

Abdul Qayyum Rana
Peter Hedera

Differential Diagnosis of Movement Disorders in Clinical Practice

 Springer

Differential Diagnosis of Movement Disorders in Clinical Practice

Abdul Qayyum Rana • Peter Hedera

Differential Diagnosis of Movement Disorders in Clinical Practice

 Springer

Abdul Qayyum Rana
Parkinson's Clinic
of Eastern Toronto
Toronto, ON
Canada

Peter Hedera
Department of Neurology
Division of Movement
Disorders
Vanderbilt University
Nashville, TN
USA

ISBN 978-3-319-01606-1 ISBN 978-3-319-01607-8 (eBook)
DOI 10.1007/978-3-319-01607-8
Springer Cham Heidelberg New York Dordrecht London

Library of Congress Control Number: 2013950487

© Springer International Publishing Switzerland 2014

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed. Exempted from this legal reservation are brief excerpts in connection with reviews or scholarly analysis or material supplied specifically for the purpose of being entered and executed on a computer system, for exclusive use by the purchaser of the work. Duplication of this publication or parts thereof is permitted only under the provisions of the Copyright Law of the Publisher's location, in its current version, and permission for use must always be obtained from Springer. Permissions for use may be obtained through RightsLink at the Copyright Clearance Center. Violations are liable to prosecution under the respective Copyright Law.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

While the advice and information in this book are believed to be true and accurate at the date of publication, neither the authors nor the editors nor the publisher can accept any legal responsibility for any errors or omissions that may be made. The publisher makes no warranty, express or implied, with respect to the material contained herein.

Printed on acid-free paper

Springer is part of Springer Science+Business Media (www.springer.com)

Preface

Movement disorders are neurological conditions characterized by excessive involuntary movements or paucity of normal movements. Movement disorders may be complex, and thus diagnosing them can be a challenge at times. Movement disorders have wide phenomenology and have been known in human history for long time since they attract visual attention. Although some patients could have multiple types of movements, most patients present with one dominant type of abnormal movement.

We have been students of movement disorders for several years and have realized that recognizing the abnormal movement and classifying it makes the basis of differential diagnosis. Thus, closely observing the abnormal movement is very important. There are five common types of movements which patients may present with in a neurology clinic. Therefore, this handbook has been divided into five chapters. Each chapter begins with the definition of abnormal movement, and then classification, investigations, and other relevant information are given. This is followed by a brief discussion of commonly associated neurological conditions in which the given phenomenology is seen. Since this guide discusses the etiology, pathophysiology, symptoms, and treatments of these conditions only briefly, readers are encouraged to use other textbooks if the content of this manual is found insufficient.

This guide may be used by medical students, general practitioners, and other healthcare professionals. The information in this manual has been simplified to a great extent. Some of the information in this guide may represent an overview of the work of many experts in this field. Every effort has been

made to present correct and up-to-date information in this handbook, but medicine is a field with ongoing research and developments; therefore, readers should refer to other sources for latest information.

Most of the information presented in this manual is considered generally accepted practice; however, the author and the publisher are not responsible for any errors, omissions, or consequences from the application of this information and make no expressed or implied warranty of the accuracy of the contents of this publication. The reader is advised to check the package insert of each drug for its indications, dosage, and warnings. Some of the information in this booklet is based upon the author's personal observation and may not be in accordance with the experience of the experts in this field.

Toronto, ON, Canada
Nashville, TN, USA

Abdul Qayyum Rana
Peter Hedera

Contents

1 Tremor	1
1.1 Introduction.....	1
1.1.1 Classification.....	3
1.1.2 Description.....	4
1.2 Frequencies of Various Tremor Syndromes... ..	5
1.2.1 Investigations.....	5
1.2.2 Causes.....	6
1.3 Essential Tremor.....	6
1.3.1 Etiology and Pathogenesis.....	8
1.3.2 Clinical Features.....	9
1.3.3 Characteristics of Essential Tremor... ..	10
1.3.4 Diagnosis of Essential Tremor.....	11
1.3.5 Treatment of Essential Tremor.....	12
1.4 Enhanced Physiological Tremor.....	19
1.5 Parkinson's Disease.....	19
1.5.1 Etiology and Pathophysiology of Parkinson's Disease.....	22
1.5.2 Nonmotor Symptoms of Parkinson's Disease.....	24
1.5.3 Treatment of Parkinson's Disease.....	27
1.6 Pharmacological Treatments.....	27
1.7 Rehabilitation Treatment of Parkinson's Disease.....	29
1.8 Surgical Treatments of Parkinson's Disease ..	30
1.9 Cerebellar Tremor.....	30
1.10 Dystonic Tremor.....	30
1.11 Task-Specific Tremor.....	31

1.12	Medication-Induced Tremor	32
1.12.1	Frequent Causes of Drug-Induced Tremors and Parkinsonism.....	32
1.12.2	Infrequent Causes of Drug-Induced Tremors and Parkinsonism.....	33
1.13	Primary Orthostatic Tremor.....	33
1.14	Holmes Tremor	34
1.15	Alcoholic Tremor	35
1.16	Fragile X Tremor/Ataxia Syndrome (FTAX)	35
1.17	Psychogenic Tremor	36
	Bibliography.....	37
2	Dystonia	43
2.1	Classification	44
2.1.1	Special Features of Dystonia	45
2.2	Investigations.....	46
2.3	Causes.....	47
2.3.1	Primary Idiopathic Torsion Dystonia (DYT-1)	47
2.3.2	Primary Idiopathic Dystonia (DYT-6)	48
2.3.3	Dopa-Responsive Dystonia	48
2.3.4	Writer's Dystonia	49
2.3.5	Musician's Dystonia	50
2.3.6	Spasmodic Dystonia	50
2.3.7	Blepharospasm.....	50
2.3.8	Cervical Dystonia	51
2.3.9	Neurodegenerative Disorders Associated with Limb Dystonia	63
2.3.10	Psychogenic Dystonia.....	63
2.3.11	Pseudodystonia	64
	Bibliography.....	65
3	Chorea.....	69
3.1	Classification	70
3.2	Investigations.....	70
3.3	Differential Diagnosis of Phenomenology of Chorea	70

3.4	Causes.....	71
3.4.1	Genetic Causes of Chorea	71
3.4.2	Nongenetic Causes of Chorea	77
	Bibliography.....	81
4	Tics.....	87
4.1	Classification	87
4.2	General Presentations	88
4.3	Investigations.....	89
4.4	Causes.....	89
4.4.1	Tourette Syndrome	89
	Bibliography.....	90
5	Myoclonus.....	95
5.1	Classification	96
5.1.1	Classification Based upon Site of Origin	96
5.1.2	Classification Based upon Clinical Features.....	97
5.1.3	Classification Based upon Provocative Factors	99
5.1.4	Classification Based upon Etiology....	99
5.2	Investigations.....	100
5.3	Causes.....	100
5.3.1	Opsoclonus Myoclonus Syndrome ...	100
5.3.2	Exaggerated Startle Syndromes.....	101
5.3.3	Hyperekplexia	101
5.3.4	Corticobasal Degeneration.....	101
	Bibliography.....	103
Index	107

Chapter 1

Tremor

Abstract Tremor is the most common abnormal movement and is defined as an involuntary, rhythmic oscillation of a body part. This chapter reviews main characteristics of tremor, such as body segments affected by tremor (arm, head vocal cords), segment position when tremor is observed (resting and action tremor), frequency and amplitude of tremor. The main emphasis is on the most common causes of tremor, essential tremor and Parkinson's disease. We discuss main clinical features of tremor seen with these two common neurologic problems, their differential diagnosis, and main therapeutic options. Additionally, other less common causes of tremor are also briefly discussed.

Keywords Tremor • Therapy • Essential tremor • Parkinson's disease • Deep brain stimulation

1.1 Introduction

Tremor is defined as an involuntary, rhythmic oscillation of a body part. Tremor is the most commonly seen abnormal movement among all of the movement disorders. Essential tremor is the most common movement disorder. Tremor can be of many different types as follows:

I. *Resting tremor*

A resting tremor usually manifests itself when the affected body part is not voluntarily activated and is in a state of completed relaxation supported against gravity. The resting tremor either diminishes or completely disappears during the onset of voluntary activity. Some of the common causes of resting tremor are as follows:

- (a) Parkinson's disease.
- (b) Parkinson plus syndromes.
- (c) Midbrain tremor (Holmes tremor).
- (d) Wilson's disease.
- (e) Essential tremor, when severe, may have a resting component, and resting tremor can be seen in about 10–15 % of patients with essential tremor. Patients with essential tremor typically do not manifest resting tremor while walking, and this may be a useful feature to distinguish it from resting tremor in Parkinson's disease, which persists during gait.

II. *Action tremor*

An action tremor occurs during voluntary activity of the affected body part or when the affected body part is maintaining a steady posture against gravity and either diminishes or completely disappears at rest. This includes postural and kinetic tremors.

A postural tremor occurs when the affected body part is voluntarily maintaining a position against gravity. True postural tremor appears without any time delay, and it needs to be distinguished from a reemergent tremor, which is observed after 5–20 s and represents resting tremor.

A kinetic tremor occurs when the affected body part is performing a voluntary activity which could be goal directed or non-goal directed.

Some examples of action tremor are:

- (a) Essential tremor
- (b) Primary writing tremor

- (c) Enhanced physiological tremor:
 - (i) Emotional stress or anxiety
 - (ii) Endocrine: thyrotoxicosis, hypoglycemia, adrenocorticosteroids, and pheochromocytoma
- (d) Cerebellar tremor
- (e) Drug- and toxin-induced tremor such as beta agonists, lithium, neuroleptics, theophylline, valproic acid, lead, mercury, manganese, and arsenic
- (f) Peripheral neuropathy

III. *Intention tremor*

An intention tremor is present if the amplitude of the tremor increases when the affected body part is approaching the target. Intention tremor is seen in cerebellar pathology.

IV. *Task-specific kinetic tremor*

A task-specific kinetic tremor occurs during a specific activity, such as primary writing tremor.

1.1.1 *Classification*

According to Position of the Body Part Affected by Tremor

Tremor is categorized as resting tremor (if the tremor occurs while the affected body part is in complete repose), postural tremor (if the tremor occurs while the affected body part is in steady posture), or kinetic tremor (if the tremor occurs while the affected body part is exerting a movement).

According to the Regions of Body Affected

Tremor may affect different body parts including the limbs, head, tongue, jaw, vocal cords, and palate. The parts of the body that are affected by tremor depend upon the underlying neurological condition.