TUMORS *and* TUMOROUS CONDITIONS *of the* BONES *and* JOINTS

By

Henry L. Jaffe, M.D.

Director of Laboratories and Pathologist, Hospital for Joint Diseases, New York, N. Y., Consultant, Armed Forces Institute of Pathology, Washington. D. C.

701 Illustrations on 194 Figures

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Preface

THIS book is intended to present an integrated account of the tumors and tumorous conditions affecting the bones and joints. The clinical, roentgenographic, and pathologic findings pertaining to the various lesions are discussed and correlated. Facts about the clinical findings are given with attention to their value in narrowing down the diagnostic possibilities. The roentgenographic appearance presented by a particular lesion is explained as much as possible in terms of the actual gross anatomic changes. In addition, the microscopic findings will often be found coordinated with the gross findings and sometimes even with the roentgenographic reflections of the latter. On the other hand, in regard to the pathologic findings, an effort has been made to avoid distorting the account as a whole by an excess of histologic detail relating to the lesion. Altogether, the presentations have been guided by the idea that the problems of diagnosis and differential diagnosis raised by the skeletal tumors can best be met if, in the interpretation of a given lesion, its clinical, roentgenographic, and pathologic features are considered and evaluated together. In regard to treatment, attention is devoted mainly to the choice of procedure appropriate for each condition.

In the past, the author (alone or with colleagues) has contributed various articles to the periodical literature on the subject of the skeletal tumors. However, this book represents a fresh presentation, written in continuity. In the exposition of the various lesions, a great deal of reliance has been placed on the illustrative material. The illustrations have been arranged on full-page plates to permit the simultaneous display of a number of pictures for purposes of comparison. The accompanying legends give details about each illustration, and many of them embody sufficient information to constitute case histories. Indeed, though integrated with the text, the illustrations and their legends compose in themselves something like an atlas of the skeletal tumors.

In large measure, this book conveys the author's personal conceptions of the lesions discussed. Due cognizance has, of course, been taken of the work of others in the field, and many key articles have been mentioned. However, the pertinent literature is vast, and it has not been found practical to give detailed citations from it. In any event, it is hoped that pathologists, radiologists, and orthopedic surgeons will find the book useful in their work, and that it will also be of interest to other colleagues who are confronted from time to time by problems relating to skeletal tumors.

Throughout the composition of this book, I have had the unflagging assistance of Miss E. Marion Pilpel, who has worked closely with me for the past twenty-five years. The preparation of the manuscript for the press has received the meticulous and unstinting care of Miss Edith Ross. To both Miss Pilpel and Miss Ross I am also indebted for their aid in the correction of the proof and preparation of the index.

For the preparation of many of the gross specimens for photography and detailed study, my thanks are due to my associate, Dr. Golden Selin, who has shown special ingenuity in this connection. The tissue sections from which the photomicrographs were made represent the skilled work of Mrs. Rose Afford. Practically all of the illustrations were made by Mr. Julius Weber, who spared no effort in their production. I also wish to express my gratitude to the many colleagues in the Hospital for Joint Diseases and elsewhere who have so kindly permitted me to draw upon their experience and material relating to individual cases. Finally, it is a pleasure to acknowledge the patience and cooperation received from the publishers during the years in which this book was being planned and written.

HENRY L. JAFFE

New York, N. Y.

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Chapter

1

Introduction

In this chapter we shall consider questions relating to: the classification of the skeletal tumors; the matter of how their diagnosis should be approached; and the obtainment and interpretation of pertinent biopsy specimens.

CLASSIFICATION

The tumors of the bones and joints make up a large and intriguing group of lesions. Their scope is indicated by the listing given below. This has been made on the basis of the different ways by which tumors (in the broadest sense of the term) may come to occur in the skeleton.

Tumors Developing as Primary Lesions in Bones
Tumors Developing at Sites of Pre-existing Bone Disease
Tumors Developing at Sites of Damage to Bone from Ionizing Radiation
Tumors Invading Bones from Overlying Soft Parts
Tumors Developing as Primary Lesions in Joints and Related Structures
Tumors Metastatic to the Skeleton

When the diagnostic problem relating to an individual skeletal tumor first presents itself, it is useful to keep such a comprehensive listing in mind. If one does this, one can often avoid the common error of concluding too hastily in a given case that the lesion in question is necessarily a primary bone tumor. However, the variety of the primary bone tumors (that is, the large number of different clinicopathologic entities composing the group) makes that group the one around which attention has largely centered. It also accounts for the fact that the schemata of classification in general use do not encompass the full scope of the skeletal tumors, but pertain mainly to the primary bone tumors.

The first systematic classification (at least in this country) of the tumors primary in bones was that proposed by the Registry of Bone Sarcoma of the American College of Surgeons. The Committee of the Registry (whose original members were Ewing, Codman, and Bloodgood) was formed to deal with the confusing nomenclature existing around 1920. If one studies the summarizing report of 1925 by Codman, one cannot fail to be impressed by the complexity of the Committee's task and by the advancement already represented by its original classification. Codman stressed the idea that the Committee's proposals would, of necessity, require modification as the understanding of skeletal tumors advanced. In 1939, Ewing published an official revision of the Registry's classification. This revision has undergone further modifications by others, but still constitutes the core of many of these later versions. For this reason, and also because it is of historic interest, the Registry's classification of 1939 is reproduced below.

REGISTRY'S REVISED CLASSIFICATION

	Malignant	*Benign*
Osteogenic series	Osteogenic sarcoma Medullary and subperiosteal Telangiectatic Sclerosing Periosteal Fibrosarcoma (*a*) Medullary (*b*) Periosteal Parosteal, capsular	Exostosis Osteoma
Chondroma series	Chondrosarcoma Myxosarcoma	Chondroma
Giant cell tumor series	Malignant	Epiphyseal giant cell tumor
Angioma series	Angioendothelioma Diffuse endothelioma	Cavernous angioma Plexiform angioma
Myeloma series	Plasma cell Myelocytoma Erythroblastoma Lymphocytoma	
Reticulum cell lymphosarcoma		
Liposarcoma		

The modifications embodied in the classification proposed by Phemister in 1949 took cognizance of new concepts (many in line with the writer's published views) which had already evolved in the field of the bone tumors. These modifications included specifically: a stricter interpretation of what should be called a giant-cell tumor; the abolition of the many subtypes of osteogenic sarcoma; the transfer of fibrosarcoma from the osteogenic sarcoma category to an independent status; the reduction of the myeloma series to the so-called plasma cell myeloma alone; and formal recognition of the doubts that the Ewing sarcoma was a tumor derived from vascular endothelium, and classification of it as a tumor apparently derived from primitive mesenchymal cells instead.

PHEMISTER'S CLASSIFICATION

Tissue	*Benign*	*Malignant*
Bone	Exostosis, osteoma	Osteogenic or osteosarcoma
Cartilage	Chondroma hyaline chondroblastoma	Chondrosarcoma
Fibroblast	Fibroma	Fibrosarcoma
Giant-cell	Benign giant-cell tumor	Giant-cell sarcoma
Vascular	Hemangioma cavernous organoid	Hemangioendothelioma (Ewing's sarcoma?) Cavernous angiosarcoma
Marrow		Solitary myeloma and multiple myelomas
Reticulo-endothelial	(?)	Reticulum-cell sarcoma
Lymphatic	(?)	Lymphosarcoma
Fatty	Lipoma (?)	Liposarcoma (?)
Undifferentiated-cell (mesenchyme)	(?)	Mesenchymal-cell sarcoma (Ewing's sarcoma?)

In both the Registry's and Phemister's classification (as in most of the more recent classifications founded upon them), the primary bone tumors are arranged in groups or series on the basis of the fundamental cell or tissue type, under the two general captions "Benign" and "Malignant." While the writer recognizes that classifying them in this manner has some value for general orientation, the practical utility of such classifications is open to question. Actually, the individual primary bone tumors are brought into sharper focus, and certain confusions about them are avoided, if one considers them outside of the framework of such classifications and views them simply as clinicopathologic entities. Toward the delimitation of these entities, the clinical findings, the roentgenographic picture, and the dominant histologic pattern all contribute.

For instance, if chondroma and chondrosarcoma are juxtaposed as the representatives of the cartilage series of tumors, one is merely contrasting the fundamentally benign with the malignant cartilage tumors. There is nothing in such an alignment to indicate that the essentially benign cartilage tumors include both solitary chondroma (enchondroma) and multiple enchondromatosis, which differ so importantly in regard to their clinical and roentgenographic aspects, as well as, incidentally, in regard to certain nuances in the histologic picture. Furthermore, in addition to the benign and malignant central cartilage tumors, there are benign and malignant cartilage tumors which develop in relation to the periosteum and which may be designated as juxtacortical chondroma and juxtacortical chondrosarcoma. Altogether, little is contributed to the understanding of the various cartilage tumors by considering them merely as members of a "cartilage series" of tumors. Furthermore, the restrictiveness of this type of classification invites the arbitrary placement of certain lesions in one category or another. Thus, in some of the more recent classifications based on tissue type, benign chondroblastoma and even chondromyxoid fibroma are forced into the category of the cartilage growths. Actually, as was brought out in the original descriptions of these lesions, they are not exactly cartilage tumors, despite the implication of "cartilage" in their names.

Let us now consider the conventional classification of lesions which have osseous tissue as an essential constituent. For example, one commonly finds exostosis and osteogenic sarcoma juxtaposed as the benign and malignant forms of the "bone" or "osteogenic" series of lesions. The alignment of these two lesions in this way implies a relationship which actually does not exist and obscures fundamental differences between them. An osteogenic sarcoma represents a bone-forming connective-tissue sarcoma. On the other hand, it is only in a superficial sense that a solitary exostosis (osteocartilaginous exostosis) is a tumor at all. Actually, it represents the expression of a developmental aberration of periosteal activity. Also, if one lists exostosis alone as the benign form in the "osteogenic" series of lesions, cognizance is not being taken of hereditary multiple exostosis. Though the latter bears a pathologic kinship to solitary exostosis, it reveals itself as a strikingly different disorder when viewed in its entirety as a clinicopathologic complex.

Still another illustration of difficulty which may arise from classification on the basis of fundamental tissue or cell type appears in connection with lesions containing multinuclear giant cells. The true giant-cell tumor of bone stands apart as a clinicopathologic complex from all the other lesions in which one may find multinuclear giant cells. It should not be classified even in the most general way with such other lesions containing giant cells as the fibrous cortical defect, the non-ossifying fibroma, and the giant-cell reparative granuloma. To avoid an alignment which invites confusion among the various lesions containing multinuclear giant cells is important both theoretically and practically.

These examples may suffice to indicate why a more dynamic approach to the primary bone tumors is attained if one turns away from the conventional classifica-

tions and considers each type of lesion as a clinicopathologic entity in its own right. The advantage of this approach is illustrated when one considers juxtacortical osteogenic sarcoma in relation to the common form of osteogenic sarcoma. It is now generally recognized that an osteogenic sarcoma which starts in the interior of a bone (the common form) represents a clinicopathologic entity entirely different from the so-called juxtacortical (or parosteal) osteogenic sarcoma. The latter, while a bone-forming sarcoma, is at first merely oriented to the regional bone in the sense of starting just beyond the confines of the cortex, though later the tumor may erode the cortex of the regional bone and even invade the marrow cavity. The juxtacortical osteogenic sarcoma and the common central osteogenic sarcoma also differ in respect to age incidence and clinical course. Indeed, the former has a much more favorable prognosis than the central osteogenic sarcoma, and even on this basis alone should be held apart from the latter.

Finally, let us consider fibrous dysplasia of bone. This is a lesion in which the basic tissue is a mixture of bone-forming connective tissue and osseous tissue in varying proportions. Should such a lesion be encompassed in the "osteogenic" series of lesions or placed among the "fibrous" lesions? Actually, it does not belong in either category. On the other hand, as a clinicopathologic entity it stands out as a condition which may occur in one, several, or many bones and which may or may not be associated with abnormal pigmentation and/or endocrine disorders.

DIAGNOSTIC APPROACH

When one is viewing the primary tumors and tumor-like lesions of bone as clinicopathologic entities, the clinical findings are helpful in narrowing down the diagnostic possibilities in accordance with the age of the patient, the duration of the complaints, the particular bone or bone area affected, *etc.* The roentgenographic picture is more directly helpful. Indeed, one may well regard the latter as a sort of blueprint (sometimes sketchy and sometimes quite elaborate) of the gross pathology of the lesion. Specifically, the roentgenographic picture: (1) reveals where the lesion is located in relation to the bone as a whole; (2) shows what the lesional tissue has done to the original osseous tissue at the site of development of the tumor; (3) indicates any response which the lesion may be provoking in the perilesional bone area; and (4) often permits one to deduce a good deal of information about the gross character of the lesional tissue itself.

In regard to the original osseous tissue, the *x*-ray picture may show, for instance, "mottled" rarefaction indicating that that tissue is undergoing spotty dissolution. Or perhaps instead, one may find large individual foci of radiolucency reflecting mass dissolution of cortex and/or spongy trabeculæ. As to the perilesional response, this too varies with the nature of the provoking condition. The periphery of a lesion may be rather vague or, on the contrary, may reveal so-called "margination" indicating reactive densification of the perilesional osseous tissue. Then again, the roentgenograph may show that the presence of the lesion is associated with subperiosteal new bone deposition, while another lesion may give no evidence of such a reaction. In respect to the lesional tissue proper, the extent to which the *x*-ray picture shows radiopacity, for instance, represents the extent to which that tissue has undergone calcification and/or ossification.

The histologic tissue pattern remains, of course, the decisive factor in the diagnostic interpretation of any particular lesion. However, in evaluating the pattern in a given case, judgment should be based on the dominant histologic appearances, as determined by examination of tissue areas from various parts of the lesion. One should recognize the subordinate character of local variations in the total

histologic pattern, and also the fact that, here and there in the lesion, tissue elements may be encountered which are not indigenous to it.

In illustration of these general histologic facts, let us consider a tissue section prepared from the actively growing peripheral portion of an osteogenic sarcoma of the common type. In such a section the tissue may be found to be composed largely of cartilage. If it is, and if undue diagnostic importance is attached to this finding, the lesion might be mistakenly interpreted as a chondrosarcoma instead of an osteogenic sarcoma. This mistaken impression may already be controverted by reference to the roentgenographic appearance of the lesion. Furthermore, the age of the patient is also an important consideration. If the subject is an older child or an adolescent, the great likelihood is that the lesion is an osteogenic sarcoma despite the impression created by the tissue section. Indeed, late childhood and adolescence is the period during which osteogenic sarcoma usually appears, except when it occurs as a complication of Paget's disease—a disorder rarely noted in persons under 40 years of age. Chondrosarcoma, in contrast to osteogenic sarcoma occurring *de novo*, appears mainly in persons of middle age, although occasionally it does occur in a young person. Of course, if the lesion is an osteogenic sarcoma, tissue sections taken from other parts of it will show the pattern which is characteristic of that tumor. That is, it will present the pattern of a sarcomatous connective-tissue growth in which the malignant stromal cells have osteogenic potentialities, as demonstrated by the presence of tumor osteoid and tumor bone.

To take another example, the value of correlating the impression gained from the histologic pattern with the clinical and *x*-ray findings stands out rather well in relation to the diagnosis of giant-cell tumor of bone. If one is dealing with a lesion thought to be a giant-cell tumor on the basis of the histologic findings, and if the clinical and *x*-ray findings deviate from those characteristic for that condition, the histologic findings should be re-evaluated in the light of these discrepancies. Consider, for example, a case in which the patient is a child or adolescent and in which the lesion, while in a long bone, is located in the shaft of the bone (somewhat eccentrically) and does not involve the epiphysial end of the bone. In such a case, even if the histologic tissue findings might lead one to conclude that the lesion was a giant-cell tumor, the great likelihood is that one is actually dealing with a non-ossifying fibroma. Indeed, a giant-cell tumor rarely occurs in persons under 20 years of age and nearly always involves the actual epiphysial end of the bone, along with some part of the adjacent metaphysis. Furthermore, the eccentrically located shaft lesion in question is likely to appear rather "loculated" and clearly marginated in the *x*-ray picture. When one once questions the histologic diagnosis in the light of these other findings and re-examines the tissue sections, certain details will now stand out which will turn one away from the diagnosis of giant-cell tumor toward that of non-ossifying fibroma. In particular, it will be noted that the multinuclear giant cells are small, sparse, and often concentrated about small areas of hemorrhage. Furthermore, the stromal cells will be found to differ from the stromal cells of a typical giant-cell tumor in that they are smaller, more drawn out, and often arranged in interlacing bundles.

These examples will probably suffice to make clear the necessity of bringing to the diagnosis of the bone tumors the light which can be shed jointly by the disciplines of radiology, pathology, and surgery. Moving forward in line with this approach, it appears to the writer, however, that, in evaluating a particular lesion, we should try to achieve something more than a collection of individual, compartmented opinions representing strictly each of these three disciplines. That is, we should strive for something more than a joint picture to which each person contributes from his point of view alone, as illustrated by the following diagram.

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