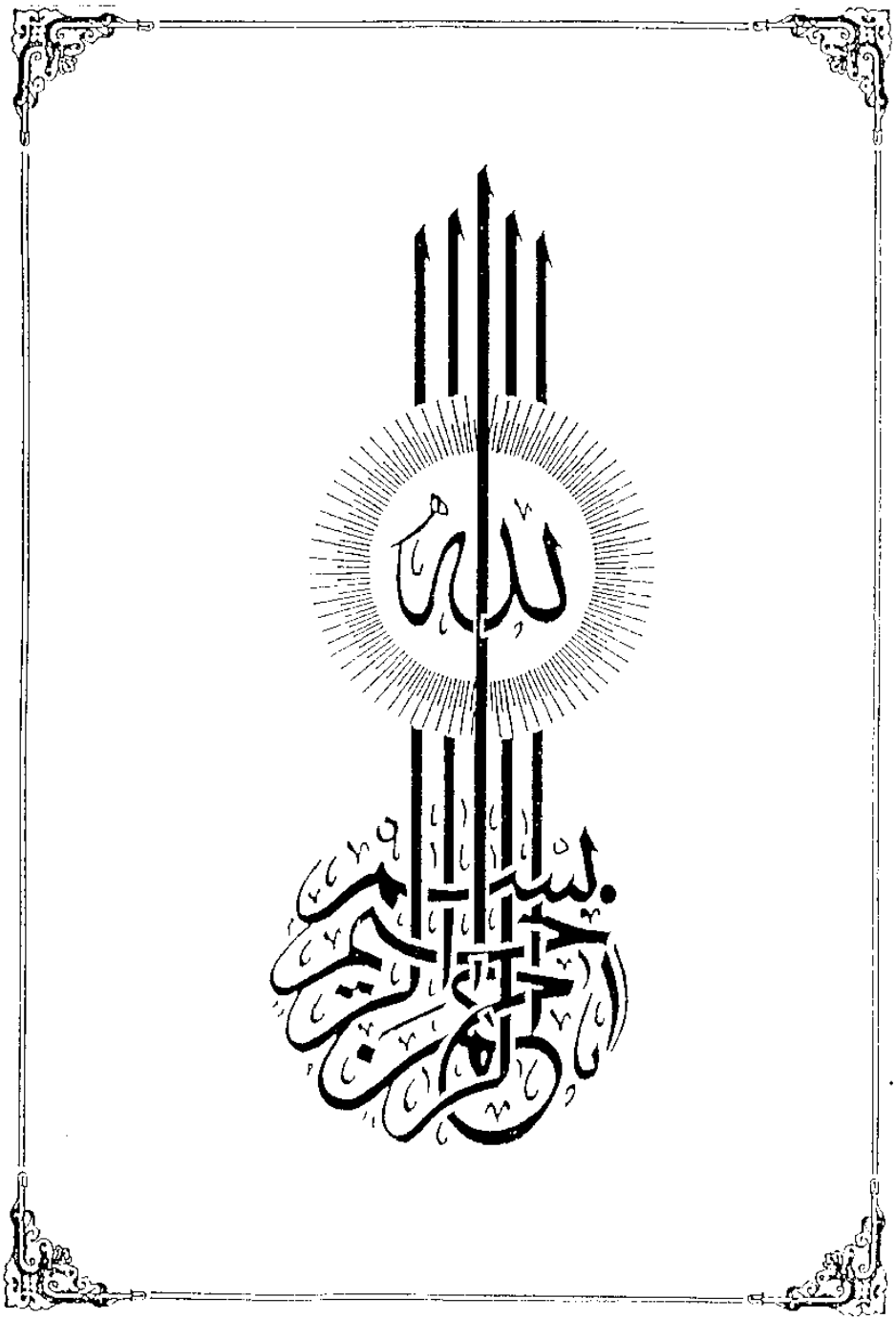
Prof. Dr. Amira Ahmed Zaki
Prof. of Neurology
Dept. of Neurosychiaty
Faculty of Medicine-Ain Shams University

Faculty of Medicine
Ain Shams University
1993

بسم الله الرحمن الرحيم

قالوا سبحانك لا علم لنا إلا ما علمتنا
إنك أنت العليم الحكيم

صدق الله العظيم
(آية ٣٢ من سورة البقرة)



ACKNOWLEDGMENT

Thanks to God who granted me the ability to perform the work of this thesis and for the helpful opportunities he has prepared for me through my sincere supervisors.

I would like to express my supreme gratitude and respect to *Professor Dr. Adel Sadek Amer, Professor of Psychiatry, Department of Neuropsychiatry, Faculty of medicine, Ain Shams University,* for his close supervision, and valuable instructions throughout this thesis.

I would like also to express my deep thanks and supreme gratitude to **Professor Dr. Amira Ahmed Zaki**, *Professor of Neurology, Department of Neuropsychiatry, Faculty of medicine, Ain Shams University,* for her supervision and encouragement throughout this work.

CONTENTS

Introduction & Aim of the Work

INTRODUCTION

Diseases of the myelin is subdivided into myelinoclastic (demyelinating) diseases and dysmyelinating diseases **(Herndon and Rudick, 1990).**

The aetiology of most of the demyelinating diseases remain obscure, though many seem to be due to complex interrelation of genetic infective, immunological and biochemical mechanisms. The pathological feature common for all demyelinating diseases are foci of destructed myelin sheath of the nerve fibre mainly situated in white matter. Multiple sclerosis (MS) is by far the most frequent of the demyelinating diseases and indeed is one of thu commonest diseases of the nervous system in temperate climate **(Walton, 1985).**

Both cognitive and psychiatric changes apparently occur in patients with MS. The cognitive changes are being recognized more widely as a result of increased sophistication of neuro-psychologic measurement techniques and heightened awareness of cerebral involvement in MS which became evident by studying brain pathology in vivo with new imaging techniques as magnetic resonance imaging (MRI). In the past, many studies of patients with MS did not adequately assess mental function because either cognitive

changes were not recognized as being an essential part of the disease or the emphasis was on the physical symptoms **(Petersen and Kokmen, 1989).**

The diagnostic criteria of MS typically do not include changes in mental status as a clinical sign in the same fashion as pyramidal, brain stem, or cerebellar findings. Consequently, the diagnosis may be overlooked when cognitive changes are the initial manifestation or implicate additional sites of lesions **(MacKay and Hirano, 1967; Lyon-Caen et al., 1986).**

Do changes in mental functions remit and relapse, as do other signs in some types of MS? Are changes in mental status more profound in one clinical profile, such as chronic progressive MS, than other profiles? All these issues will be discussed.

The issue about whether affective changes are a reaction to the disorder or part of the disease process itself remain controversial. Several studies **(Rabins et al., 1986; Honer et al., 1987)** have suggested that cerebral demyelinating plaques may be causing the disturbances, and with more precise neuropsychologic testing capable of localizing deficits. A portion of the patients' affective

changes may be related to their recognition that they have a clinically progressive debilitating disorder, yet this is only a partial explanation. Thus MS will be seen as a disorder of the cerebrum as well as of the brain stem, cerebellum and the spinal cord.

Attention has also been directed to the possibility that psychological factors may be important in precipitating fresh relapses of the disease **(Maybury and Brevin, 1984).** There is a little evidence that MS makes a specific imprint on the personality **(Lishman, 1987).**

Psychotic illnesses have been described in the disease, sometimes late in the course, but interestingly it may be the presenting feature **(Mathews, 1979).**
A recent epidemiological study into cases of MS first presenting to psychiatrists have revealed that physical symptoms and signs are almost always present when the patients complained of affective symptoms, but were often overlooked or misinterpreted **(Skegg et al., 1988).**
After cognitive and psychiatric abnormalities in MS became evident, new strategies of management will be designed.

Lastly, the dysmyelinating diseases with their cognitive and psychiatric pictures will be discussed.

AIM OF THE WORK

To highlight the different cognitive and psychiatric manifestations of demyelination diseases in order to pick up cases presenting early with cognitive and/or psychiatric manifestations and to plan better strategies of management.

Diseases of Myelin.

DISEASES OF MYELIN

Definition and Classification

Diseases of myelin constitute a significant proportion of all disabling neurologic diseases of young adults. They can be defined by failure of formation or destruction of myelin with relative preservation of axons. This group of diseases is frequently subdivided into:

Myelino clastic (demyelinating) diseases, in which normal myelin is destroyed, and dysmyelinating diseases, in which a metabolic abnormality causes breakdown of abnormal myelin or frank failure of myelination.

As with most of the medical classifications, nosology of demyelinating diseases is imperfect, since our knowledge of etiology and pathogenesis is in most instances limited **(Herndon and Rudick, 1990).**

The following classification, as a working overview of the major diseases of myelin,begins with a group of disorders of unknown cause with a presumed autoimmune pathogenesis because this is the most common and important group and ends with metabolic diseases of myelin (dysmyelinating diseases).

Diseases of Myelin

(A) Idiopathic, presumably autoimmune diseases of myelin:

. Recurrent demyelination (MS)

. Monophasic sclerosis

- Single sclerosis.
- Optic neuritis.
- Transverse myelitis.

. Acute disseminated encephalomyelitis (ADEM):

a- Post exanthem (Rubella, Varicella, Scarlet fever).

b- Following non specific infection (influenza, upper respiratory infections).

c- Post vaccinial encephalitis (Vaccinia pertussis).

d- Post inoculation with brain tissue (Rabis immunization).

(B) Infection causing demyelination:

a- Progressive multifocal leukoencephalopathy.

b- Subacute sclerosing pan encephalitis (SSPE).

c- Acquired immune deficiency syndrome-dementia complex (AIDS- dementia complex).

(C) Metabolic diseases of myelin (dysmyelinating diseases):

. Restricted to central myelin:

a- Adrenoleukodystrophy.

b- spongy degeneration.

c- Pelizaeus Merzbacher disease.

d- Fibrinoid type (Alexander's disease).

. Central and peripheral myelin:

- Metachromatic leukodystrophy.

- Globoid leukodystrophy.

(Herndon and Rudick, 1990).

Because MS is the commonest disease known in the demyelinating diseases in the temperate climate we will take it as an example and discus its cognitive and psychiatric aspects in details.