

Movement disorder Emergencies

Essay submitted for partial fulfillment of master degree of
Neuropsychiatry

By
Mohammed Salim Farrag
M.B.B.ch
Sohag faculty of medicine

Under supervision of
Prof. Dr. Taha Kamel Alloush
Professor of neuropsychiatry,
Faculty of medicine - ain shams university

**Prof. Dr. Mahmoud Hemida
Elrakawy**
Professor of neuropsychiatry,
Faculty of medicine- ain shams university.

Prof .Dr. Ayman Mohamed Nassef
Professor of neuropsychiatry,
Faculty of medicine - ain shams university.

Ain shams university
Faculty of medicine
2010

طوارئ اضطراب الحركة

رسالة

توطئة للحصول على درجة الماجستير في طب الأعصاب والطب النفسي

مقدمة من

الطبيب / محمد سالم فراج
بكالوريوس الطب والجراحة - جامعة سوهاج

تحت إشراف

أ.د. طه كامل علوش

أستاذ الأمراض العصبية والنفسية

كلية الطب - جامعة عين شمس

~.د. محمود حميدة الرقاوي

أستاذ الأمراض العصبية والنفسية

كلية الطب - جامعة عين شمس

~.د. أيمن محمد ناصف

أستاذ الأمراض العصبية والنفسية

كلية الطب - جامعة عين شمس

كلية الطب
جامعة عين شمس

2010

CONTENTS

Acknowledgments	I
List of Abbreviations	II
List of Tables	IX
List of Figures.....	X
Introduction and Aim of the work	1
Chapter1:Generalprinciple.....	4
Chapter2:Disorders presenting wih stiffness or rigidity.....	19
Chapter3:Disorderspresentingwith dystonia.....	47
Chapter 4: Disorders presenting with hyperkinetic movements.....	57
Chapter 5: Disorders presenting with psychiatric manifestations...	86
Discussion.....	97
Recommendations.....	103
Summary.....	105
References	108

List of Table

<i>Page No.</i>	<i>Comment.</i>	<i>Table No.</i>
21	Etiologies for Acute Parkinsonism	1
27	Classification of Infectious Causes of Parkinsonism	2
30	Causes of Viral Encephalitis related to parkinsonism	3
35	Differential diagnosis of neuroleptic malignant syndrome	4
41	Drugs and drug interactions associated with the serotonin syndrome	5
43	Comparison of the Serotonin Syndrome and the Neuroleptic Malignant Syndrome	6

60	<i>Causes of hemiballism</i>	7
68	<i>Clinical Spectrum of Poststreptococcal Central Nervous System Disorders</i>	8
82	<i>Tic exacerbating factors</i>	9
83	<i>Drugs Implicated in Tic Exacerbations</i>	10
90	<i>Disorders Associated With Malignant Catatonia Syndrome</i>	11

List of figures

<i>Page No.</i>	<i>Comment.</i>	<i>Fig No.</i>
9	The basal ganglia	1
11	Basal ganglia circuits	2
15	Pathophysiology of neuroleptic malignant syndrome	3
51	Algorithm shows management of status dystonicus	4
66	Algorithm shows management of hemiballism	5

List of Abbreviations

ADHD	Attention-Deficit Hyperactivity Disorder
BMT	bone marrow transplantation
cpk	Creatine phosphokinase
CSF	Cerebro spinal fluid
CT	Computed tomography
D1	Dopamine 1 receptors
D 2	Dopamine 2 receptors
DIP	Drug induced Parkinsonism
DSM)	Diagnostic and Statistical Manual
DYT1	Gene DYT1
ECT	electroconvulsive therapy
ED	Economo's disease
EEG	Electroencephalography
Gpi	globus pallidus interna
HD	Huntington's disease
ICU	Intensive care unit
LSD	Lysergic acid diethylamide

MC	Malignant catatonia
MDMA	Methylioxyamphetamine
MPTP	1-methyl-4-phenyl- 1,2,3,6-tetrahydropyridine
MRI	magnetic resonance image
NMS	Neuroleptic malignant syndrome
NMLS	neuroleptic malignant-like syndrome
NMDA	N-methyl-D-aspartic acid
PANDAS	Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections
PEP	Postencephalitic parkinsonism
PHS	parkinsonism-hyperpyrexia syndrome
SC	Sydenham's chorea
SNC	substantia nigra pars compacta
SNr	substantia nigra pars reticulata
SSRIs	Selective serotonin-reuptake inhibitors
STN	subthalamic nucleus

Acknowledgments

Firstly, I would like to thanks to great **ALLAH** for his care and blessing.

I would like to express my heartily gratitude to **Prof. DR.Taha Kamel Allush** for his guidance and support as well as his constructive remarks. His invaluable suggestions paved the way for this review to appear as it is now.

Words can not express my deepest appreciation and thankfulness to my mentor **Prof. Dr. Mahmoud Hemida Elrakawy** whose encouragement, guidance and support from the initial to the final stages enabled me to carry on. His perfectionism and originality has inspired me all through the way. I extend to him my feelings of being honored to work under his considerate supervision.

I would also like to express my deepest gratitude to **Prof. Dr. Ayman Mohamed Nasef** for his immense patience and great care. I extend to him my feelings of being thankful to enable me to fulfill my goals in this work.

Furthermore, I want to express my genuine gratitude to all my professors and colleagues at the **department of neuropsychiatry, Ain Shams University**.

Also, I extend sincere thanks to **Prof. Dr. Ali shalash** for his constant support.

Finally, I offer my regards and blessings to my family; for their utmost encouragement, **my loving wife** for warmly escorting me through hard times and **my wife's family** for their great supportiveness and kind.

Introduction

Movement disorder emergency is defined as any neurological disorder, evolving acutely or subacutely, in which the clinical presentation is dominated by a primary movement disorder, and in which failure to accurately diagnose and manage the patient may result in significant morbidity or even mortality. Movement disorder emergencies include such diverse entities as acute forms of Parkinsonism, chorea, neuroleptic malignant syndrome, malignant catatonia, akinetic crisis, Sydenham's chorea (*Frucht and Fahn, 2005*).

The key to success in diagnosing and managing a patient presenting with a disorder of movement is to establish the phenomenology of the problem. Although the broad definition of patients into those who move too much (hyperkinetic disorder) or too little (hypokinetic or akinetic-rigid disorder) is relatively straightforward, differentiating jerky dystonia from tremor, or tics from chorea or myoclonus, for example, may not be a simple task to the inexperienced physician. To make matters more complicated, the movement disorder may sometimes be 'mixed' (for example, myoclonic dystonia or dystonic tremor) (*Burn, 2006*).

Clinical diagnosis of these cases is the best after good observation to the single or complex movements.

Acuet parkinsonism is a term used to describe secondary

Introduction and Aim of The Work

causes of parkinsonism , sometimes also referred to as pseudo-parkinsonism, is the second most common cause of akinetic rigid syndrome in the western world. Its prevalence is increasing due to an aging population and the rise of polypharmacotherapy (*Mena and Yebenes, 2006*).

Neuroleptic malignant syndrome (NMS) is a rare but potentially lethal adverse reaction arising from the use of medications that involve the central dopaminergic system such as phenothiazines, butyrophenones and the more recent atypical agents (*Morita et al., 2004*).

Status dystonicus is a life threatening disorder that develops in patients with both primary and secondary dystonia, characterized by acute worsening of symptoms with generalized and severe muscle contractions, how to diagnose and recent approaches in management (*Mariotti et al., 2007*).

Some of these disorders presented with psychiatric manifestations as depression, malignant catatonia, conversion and hallucination and psychosis in patient with Parkinson disease.

Prompt recognition of these emergencies is crucial, and diagnosis is based on history and phenomenology. Supportive and temporizing measures must be implemented immediately before disease-specific therapy is begun(*Hu and Frhcht, 2008*).

Aim of the work

- To review the different methods in diagnosis of movement disorder emergencies in different situations.
- To review the management of movement disorder emergencies for better treatment of such cases.

General principle

The term movement disorders often are used synonymously with basal ganglia or extrapyramidal diseases, but neither of those terms adequately encompasses all the disorders included under the broad umbrella of movement disorders. Movement disorders are neurological motor disorders manifested by slowness or poverty of movement (bradykinesia or hypokinesia, such as that seen in parkinsonian disorders at one end of the spectrum and abnormal involuntary movements (hyperkinesias) such as tremor, dystonia, athetosis, chorea, ballism, tics, myoclonus, akathisia, and other dyskinesias at the other(*Jancovich and Lang, 2004*).

Although motor dysfunctions resulting from upper and lower motor neuron, spinal cord, peripheral nerve and muscle diseases usually are not classified as movement disorders. Abnormalities in muscle tone (e.g., rigidity, spasticity, and stiff man syndrome), incoordination (cerebellar ataxia; and complex disorders of execution of movement denoted by the term apraxia) are now included among movement disorders (*Jancovich and Lang, 2004*).

Classification

In general movement disorders can be classified into:

Hypokinetic disorders

Akinesia, hypokinesia, and bradykinesia are terms used to describe patients with an absence or paucity of movement. The latter term is most commonly used, and refers to patients with Parkinsonism. (*Kumar and Calne, 2004*)

Hyperkinetic disorders

Once the examiner has determined that a patient has a hyperkinetic movement disorder, the next question is: which one is it? The major categories of hyperkinetic disorders include five conditions: dystonia, chorea, tics, myoclonus, and tremor. Rarer hyperkinetic movement disorders include entities such as paroxysmal dyskinesias, stereotypies, and episodic ataxia, and restless leg syndrome, periodic limb movements of sleep, hemifacial spasm, and hyperekplexia. Of these, only hyperekplexia (exaggerated startle syndrome) qualifies as a movement disorder emergency (*Frucht and Fahn, 2005*).

Basal ganglia considered being the seat of most movement disorders, Vesalius and Piccolomini distinguished

subcortical nuclei from cortex and white matter in the 16th century. Willis' mistaken concept in the late 17th century that the corpus striatum was the seat of motor power persisted for 200 years and formed the basis of mid-19th-century localizations of movement disorders to the striatum. By the late 19th century, many movement disorders were described but for most no pathologic correlate was known (*Lanska, 2009*).

The globus pallidus, named for its pale appearance, is a dense wedge of nerve tissue that occupies the center of the basal ganglia region. The deepest portion of the globus pallidus, named the posteroventral medial globus pallidus interna (GPi), is the site of the pallidotomy operation, and represents the main outflow connection from the globus pallidus to the thalamus. The globus pallidus is a larger and more complex structure than subthalamic nucleus (STN), with a complicated internal circuitry. Like STN stimulation, globus pallidus stimulation has broad beneficial antiparkinsonian effects (*Ford, 2009*). Basal ganglia structures shown in figure (1):