Marc Sindou
Yves Keravel
Aage R. Møller (eds.)

Hemifacial Spasm

A Multidisciplinary Approach

SpringerWienNewYork

Prof. Dr. Marc Sindou
Hôpital Neurologique, Lyon, France

Prof. Dr. Yves Keravel
Hôpital Henri Mondor, Creteil, France

Prof. Dr. Aage R. Møller
Department of Neurological Surgery, University of Pittsburgh, Pittsburgh, U.S.A.

Typesetting: Thomson Press (India) Ltd., New Delhi 110001

Printing: DAN, Ljubljana, Slovenia

Graphic design: Ecke Bonk

Cover illustration: Posterior operative view of the left cerebello-pontine angle in a patient operated on for a left Hemifacial spasm due to a neuro-vascular conflict between the facial nerve and the PICA. [**7**: facial nerve, **8**: cochleo-vestibular nerve, **9**: glosso-pharyngeal nerve, **10**: vagal nerve; **floc.**: flocculus; **V**: vertebral artery, **pica**: postero-inferior cerebellar artery, **L**: labyrinthine artery] Left: A pica loop is cross-compressing infero-laterally (arrow-heads) the Root Exit Zone (REZ) of the facial nerve, hidden by the cochleo-vestibular nerve. Right: The offending PICA loop has been pushed away (downward) from the facial REZ (7) with a microsucker.

Printed on acid-free and chlorine free bleached paper

With 95 partly coloured Figures

Library of Congress Cataloging-in-Publication Data

A hemifacial spasm : a multidisciplinary approach / M. Sindou, Y. Keravel. A.R. Møller (eds.).
p. cm.
Includes bibliographical references.
ISBN-13:978-3-7091-7416-6 e-ISBN-13:978-3-7091-6829-5
DOI: 10.1007/978-3-7091-6829-5

1. Face—Movement disorders. 2. Spasms. I. Sindou, Marc, 1943– II. Keravel, Yves, 1944– . III. Møller, Aage R.
[DNLM: 1. Facial Muscles—physiopathology. 2. Facial Muscles—innervation. 3. Spasm. 4. Facial Nerve—physiopathology. WE 705 H488 1997]
RC418.H46 1997
617.5'2—dc21

ISBN-13:978-3-7091-7416-6

Preface

Hemifacial spasm constitutes a severe disability, not only aesthetic because of the visible involuntary contractions affecting the muscles on one side of the face, but also functionally for daily life. This chronic disease has no curative medical treatment and is almost never, even slightly, favourably influenced by sedative medication. Its psychological consequences are really devastating, isolating the patient from his environment and destroying his social life.

Until recently, this peculiar syndrome had remained enigmatic. But during the last two decades, important advances have been made in the comprehension of its mechanisms and the efficacy of its treatment. A neurovascular conflict has been recognized as the almost constant cause of the disease. Microsurgical vascular decompression by liberating the nerve from the cross-compression exerted by an elongated neighbouring artery at the root exit zone of the facial nerve from the brain stem is the curative treatment. Botulinic toxin injected in the facial muscles turned out to be helpful to alleviate the facial contractions, even though its effects are generally transient. Taken as a palliative treatment, botulinic toxin injections can be indicated in those patients who do not want or would not tolerate an open surgery.

A wide range of specialists are involved in the diagnosis and the management of this disease: neurologists, ophthalmologists, neurophysiologists, electromyographists, neuroradiologists and, of course, neurosurgeons. For these specialists, this multidisciplinary review of the topic will be of interest.

Marc Sindou
Yves Keravel
Aage Møller

Contents

Introduction

Hemifacial spasm (HFS) is a rare disorder (its incidence is less than 1 in 100,000 in the U.S.A.) that is *characterized* by involuntary contractions of muscles on one side of the face. As defined by Wilkins (Wilkins, R. H. Hemifacial spasm: A review. Surg. Neurol. 36: 251–277, 1991), HFS is a syndrome of spontaneous and gradual onset that has as its hallmark the intermittent twitching of the muscles of facial expression on one side of the face. Typical HFS – which is the most common form of this disorder – begins in the orbicularis oculi muscle and subsequently spreads to involve other muscles that are innervated by the ipsilateral facial nerve. Voluntary facial movements often trigger the involuntary contractions of the face. Characteristically, the frequency and intensity of the spasms increase over a period of 1–10 years, and they may be followed by a sustained spasm. The spasms may fluctuate in severity and they are often affected by stress, but generally the spasm becomes more pronounced and truly disabling over time. HFS has *great cosmetic and psychological effects*, and in its advanced forms the patient's vision can be affected. HFS occurs almost exclusively in *adults*; the average age of patients in HFS studies is in the mid-fifties. The *sex ratio is slightly higher in females* than in males (incidence of 0.81 versus 0.74 per 100,000) (Auger, R. G. and Whisnant, J. P. Hemifacial spasm in Rochester and Olmsted County, Minnesota, 1960 to 1984. Arch Neurol 47: 1233–1234, 1990). The *left side of the face is affected twice* as often as the right side. The objective *electromyographic signs* of HFS are rhythmically-occurring discharges at high frequencies (20 to 200 Hz), with almost complete synchronization of the discharges within the ipsilateral facial muscles (i.e., *synkinesis*). There is a characteristic abnormal muscle contraction in patients with HFS that can be demonstrated by electrically stimulating one branch of the facial nerve while recording EMG responses from a muscle that is innervated by a different branch of the facial nerve.

Previous to the last two decades this peculiar disease had remained an enigma and was *resistant to all kinds of treatments*. But *within these two last decades important advances* in ascertaining the pathophysiology of HFS have been made which has led to an increase in the efficacy of treatment.

There have been two main *hypotheses* regarding the mechanisms involved in HFS. Gardner (Gardner, W. J. Crosstalk – the paradoxical transmission of a nerve impulse. Arch. Neurol. 14:149–156, 1966) proposed that an abnormal cross-transmission between individual fibers of the facial nerve (ephaptic transmission) gives rise to the spasm and synkinesis that are typical in patients with HFS. This hypothesis was supported by subsequent electrophysiological studies (e.g., Nielsen, V. K. Pathophysiological aspects of hemifacial spasm. Part I. Evidence of ectopic excitation and ephaptic transmission. Neurology 34: 418–426, 1984) that were interpreted to indicate that peripheral ectopic excitation and ephaptic transmission occurs within the facial nerve fibers of patients with HFS. Ferguson proposed the hypothesis that it is an increased excitability in the facial motonucleus that causes the signs of HFS (Ferguson, J. H. Hemifacial spasm and the facial nucleus. Ann. Neurol. 4: 97–103, 1978). This increased excitability was assumed to be induced by abnormal peripheral nervous activity in the facial nerve. Intraoperative electrophysiological studies in patients undergoing microvascular decompression (MVD) to relieve HFS have supported the latter hypothesis (e.g., Møller, A. R. and Jannetta, P. J. On the origin of synkinesis in hemifacial spasm: Results of intracranial recordings, J. Neurosurg. 61: 569–576, 1984; Møller, A. R. Hemifacial spasm: Ephaptic transmission or hyperexcitability of the facial motor nucleus? Exp. Neurol. 98: 110–119, 1987).

Micro Vascular Decompression (MVD) *of the seventh cranial nerve* at its root exit zone (REZ) from the brainstem has been recognized for the last two decades as being the most effective treatment for HFS. MVD has a high success rate and a low rate of complications when the procedure is performed by experienced surgeons and when the abnormal muscle response and brainstem auditory evoked potentials (BAEP) are being monitored. Although disorders such as trigeminal neuralgia can be controlled, at least for a certain period of time, by medications, especially those of the anti-epileptic type, there is no known medical treatment for HFS.

More recently, the use of *botulinum toxin* in treating HFS was introduced. This treatment has been shown to be helpful in alleviating the spasm in patients with HFS, although its effects are most often transient and there are considerable side-effects, but for some patients, botulinum toxin injections may provide enough relief to avoid surgery, at least temporarily.

The *vascular conflict* that is usually found at the REZ of the facial nerve during operations on patients with HFS has been recognized as being the cause of the disease. The REZ of the seventh cranial nerve is known as a "critical anatomical site," as it corresponds to the junction of oligodendrocyte myelin and Schwann cell myelin.

The probability that vascular compression is directly involved in creating the symptoms and signs of HFS is supported by the observation that vascular decompression of the facial nerve causes the abnormal muscle response to disappear (Møller, A. R. and Jannetta, P. J. Microvascular decompression in hemifacial spasm: Intraoperative electrophysiological observations. Neurosurgery 16: 612–618, 1985) thus indicating that the facial motonucleus is no longer being subjected to abnormal antidromic impulses. With the technology of modern imaging techniques (Magnetic Resonance Imaging, MRI), images are sufficiently refined to demonstrate the neurovascular conflict(s).

June 1996

M. Sindou
Y. Keravel
A. Møller

Acknowledgements

The editors want to especially acknowledge the ANPP Foundation (Paris) and Mrs Hélène OLLAT (M.D.), its scientific director, for their generous support which allowed contributors to meet together in Chinon, France, to prepare this book.

The Editors thank Cleat **Szczepaniak** and Joyce **Beatty** for their superb secretariat assistance. Particular thanks are given to **Cleat Szczepaniak** for her excellent and meticulous copy editing of the entire volume.