بسم الله الرحمن الرحيم

MONA MAGHRABY

شبكة المعلومات الجامعية

التوثيق الالكتروني والميكروفيلم

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جامعة عين شمس

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Clinical characteristics of Neuromyelitis optica spectrum disease (NMOSD) and factors affecting treatment response

Thesis

Submitted for Partial Fulfillment of master's degree in Neuropsychiatry

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الخصائص الاكلينيكية لاضطرابات التهاب النخاع والعصب البصري والعوامل التي تؤثر على الاستجابة للعلاج

رسالة

توطئة للحصول على درجة الماجستير في طب المخ والأعصاب والطب النفسي

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Contents

List of Abbreviations

Abb.		Full term
ADCC	:	Antibody-Dependent Cellular Cytotoxicity
ADEM	:	Acute Disseminated Encephalo-Myelitis
AHL	:	Acute Haemorrhagic Leucoencephalitis
ANHE	:	Acute Necrotizing Hemorrhagic Encephalomyelitis,
anti-TNFα	:	Anti-tumor necrosis factor-alpha Anti-tumor necrosis factor-alpha
APL	:	Anti-phospholipid
APS	:	Area Postrema Syndrome
AQP	:	Aquaporin
AQP4 IgG	:	Aquaporin-4 Immunoglobulin G
AQP4-Ab	:	Aquaporin 4 antibody
ARR	:	Annualized Relapse Rate
ATG	:	Anti-Thymocyte Globulin
AZT	:	Azathioprine
BBB	:	Blood-Brain Barrier
BBB	:	Blood Brain Barrier
BD	:	Behcet's Disease
CDC	:	Complement-Dependent Cytotoxicity
CDR	:	Complementarity-Determining Region
CIS	:	Clinical Isolated Syndrome
CNS	:	Central Nervous System
CNS	:	Central Nervous System
CPM	:	Central Pontine Myelinolysis
CR	:	Complete Response
CRP	:	C-Reactive Protein
CSF	:	Cerebrospinal fluid
CVST	:	Cerebral Venous Sinus Thrombosis
DMT	:	Disease Modifying Therapy
EDSS	:	Expanded Disability Status Scale
ELISA	:	Enzyme-linked Immunosorbent Assay
EMA	:	European Medicines Agency
EPM	:	Extra-Pontine Myelinolysis
ESR	:	Erythrocyte Sedimentation Rate
FDA	:	Food and Drug Administration
FDG	:	2-18F-fluoro-2-deoxyglucose
FLAIR	:	fluid-attenuated inversion recovery
GA	:	Glatiramer Acetate
GFAP	:	Glial Fibrillary Acidic Protein

Abb.		Full term
HELLP	:	Hemolysis, Elevated Liver enzymes and Low Platelet levels
HIV	:	Human Immunodeficiency Virus
HLA	:	Human Leukocyte Antigen
HSCT	:	Hematological Stem Cell Transplantation
ICAM1	:	Intracellular Adhesion Molecule 1
ICBD	:	The International Criteria for Behcet's Disease
ICU	:	Intensive Care Unit
IFNb	:	Interferon b
IgG	:	Immunoglobulin G
IIDDs	:	Idiopathic Inflammatory Demyelinating Diseases
IIF	:	Indirect Immunofluorescence
IL	:	Interleukin
IL-6	:	Interleukin-6
IM	:	Intramuscular
IPMSSG	:	The International Pediatric Multiple Sclerosis Study Group
IPND	:	International Panel for NMO Diagnosis
IPND	:	International Panel for Neuromyelitis optica Diagnosis
ISG	:	International Study Group for Behcet's Disease
ITM	:	Idiopathic Transverse Myelitis
IVIg	:	Intravenous Immunoglobulins
IVMP	:	Intravenous Methylprednisolone
LETM	:	Longitudinally extensive transverse myelitis
LETM	:	Longitudinal Extensive Transverse Myelitis
mAb	:	monoclonal antibody
MAC	:	Membrane Attack Complex
MM	:	Mycophenolate Mofetil
MOG	:	Myelin Oligodendrocytes Glycoprotein
MOGAD	:	Myelin Oligodendrocyte Glycoprotein antibody disorder
MRI	:	Magnetic Resonance Imaging
MRI	:	Magnetic Resonance Imaging
MS	:	Multiple Sclerosis
MS	:	Multiple Sclerosis
NBD	:	Neuro-Behcet's Disease
NMO	:	Neuromyelitis Optica
NMOSD	:	Neuromyelitis Optica spectrum disease
NPSLE	:	Neuropsychiatric Systemic lupus erythematosus
ON	:	Optic Neuritis

Abb.		Full term
ON	:	Optic Neuritis
ONTT	:	Optic Neuritis Treatment Trial
OSMS	:	Optico-Spinal form of Multiple Sclerosis
PET	:	Positron Emission Tomography
PLEX	:	Plasma Exchange
PML	:	Progressive Multifocal Leukoencephalopathy
PNS	:	Peripheral Nervous System
PPMS	:	Primary Progressive Multiple Sclerosis
RA	:	Rheumatoid Arthritis
RMS	:	Relapsing Multiple sclerosis
RRMS	:	Relapsing Remittent Multiple sclerosis
SC	:	Subcutaneous
SD	:	Standard deviation
SLE	:	Systemic lupus erythematosus
SPMS	:	Secondary Progressive Multiple Sclerosis
Th1	:	T-helper 1
Th17	:	T Helper 17
Th2	:	T-helper 2
TM	:	Transverse Myelitis
TM	:	Transverse myelitis
TNF	:	Tumor Necrosis Factor
UK	:	United Kingdom
URTI	:	Upper Respiratory Tract Infection
US	:	United States
UTI	:	Urinary Tract Infection
VCAM-1	:	Vascular Cell Adhesion Molecule 1
VEGF-A	:	Vascular Endothelial Growth Factor A
VEP	:	Visual Evoked Potential
WHO	:	World Health Organization

List of Table

List of Figures

Figure No	Subjects	Page

ABSTRACT

Background: Neuromyelitis Optica (NMO) previously known as Devic's syndrome is an autoimmune, inflammatory and demyelinating disorder of the central nervous system with predominant affection of the optic nerves and spinal cord. The global incidence and prevalence of NMO is incompletely characterized. To be more specific, it is unclear whether disease severity, frequency, clinical picture, and gender predominance vary between geographical regions. It's really challenging to obtain epidemiological and demographic data for a disorder like NMO, as it's relatively uncommon.

Aim of the Work: Study the demographic data of the patients affected with NMOSD attending ASU neurology MS unit, Document patient's response to treatment. and to estimate the incidence rate of NMOSD, according to the new international consensus NMO Diagnosis criteria.

Patients and Methods:

Patient attending ASU MS unit, during one year study time, Age: 18 to 65 years old, with Diagnosis of NMODS according to the International consensus on NMODS.

Results: 50 NMO patients were enrolled in this study, Mean age of NMO patients was 33.140 ± 9.136, ranging from 20 to 59 years, 39 were females representing 78% of the study group while 11 were males representing 22%. EDSS showed significant relation with treatment during acute attack and receiving DMD as a preventive therapy with p-value 0.052 and 0.024 respectively.

Conclusion: Since the landmark discovery of AQP4-IgG, the clinical spectrum of NMO has expanded, and non- optico-spinal clinical manifestations are recognized, accordingly rational targeted treatments have been identified that are now being delivered to patient care.

Keywords: NMO, Aquaporin, Treatment response, Rituximab, Optic neuritis, Myelitis.