CURRENT THERAPY IN NEUROLOGICAL SURGERY

1985 • 1986

DONLIN M. LONG, M.D., PH.D.

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B.C. DECKER INC. • Toronto • Philadelphia
THE C.V. MOSBY COMPANY • Saint Louis • Toronto • London

Publisher: B.C. Decker Inc.
3228 South Service Road
Burlington, Ontario L7N 3H8

Publisher: B.C. Decker Inc.
P.O. Box 30246
Phildelphia, Pennsylvania 19103

North American and worldwide sales and distribution

The C.V. Mosby Company
11830 Westline Industrial Drive
Saint Louis, Missouri 63141

In Canada: The C.V. Mosby Company, Ltd.
120 Melford Drive
Toronto, Ontario M1B 2X5

Current Therapy in Neurological Surgery ISBN 0-941158-49-7

Library of Congress catalog card number: 85-071353

10 9 8 7 6 5 4 3 2 1

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The Unstable Low Back

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Subarachnoid Hemorrhage From An Arteriovenous Malformation of the Brain or Spinal Cord

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Lipoma and Related Abnormalities Such As Dermal Sinuses

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Achondroplasia: Neuroradiological Investigation and Surgical Management of Craniocervical Junction and Pan-Spinal Stenosis

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ACTH Tumors and Nelson's Syndrome

Intracerebral Hemorrhage

PREFACE

Why another text in neurosurgery? There are several authoritative and comprehensive works that provide detailed descriptions of neurosurgical disease, and a wide range of monographs on specific subjects are also available. However, there is no source where the interested practitioner can find a specific opinion by an expert which suggests an effective mode of therapy for an individual patient. *Current Therapy in Neurological Surgery* will fill this need. The topics have been chosen because of their clinical importance or the frequency of their occurrence; the authors have been chosen for their documented expertise in managing these specific problems, and because of their stature in the field of neurosurgery. The purpose of the book is to provide a ready reference for any physician who is confronted with a question of therapeutic management of a diagnosed neurosurgical condition. Detailed discussion of etiology, pathophysiology, diagnosis, and natural history has been waived in order to sharpen the focus on therapy. For many of these conditions there are alternative techniques, but the descriptions in this book provide an effective treatment utilized and verified by an expert in the field.

The book is not all inclusive. There are topics which have been omitted deliberately. Some, fortunately a small number, were omitted owing to the pressures of the publication schedule. Undoubtedly some do not appear because of my own neglect. Still, the topics provide a broad overview of the most common and important neurosurgical problems.

I hope that the book will prove useful to those practicing neurosurgery, and that it will have special merit for residents and students confronted with neurosurgical problems during their training. The Current Therapy concept also provides a ready reference for the neurologist, internist, or other physician confronted with a patient with a neurosurgical disease. The management techniques are directly transferrable to individual patients with the assurance that a master of the field recommends a proven method of therapy.

I wish to thank the contributors of this volume for their compliance with my request for manuscripts and for the clarity and brevity of their contributions. Such authors make the editor's job both pleasant and educational. I also wish to thank Mr. Brian Decker and his outstanding staff. Their attention to detail and the quality of their work simplified my role as editor and expedited the publication of this volume. My secretary, Mrs. Shawne Tubinis, contributed greatly to the organization and record keeping of the enterprise.

Donlin M. Long, M.D., Ph.D.

CONTENTS

Spinal Disorders

Pain

Congenital Disorders

Peripheral Nerve Disorders

INTRACRANIAL TUMORS

MENINGIOMA OF OLFACTORY GROOVE AND PLANUM SPHENOIDALE

DONLIN M. LONG, M.D., Ph. D.

Meningiomas of the olfactory groove and planum sphenoidale are the most common basal meningiomas. Symptoms and signs are both subtle and insidious so that the tumors often grow to enormous size before they are found. The patient frequently notices an inability to smell or has a visual complaint. Headache may occur. Family members often notice personality change. Seizures are sometimes the presenting sign, and uncinate attacks are not unknown. There is a paucity of physical findings. Posterior extensions of these tumors are commonly associated with both unilateral and bilateral visual change. There is often a diminution in acuity, and a broad spectrum of field defects are present. There is nothing characteristic about the visual examination. The Foster-Kennedy syndrome has been described as typical for these tumors. In fact, the combination of ipsilateral optic atrophy associated with contralateral papilledema and anosmia is extremely rare and certainly cannot be considered diagnostic for these tumors.

The diagnosis is now made by CT scan, although angiography is still useful. Definition of the blood supply for possible embolization and the location of the major intracranial vessels in relation to the tumor are both important. A bolus scan now serves the latter purpose, but angiography is required to determine the location of the feeding vessels perforating the base of the skull and to determine whether embolization is a feasible adjunct. Our radiographers are prepared to carry out embolization at the time of angiography if it appears that the tumor can be significantly reduced by interrupting the blood supply.

The decision for surgery usually is based simply on the discovery of the tumor. However, there are circumstances in which removal of the tumor may not be indicated. The aged patient or one with serious intercurrent disease, which makes the surgical procedure unduly risky, may be followed expectantly with CT scan, and surgery is undertaken only for disabling symptoms. For most patients, surgery is the only answer and should be undertaken when the tumor is discovered. Growth of the tumor simply makes surgery more difficult, and any tumor that has become symptomatic should be treated. There is no alternative to surgical removal.

SURGICAL THERAPY

The surgical approach to these tumors depends on their location and size. If the tumor is largely unilateral, a unilateral frontal approach, carried to the midline, is required. Most are bilateral, however, requiring a bifrontal flap to allow complete exposure of the tumor. A coronal skin incision hidden behind the hairline is best, irrespective of the bony approach. Most of these tumors can be removed by simply elevating the frontal lobes, leaving the falx and sinus intact. An extremely large tumor requires division of the sagittal sinus at its most anterior extremity and section of the falx to allow adequate retraction of both frontal lobes and tumor removal. It is important to avoid injury to the large draining frontal veins during this maneuver.

The tumors are of two basic varieties. In one group the tumors are extremely firm, adherent to the base of the skull, and very difficult to remove by standard techniques. In the other group the tumors are soft and suckable; even conventional suction usually is adequate to remove them totally. The laser and ultrasonic suctions have greatly expedited the removal of all these tumors.

Three major technical problems confront the surgeon in the removal of such large frontal basal tumors.

1. The tumors may have a posterior extension that involves the anterior visual apparatus. The CT scan usually demonstrates this well. Even when symptoms are unilateral, the involvement is probably bilateral. At the posterior limits of the tumor, the surgeon must be prepared to dissect the tumor free from the optic nerves and chiasm.
2. The anterior cerebral arteries and the anterior portion of the circle of Willis are sometimes involved by this same posterior extension. This can be judged by the bolus CT scan or by the angiogram. Freeing these vessels from the posterior rim of tumor is a challenging exercise.
3. A major technical problem is the degree of involvement of the floor of the frontal fossa, particularly the ethmoid and sphenoid sinuses. When the sinuses are involved, a radical removal of bone and tumor from the sinuses with repair of the anterior fossa is required, if total tumor removal is to be achieved.

Patients undergoing removal of these tumors are virtually certain to become anosmic, even with tumors that are largely unilateral. Cerebrospinal fluid fistula is a risk when an extensive frontal fossa resection is undertaken. Injury to the anterior visual apparatus and anterior circle of Willis may occur. Bifrontal damage from

retraction or venous stasis and edema is possible. A rare but very real complication is progressive sinus thrombosis, secondary to an operative injury to the sinus or its major anterior tributaries. These patients usually awaken from surgery and do well for several days until subtle personality change signals the onset of difficulties. Their condition deteriorates to a state that verges on autism; they demonstrate quadriparesis and gradually lapse into coma. If the propagation continues far enough posteriorly, death from this complication is virtually certain.

OPTIC NERVE MENINGIOMA

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Once considered rare, optic nerve sheath meningiomas are now diagnosed with increased frequency owing primarily to the advent of high-resolution computed tomography. These tumors arise from the cap cells of the arachnoid villi of the optic nerve and may take origin at any site from the intracanalicular portion of the nerve to the globe.

The growth of these tumors may take one of three directions.

1. Most are located in the subdural space and extend axially anteriorly and posteriorly along the nerve.
2. Eventually the tumor may break through the dura and present as a combined exophytic and subdural mass.
3. Rarely the tumor may be primarily dural and extradural with very little if any subdural component.

The clinical presentation depends somewhat on the mode of growth of the tumor. As it expands within the subdural space, compression of the nerve occurs, and early progressive visual loss develops. With the chronic compression of the nerve and its blood supply, cilioretinal shunt veins develop which may be seen on the optic disc. These shunt vessels have been described as characteristic of optic nerve sheath meningiomas and are nicely demonstrated by fluorescein angiography. With progressive compression of the normal circulation and invasion of the optic nerve septa, secondary glaucoma may develop. When the mass is exophytic or is associated with progressive growth, proptosis may occur but almost always is preceded by visual impairment. The clinical triad of progressive visual loss, optic atrophy with edema, and optociliary shunt vessels is considered diagnostic of optic nerve sheath meningioma. For unexplained reasons, primary optic nerve sheath meningiomas occur predominantly in middle-aged women and are rarely encountered in children. It is suggested that when they do occur in children, the course is more aggressive. In our experience with over 35 cases, however, we have documented none under the age of 20.

The differential diagnosis includes primary optic nerve atrophy from vascular occlusive disease, primary optic nerve glioma, detached retina, multiple sclerosis, as well as many other conditions associated with the aforementioned findings. The diagnosis is established primarily by CT scanning. Although hypocycloidal polytomography and arteriography are occasionally used, they have been of little value. Ultrasonic scanning of the orbit may be helpful in eliminating other diagnostic possibilities. The CT scan characteristics include a tubular enlargement of the optic nerve or a bulbous appearance anywhere along the nerve sheath. Most commonly, the tubular enlargement contrasted with the normal optic nerve extends along the entire nerve. Detection of eccentric enlargements, however, is extremely important for therapy. On coronal sections of the nerve an area of hypodensity representing the nerve, surrounded by a more peripheral dense area, is considered diagnostic of primary perioptic meningioma.

The management of patients with primary optic nerve sheath meningioma is in a transitional phase. Formerly, radical surgical excision of the optic nerve and tumor at the time of suspected diagnosis was considered necessary. With a better appreciation of the growth characteristics of the tumor, the observation that they may be present for many years in a rather dormant state, and the capability of following their growth pattern with CT and now MRI scans, a less aggressive approach may be justified.

We have now treated over thirty cases of primary optic nerve meningiomas, and the form of treatment has been based primarily on the location of the tumor in the orbit, the status of frequent visual acuity examinations, and the informed consent of the patient after the various options of treatment are fully explained. If the tumor is located in the mid to anterior portion of the optic nerve and the visual acuity is stable and not seriously compromised, we usually elect to observe the patient with CT scans at 4- to 6-month intervals with CT scans once per year. If the tumor is thought to be located primarily in the mid to anterior portion of the nerve and there is documented progressive vision loss, we consider a lateral microsurgical approach to remove the tumor from its extradural as well as subdural location.

If the tumor is located in the apex and there is normal or near-normal vision, we follow the patient with serial checks of vision and CT scans. If the tumor is