DIAGNOSIS AND TREATMENT OF POLYOSTOTIC SPINAL TUMORS

Kent K. Wu, M.D.

Senior Orthopaedic Surgeon
Department of Orthopaedic Surgery
Henry Ford Hospital
Detroit, Michigan

Built on the experiences of many Henry Ford Hospital clinicians and researchers, this book provides singularly broad coverage of the diagnosis and treatment of polyostotic spinal tumors. The initial chapter explains the spectrum of current treatments for primary and metastatic tumors of the spine: surgery, chemotherapy, radiotherapy, immunotherapy, hormonal treatment, cryotherapy and hyperthermia. Separate sections then focus on metastasis, malignant lymphomas, leukemia, chordoma, and myeloma. Each encompasses incidence, clinical signs and symptoms, laboratory findings, roentgenographic appearances, pathology, treatment, and prognosis. Augmenting the text are abundant roentgenograms, photomicrographs, clinical photographs, and pictures of autopsy specimens as well as some 2,500 pertinent and current references to the literature.

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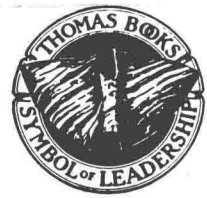

Charles C Thomas • Publisher
Springfield • Illinois • U.S.A.

Published and Distributed Throughout the World by

CHARLES C THOMAS • PUBLISHER
2600 South First Street
Springfield, Illinois 62717 U.S.A.

ISBN 0-398-04671-9

Library of Congress Catalog Card Number: 81-23425

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Library of Congress Cataloging in Publication Data

Wu, Kent K.
Diagnosis and treatment of polyostotic spinal tumors.

Bibliography: p.
Includes index.
1. Spine — Tumors. I. Title. [DNLM: 1. Spinal neoplasms — Diagnosis. 2. Spinal neoplasms — Therapy. WE 725 W959d]
RC280.S72W8 616.99'2711 81-23425
ISBN 0-398-04671-9 AACR2

Printed in the United States of America
C-1

Diagnosis and Treatment of Polyostotic Spinal Tumors

to my wife, Judith
and my children, Jonathan, Richard, and Kimberly
whose love and understanding make life an unforgettable experience

Preface

With the exception of metastases and multiple myeloma, benign and malignant tumors of the spine are rarely encountered in clinical practice. The cases presented in this book represent a very unusual collection of spinal tumors, which my predecessors and my contemporary medical colleagues at Henry Ford Hospital had the foresight of preserving in our institutional teaching files during the past thirty-five years.

It is planned to have two separate books to cover this subject. This first book deals with the classification and general consideration of spinal tumors; the different modalities of spinal tumor therapy; and the incidence, clinical symptoms and signs, laboratory findings, roentgenographic manifestations, pathology, treatment, and prognosis of metastasis, malignant lymphomas, leukemia, myeloma, and chordoma of the spine that tend to show polyostotic involvement and systemic symptoms and frequently require nonsurgical treatments.

In contrast, the second book will cover the remainder of the spinal tumors, which usually exist in the monostotic form with rare systemic symptoms prior to their metastases. Eradicative surgical procedures coupled with reconstructive spinal stabilization procedures are commonly the treatment of choice for these tumors, and medical treatments tend to play a secondary role. Consequently, two special chapters, one dealing with surgical exposure of the spine, and the other with different spinal stabilization procedures, are included in the second book.

I am indebted to Doctors C. Leslie Mitchell, Harold M. Frost, Edwin R. Guise, and Robert Knighton for allowing me to use their cases, which form

a substantial portion of the cases presented in this book. I am grateful to Doctor Ghaus M. Malik, James Ausman, and J. Speed Rogers of our Department of Neurosurgery for cooperating with me in our attempts to eradicate different spinal tumors; Doctors Joseph W. Lewis and Donald J. Magilligan of our Department of Thoracic Surgery for helping us in transthoracic and transthoraco-abdominal exposure of the spinal column; Doctors William R. Eyler, William A. Reynolds, and Roushdy S. Boulos of our Department of Radiology for assisting me in interpreting many roentgenograms; and Doctors John W. Rebuck, Gerald Fine, and Julius M. Ohorodnik of our Department of Pathology for aiding me in verifying the diagnosis of spinal tumor slides. I also wish to give my sincere thanks to Jay Knipstein for his superb illustrations on surgical exposure and different stabilization procedures of the spine; and Arthur Bowden, Walter Harlan, and John Worsham for making the numerous photomicrographs for me. Finally, I am obliged to my outstanding secretary, Mrs. Justine Frankfurth, who tirelessly and meticulously typed the whole manuscript and skillfully handled the office of my very busy orthopaedic practice at the same time.

KENT K. WU, M.D.

Introduction

EXCLUDING metastatic tumors and myeloma of the spine, spinal tumors are uncommon lesions. A great variety of spinal tumors reported in the medical literature consists of single case reports.[1-18] The fact that the author was able to publish a chordoma of the atlas,[19] an osteochondroma of the atlas,[20] and an unicameral bone cyst of the lumbar spine[21] in two leading orthopaedic journals clearly illustrates the point. Dahlin's book, *Bone Tumors: General Aspects and Data on 6,221 Cases,*[22] provides us with a rough estimate of the relative frequency of occurrence of various types of spinal tumors. This is presented in Table I.

Although Table I shows that only myeloma, malignant lymphoma, and chordoma exceed 1 percent of the total number of tumors, the whole spinal tumor group comprises 12.65 percent (or roughly one-eighth) of the entire tumor group. This is a sizable and unique tumor group, deserving special medical attention. The same table also indicates that chordoma, osteoblastoma, myeloma, fibrous histiocytoma, and hemangioma, in descending order, show significant predilection for the spinal column.

Anatomically, the spinal column lies in the deepest portion of the body and is surrounded by various bony, visceral, myofascial, and cutaneous structures, which, when coupled with the relatively small size of spinal tumors, make these tumors hard to detect by either physical or roentgenographic examination. In addition, owing to the fact that most spinal tumors are osteolytic in nature and frequently produce compression fractures that immediately obliterate all the traces of any distinctive intraosseous architecture, very different tumors can look surprisingly alike in the

Table I

THE RELATIVE FREQUENCY OF OCCURRENCE OF BENIGN AND MALIGNANT SPINAL TUMORS IN 4,277 CASES OF BONE TUMORS.

	Type of Tumor	*Frequency of Occurrence Within the Individual Tumor Group (in percent)*	*Frequency of Occurrence Within the Entire Tumor Group (in percent)*
BENIGN TUMORS	Osteochondroma	3.28	0.40
	Chondroma	4.41	0.14
	Chondroblastoma	2.27	0.02
	Chondromyxoid Fibroma	3.33	0.02
	Osteoid Osteoma	5.69	0.21
	Benign Osteoblastoma	44.19	0.44
	Giant Cell Tumor	12.03	0.75
	Fibrous Histiocytoma	28.57	0.05
	Fibroma	0	0
	Desmoplastic Fibroma	0	0
	Hemangioma	24.64	0.39
	Lipoma	0	0
	Neurilemmoma	10	0.02
MALIGNANT TUMORS	Myeloma	38.71	3.53
	Malignant Lymphoma	14.07	1.08
	Primary Chondrosarcoma	8.94	0.87
	Secondary Chondrosarcoma	9.62	0.12
	Mesenchymal Chondrosarcoma	6.67	0.02
	Osteosarcoma	2.91	0.65
	Parosteal Osteosarcoma	0	0
	Ewing's Sarcoma	7.36	0.51
	Malignant Giant Cell Tumor	10	0.05
	Adamantinoma	0	0
	Fibrous Histiocytoma	0	0
	Fibrosarcoma	9.49	0.35
	Chordoma	63.59	2.89
	Hemangioendothelioma	20	0.12
	Hemangiopericytoma	20	0.02
Total			12.65

absence of adequate histological examination. Consequently, every effort should be made to make use of all the available clinical information in order to maximize the chances of arriving at a correct diagnosis and formulating a logical course of treatment. For example, past history of cancer surgery; exposure to excessive ionizing radiation, cytotoxic substances, and carcinogenic drugs; or the presence of diseases with tendency to exhibit malignant transformation (such as neurofibromatosis, Ollier's disease, heredity multiple exostosis, and Maffuci's syndrome) in patients

with unexplained spinal symptoms should alert the clinicians of the possible existence of benign or malignant spinal tumors. Particular attention should be paid to patients' complaints of neck and back pain; paresthesia, weakness and paralysis of upper or lower extremities; and disturbances of the normal bowel or bladder functions, which all suggest impairment of spinal cord and nerve functions and mandate prompt medical attention in order to prevent permanent disability or a potentially fatal outcome. Furthermore, the presence of constitutional symptoms, such as weight loss, anorexia, malaise, ease of fatigue, fever, night sweats, etc., favors the diagnosis of metastases, especially in patients with known history of cancer.

In examining patients suspected of having spinal tumors, one should try to find the point of maximal tenderness; the presence or absence of muscle spasm, list, sensory or motor deficit, pathological reflexes, clonus, muscular atrophy and fasciculation, and palpable mass; asymmetrical, decreased, or absent deep tendon reflexes; decreased range of motion of the spine, etc., which will enable the examiner to localize the exact level of spinal involvement and the extent of neurological dysfunction.

Roentgenographically, ill-defined and inconspicuous spinal tumors are usually not well demonstrated by routine x-rays; consequently special studies, such as laminography, myelography, angiography, bone scan, and CAT scan, are often indispensable in clearly delineating the size, shape, and the extent of the spinal involvement by the tumor under consideration. This will not only improve the accuracy of a preoperative diagnosis, but can also greatly enhance the chances of a safe and complete removal of the spinal tumor by competent surgeons. In addition, preoperative laboratory studies can also provide valuable information on the nature of the spinal tumor in question. Some specific examples are elevated serum acid phosphatase in association with metastatic prostatic carcinoma; elevated catecholamine with metastatic neuroblastoma; the presence of Bence-Jones protein and typical electrophoretic curve with multiple myeloma; increased histologically distinct but abnormal cells in blood or bone marrow with leukemias; and abnormal proliferation of malignant lymphomatous cells in lymph nodes with lymphomas. Other less specific abnormal laboratory studies, including anemia, thrombocytopenia, hypercalcemia, hyperuricemia, leukocytosis, elevated erythrocyte sedimentation rate, and alkaline phosphatase caused by destruction of tumor cells and the invaded normal tissues, are the common features shared by several malignant diseases, such as metastatic carcinomas, malignant lymphomas, multiple myeloma, leukemias, etc.

Needless to say, definitive diagnosis of any spinal tumor requires adequate tissue biopsy, which will enable pathologists to identify the types of cells in the spinal tumor and the intercellular substance associated with

these cells whose shape, number, variations, and staining characteristics help to differentiate malignant lesions from benign ones. Generally speaking, all spinal tumors fall under two categories: intrinsic and extrinsic tumors. The intrinsic are tumors originating in the spinal column; extrinsic tumors are metastatic tumors from distant primary sites or invading tumors from various adjacent soft tissues and bones. A complete list of tumors of the spine can be easily obtained if one will only remember that they are no more than the benign and malignant counterparts of various tissues present in bone, which include bone, cartilage, fibrous tissue, nerve, blood vessel, bone marrow elements, fat, etc. A comprehensive classification of benign and malignant tumors of the spine is presented in Table II.

TABLE II
CLASSIFICATION OF SPINAL TUMORS

	Tissue Type	*Malignant Tumor*	*Benign Tumor*
Intrinsic spinal tumors	Osteogenic tissue	Osteogenic sarcoma Parosteal osteogenic sarcoma	Osteoid osteoma Osteoblastoma
	Chondrogenic tissue	Chondrosarcoma	Osteochondroma Chondroblastoma Chondroma Chondromyxoid Fibroma
	Fibrogenic tissue	Fibrosarcoma	Fibrous cortical defect, non-ossifying fibroma, unicameral bone cyst, fibrous dysplasia
	Vasogenic tissue	Malignant hemangio-endothelioma Malignant hemangio-pericytoma	Hemangioma Benign hemangiopericytoma
	Neurogenic tissue	Neurogenic sarcoma (malignant Schwannoma, neurofibrosarcoma)	Neurilemmoma Neurofibroma
	Myelogenic tissue	Myeloma, Hodgkin's disease, Non-Hodgkin's lymphomas, Leukemias	None
	Lipogenic tissue	Liposarcoma	Lipoma
	Reticuloendothelial tissue	Malignant giant cell tumor Malignant fibrous histiocytoma	Benign giant cell tumor Benign histiocytoma
	Notochordal tissue	Chordoma	None
	Uncertain tissue	Ewing's sarcoma, Adamantioma	None
Extrinsic spinal tumors	Metastatic tumors Invading malignant tumors from adjacent soft tissues and bones		

Once the diagnosis of a particular spinal tumor has been firmly established, the treatment naturally depends on the nature, site, and the extent of involvement of the spinal tumor. As a rule of thumb, tumors with polyostotic involvement of the spine, such as myeloma, lymphomas, leukemias, and metastases, usually do not readily lend themselves to radical surgical eradication except for limited decompression procedure to relieve pressure on the spinal cord or nerves and stabilization procedure to prevent disastrous collapse of the vertebral column. In contrast, well-localized benign and malignant spinal tumors should be removed as completely as possible in order to prevent recurrence and metastasis, and the resulting defect should be stabilized if spinal instability has been created by the surgery. In dealing with large spinal tumors with significant soft tissue and bone involvement, complete eradication may not be practical because of the danger of damaging vital organs. Under this circumstance, every attempt should be made to remove as much tumor tissue as possible in order to restore the normal functions of the impaired organs. This palliative surgical procedure may have to be repeated when the spinal tumor recurs. Adjuvant radiotherapy and chemotherapy should be employed to treat spinal tumors which are not amendable to radical surgical ablation or highly malignant spinal tumors with high potential of producing local recurrence and distal metastases. It should again be emphasized that owing to the fact that malignant spinal tumors do invade the surrounding neurovascular and visceral structures and produce distant metastases, specialists of different medical fields, such as orthopaedists, neurosurgeons, thoracic surgeons, vascular surgeons, general surgeons, radiologists, pathologists, oncologists, etc., should closely cooperate with each other in a multidisciplinary approach to strive for the highest probability of a complete tumor eradication and the minimal degrees of functional impairment at the same time.

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Contents

Diagnosis and Treatment of Polyostotic Spinal Tumors

Chapter 1

The Armamentarium of the Different Treatments of Primary and Metastatic Tumors of the Spine

MANY DIFFERENT KINDS of malignant and benign primary bone tumors originate in the spine, which is also one of the favorite sites for metastatic carcinomas and sarcomas. A comprehensive understanding of the basic principles of various modalities of cancer therapy appears to be quite desirable prior to the detailed discussion of each individual kind of spinal tumors. From a practical point of view, there are seven basic methods of treating cancers, which include surgery, chemotherapy, hormonal therapy, radiotherapy, immunotherapy, hyperthermic therapy, and cryotherapy. As a rule of thumb, surgery, radiotherapy, hyperthermia, and cryotherapy are usually used in treating localized tumors, whereas chemotherapy, hormonal therapy, and immunotherapy are more applicable to metastatic tumors. However, there are exceptions to the rules, such as intrathecal spinal or pleural injection of immunotherapeutic and chemotherapeutic agents, systemic hyperthermic therapy, whole body irradiation, etc.

Surgery

Before any definitive surgical procedure is taken to eradicate a certain spinal tumor, the exact nature of the tumor, the extent of the spinal tumor's bony and soft tissue involvements, which can be demonstrated preoperatively by CAT scan, myelography and angiography, the presence or absence of metastasis, the patient's general health, etc., should be thoroughly investigated. It should be strongly emphasized that eradicative spinal tumor surgery is often very technically demanding. It should never