TUMORS
of the
SOFT TISSUES

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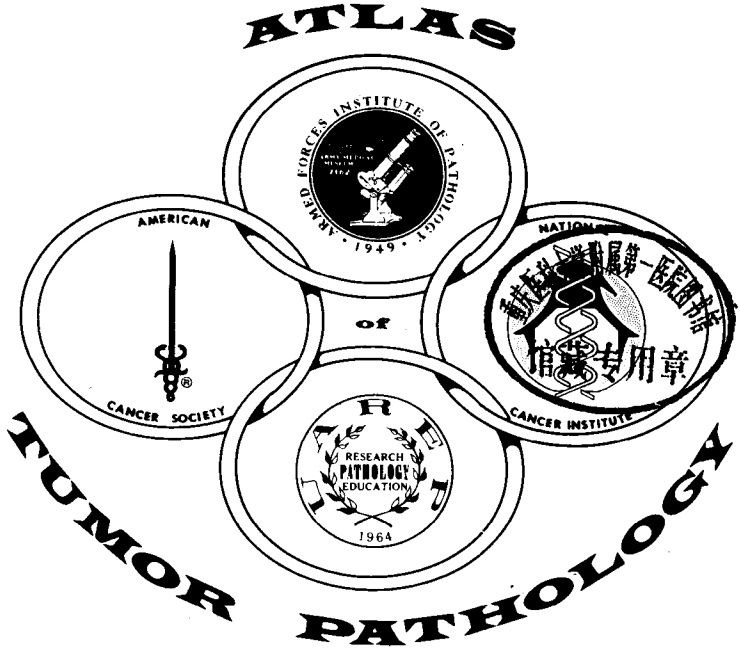

ATLAS OF TUMOR PATHOLOGY

Second Series

Fascicle I

TUMORS OF THE SOFT TISSUES

by

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EDITOR'S NOTE

The Atlas of Tumor Pathology was originated by the Committee on Pathology of the National Academy of Sciences—National Research Council in 1947. The form of the Atlas became the brain child of the Subcommittee on Oncology and was shepherded by a succession of editors. It was supported by a long list of agencies; many of the illustrations were made by the Medical Illustration Service of the Armed Forces Institute of Pathology; the type was set by the Government Printing Office; and the final printing was made by the press at the Armed Forces Institute of Pathology. The American Registry of Pathology purchased the fascicles from the Government Printing Office and sold them at cost, plus a small handling and shipping charge. Over a period of 20 years, 15,000 copies each of 40 fascicles were produced. They provided a system of nomenclature and set standards for histologic diagnosis which has received world-wide acclaim. Private contributions by almost 600 pathologists have helped to finance the compilation of an index by The Williams & Wilkins Company to complete the original Atlas.

Following the preparation of the final fascicle of the first Atlas, the National Academy of Sciences—National Research Council handed over the task of further pursuit of the project to Universities Associated for Research and Education in Pathology, Inc. Grant support for a second series was generously made available by both the National Cancer Institute and the American Cancer Society. The Armed Forces Institute of Pathology has expanded and improved its press facilities to provide for a more rapid and efficient production for the next series. A new Editor and Editorial Advisory Committee were appointed, and the solicitation and preparation of manuscripts continues.

This second series of the Atlas of Tumor Pathology is not intended as a second edition of the first Atlas and, in general, there will be variation in authorship. The basic purpose remains unchanged in providing an Atlas setting standards of diagnosis and terminology. Throughout this new series, the international nomenclature is shown as the first synonym in italics whenever a term has been chosen by the Committee on Tumor Nomenclature of the International Union Against Cancer. Hematoxylin and eosin stained sections still represent the keystone of histologic diagnosis; therefore, most of the photomicrographs will be of sections stained by this technic, and only sections prepared by other technics will be specifically designated in the legends. It is hoped that in many of the new series a broader perspective of tumors may be offered by the inclusion of special stains, histochemical illustrations, electron micrographs, data on the biological behavior, and other pertinent information for better understanding of the disease.

The format of the new series is changed in order to allow better correlation of the illustrations with the text, and a more substantial cover is provided. An index will be included in each fascicle.

It is the hope of the Editor, the Editorial Advisory Committee, and the Sponsors that these changes will be welcomed by the readers. Constructive criticisms and suggestions will be appreciated.

Harlan I. Firminger, M.D.

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The gross and microscopic illustrations in this fascicle have been made by the late Mr. Walter I. O'Neill, photographer of the Department of Surgery, and by Mr. Lewis W. Koster, photographer of the Department of Pathology, Columbia University College of Physicians and Surgeons, with the exception of Plate III–A, which was taken by Dr. Raffaele Lattes.

The tissue cultures were all prepared and interpreted by Dr. Margaret R. Murray, Professor of Anatomy and Director of the Tissue Culture Division of the Laboratory of Surgical Pathology, Columbia University.

Dr. E. B. Powell and Dr. A. J. Cracovaner have kindly given permission for the use of illustrations which were used in previous publications.

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Arthur Purdy Stout
Raffaele Lattes

TUMORS OF THE SOFT TISSUES

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TUMORS OF THE SOFT TISSUES

INTRODUCTION

The manuscript for the first series of this fascicle was completed in 1952, and the intervening years have brought to attention many new features which have shed a great deal of light on some of the tumors discussed in that fascicle. New and unsuspected varieties have also been disclosed. The first series neglected tumors in children almost entirely, on the assumption that they behaved in the same fashion as tumors in adults. The mesenchymal tumors in children have now been surveyed, and they do not always behave in the same fashion; many malignant ones are less malignant than the same types in adults. On the other hand, rhabdomyosarcomas not only have an entirely different histologic appearance but also are more malignant in children than in adults. A great deal has been learned about the true fibroblastic tumors. Very cellular nonmetastasizing fibroblastic tumors are often identified by the term fibrosarcoma. This leads to an entirely unjustifiable picture of the frequency of malignant fibrous tumors inasmuch as fibrosarcomas are actually relatively uncommon.

Much new information about fibrous growths has been recorded in the past 14 years. Generalized fibromatoses of newborn infants have been recognized, the behavior of fibromatoses in children has been studied in detail, and the existence of several pseudosarcomas such as pseudosarcomatous fasciitis and so-called paradoxical fibrosarcoma of the skin have been identified (Kempson and McGavran).

A rare new tumor type, the extraordinary elastofibroma, that until recently has been found only in the general vicinity of the latissimus dorsi and rhomboideus muscles anterior to the scapula, has been recognized (Barr). The study of the cellular origin of certain tumors has made it possible to reclassify a number of them and has facilitated the study of their behavior. The fact that the histiocyte is an unusually versatile cell, sometimes acting as a fibroblast, sometimes as a phagocyte, and sometimes exercising both functions in the same tumor, has made it possible to classify the so-called dermatofibrosarcoma protuberans, the sclerosing hemangioma, and many so-called giant cell tumors of the soft tissues as histiocytic tumors. It will be more satisfactory to call all of these tumors fibrous histiocytomas rather than fibrous xanthomas since the term xanthoma has become firmly associated with the presence of a cell with finely vacuolated foamy cytoplasm containing lipids, and many of the fibrous histiocytomas have few, if any, foam cells. Studying all of these growths together has made it possible to learn that there is a malignant variety of fibrous histiocytoma.

It is now realized that not all fibroblastic cells are derived from pre-existing fibrocytes. On occasion, not only the histiocyte can act as a facultative fibroblast but also the mesothelial cells, synovioblasts, Schwann cells, lipoblasts, reticuloblasts, and probably many others can form reticulin or collagen fibers and assume the shape and appearance of the fibroblast. This versatility has been demonstrated by tissue culture, but with the electron microscope some of the facultative fibroblasts apparently assume the ultramicroscopic features of a true fibroblast and do not retain the features of their true origin. Whatever the explanation, the hypothesis of the

facultative fibroblast is extremely helpful in understanding the complex nature of some of these tumors.

The granular cell tumors have been subjected to an unprecedented amount of study, yet still without any definite decision as to their nature, although it seems certain there are three different and unrelated varieties. It is very probable that they are not myoblastic tumors; therefore, they will be called granular cell tumors in this second series and will be described under a separate heading.

We will do our best to present these complex tumors and their myriad variations in a clear and understandable manner. We realize that the written word and photographic illustrations cannot possibly replace personal experience. The fact that a pathologist in a general hospital cannot hope to recognize all the soft tissue tumors that come his way was brought to the attention of one of us (A.P.S.) during preparation of a report on the bizarre round cell smooth muscle tumors of the stomach. Among 69 cases, some 108 different diagnoses had been made by the pathologists who first encountered them; most of the opinions favored benign tumors but the diagnoses were almost equally divided between benign and malignant.

We would like to clarify our interpretation of certain terms which will be used in regard to tumor growth. If a tumor grows entirely by the in situ multiplication of its cells, it will push the surrounding tissues aside and become enclosed with a fibrous capsule. Very few tumors are truly encapsulated. A second type of growth is by internal expansion and also by slow infiltration. In such cases, the apparent capsule will contain tumor cells. It is obvious that if such a tumor is "shelled out" from within its apparent capsule, some tumor cells may be left behind, leading to possible recurrence. We use the term circumscribed to designate this type of growth. Finally, growth may be by direct infiltration into surrounding tissues and we call this type infiltrative growth. It is important to remember that these vagaries of growth may not determine the occurrence of embolic metastasis. Thus, an encapsulated tumor may metastasize and an infiltrative tumor may not. Almost all the mesenchymal tumors that develop in the skin grow by infiltration but extremely few of them metastasize.

We realize that there are so many varieties of mesenchymal tumors and relatively so few examples of them that it is not possible to predict the behavior of each variety and to use names that will be familiar to all pathologists and surgeons. We will try to convey our meaning by the names we have selected and the observed behavior of each variety, but the subject is still far from an exact science and it is inevitable that mistakes will be made.

In order that one may understand the tumors that arise in the soft tissues of the body, it is necessary to take cognizance of certain facts. If the tumors of the epidermis and the ectodermal structures of the skin and those of the lymph nodes are excluded, all the neoplasms develop from two primitive sources: the mesoderm and the neurectodermal tissues of the peripheral nervous system. From the primitive mesenchyme come the supportive and reticuloendothelial tissues and their corresponding tumors. From the neurectoderm come the Schwannian sheath, possibly the endoneurium, and conceivably the perineurium, which in turn form the prototypes of most of the tumors of the peripheral nerves. If the various tumors developing from these tissues reproduced their prototypic tissues in pure form, even though in various stages of differentiation, recogni-

tion would be relatively simple. Unfortunately this is not always the case, and it is these aberrations which result in the formation of the metatypical conglomerations that are so difficult to recognize. It is well known that the repair which follows injury takes place by the proliferation of fibroblastic cells primarily, accompanied by endothelial proliferations forming capillaries. No other tissues, with the exception of Schwann cells, reproduce themselves with the same facility. It need not surprise us, therefore, to find that many tumors, especially the malignant ones, show a tendency in greater or lesser degree to contain fibroblastic elements. This may be carried to such an extent that these transformed areas are histologically indistinguishable from fibrosarcomas, or the metaplasia may be incomplete so that, for example, lipoblasts, leiomyoblasts, or rhabdomyoblasts may retain their distinguishing characteristics while at the same time acting as fibroblasts and producing connective tissue fibers. This is invariably the case in the synovial sarcoma; it is always made up of two elements inextricably intermingled.

This tendency to form multiple tissues is sometimes carried much further, so that one tumor may be compounded of several more or less differentiated tissues without the predominance of any one type. The mixed mesodermal tumor or mesenchymoma, which is seen in both benign and malignant examples, is produced in this fashion. The best known benign form consists of an admixture of adult fat, blood vessels, and smooth muscle in varying quantities. Malignant mesenchymomas may have an admixture of as many as five different cellular types in the same tumor. The combinations formed are almost endless and no two of these strange tumors are ever exactly alike. Inevitably one is reminded of mixed tumors and teratomas in which are found admixtures of both epithelium and tissues resembling the derivatives of the mesenchyme. These latter develop in certain definite situations such as the salivary glands, male and female genitourinary systems, the breast, and in regions where congenital malformations of development are likely to occur, such as the sacral region. The mesenchymomas may be found in these areas, but they also develop in other situations such as the thigh, where teratomas are unknown.

One might expect that tumors of certain types would arise in areas where corresponding varieties of tissue are normally found, but this is by no means necessarily the rule. It seems to hold for tumors of Schwannian and other neurectodermal cells and because connective tissue, fat, and vascular elements including smooth muscle are almost universal, there need be no surprise if tumors composed of their cells are encountered almost anywhere. Contrariwise, striated muscle, bone, and cartilage are restricted in their distribution; yet malignant tumors composed of rhabdomyoblasts, osteoblasts, and chondroblasts in pure or compound form can develop in situations where normally such tissues are never found.

While the ordinary hematoxylin and eosin stain properly carried out after good fixation will suffice to permit recognition of many of these tumors if one is thoroughly familiar with their vagaries of growth, it will not do for all. It must be supplemented in some cases by differential fiber stains, adequate silver reticulin impregnations, and stains for special substances like lipoid, mucoid or hyaluronic acid, hemosiderin, melanin, elastic tissue, amyloid, and nerve fibers. It is not yet clear whether electron microscopy will be particularly useful in a more exact classification of soft tissue tumors.

If, in addition, one can call upon the aid of tissue culture, obscure tumors can sometimes be elucidated because in most instances, no matter how anaplastic, explanted tumor cells will grow in vitro with sufficient resemblance to their normal prototypes to permit recognition. At the present time this aid is not available in most diagnostic laboratories, and there are very few individuals capable of the proper interpretation of differential tissue growth.

Even with all our knowledge about tumor cells and their growth in vivo and in vitro, there are still a few tumors that can be recognized and named although their exact nature remains obscure. Prominent among these is the tumor originally called myoblastic myoma, now better known as granular cell myoblastoma or myoma. Considerable doubt arose as to the myoblastic origin of this tumor because embryonal myoblasts do not have granules in them and the tumor sometimes grows in places where striated muscle is never found. It has been suggested that these are tumors of histiocytes (Martin et al.), of granular cell fibroblasts (Pearse), induced by parasites (Gullino), of Schwann cells (Fust and Custer), that some of them are paragangliomas (Smetana and Scott), and that they are forms of granular myolysis of muscle and not tumors at all (Roffo). Tissue culture has shown that the cellular outgrowth in vitro most nearly resembles striated muscle but it does not account for the intracellular granules, so that the exact nature of the tumor remains a mystery.

The failure to label the soft tissue tumors properly, especially the malignant ones, has led to a great deal of confusion regarding the distribution, relative frequency, and relative malignancy of many of them. This has been largely responsible for the fact that mesenchymal tumors of the soft tissues of the extremities, especially the malignant ones, are the least understood and probably the most inadequately treated of all tumors.

While the tumors of bone, bone marrow, and peripheral nerves will not be considered in detail in this fascicle because there are separate fascicles devoted to them, it should be remembered that nerve tumors grow in the soft tissues and certain tumors of bone, notably the fibrosarcoma of the periosteum and Ewing's sarcoma, secondarily invade and may form larger tumor masses in the soft tissues than in their tissue of origin.

If one is quite familiar with the usual distribution, relative frequency, and gross growth characteristics of the soft tissue tumors, it is sometimes possible to make an accurate diagnosis on physical examination alone. The surface hemangiomas are familiar to all and the deep hemangiomas or benign vascular mesenchymomas involving muscle often contain phleboliths, which give a characteristic roentgenogram; lipomas of the skin, forming pedunculated growths and lipomas of the subcutaneous tissues, forming soft diffuse and sometimes multiple masses are generally identifiable. The slow growing multinodular so-called dermatofibrosarcoma protuberans has an appearance not often assumed by other tumors. If a tumor is deep, bulky, and nodular, it often proves to be a liposarcoma; if it starts deeply and grows into the skin producing a projecting, dark red, fungating mass, it may be a rhabdomyosarcoma; if it produces a fusiform swelling, movable from side to side but not in the long axis of the extremity, it is likely to be a nerve sheath tumor, whether or not there is any interference with function or sensation; and if it is a subungual lesion producing attacks of paroxysmal pain, it will almost surely prove to be a glomus tumor.

These examples cover a relatively small proportion of the soft tissue tumors and, in most instances, may be imitated by something else. In the vast majority of instances, accurate diagnosis depends upon histologic examination.

In order to give a comprehensive picture of the neoplasms of the soft tissues, it will be necessary to enumerate not only the malignant and benign neoplasms but also the tumor-like lesions about which there is uncertainty whether they are neoplasms. In each instance, a succinct note will describe the important facts concerning each lesion. It is difficult to know where to draw the line in such an enumeration, since there are a good many infections–for example, tuberculosis, sarcoid, syphilis, and rheumatic nodules–which can produce gross lesions closely resembling neoplasms but which can be identified on microscopic examination. These have been omitted.

It is also difficult to know what to exclude from the term soft tissues. Broadly interpreted, it could mean everything except the bones. It is customary, however, to exclude all the organs, epithelial-lined tubes, epithelial structures of the skin, bone marrow, and lymph nodes. This leaves the remaining tissues covering the bones of the head, neck, trunk, and extremities, as well as the internal soft tissues. Most of the latter will be considered in other fascicles, but for the sake of completeness the distribution of some of these soft tissue tumors in the abdominal and thoracic cavities and in the orbit will be included. The benign and malignant tumors will be considered separately.

Although we have dealt with all the benign and malignant tumors of mesenchymal derivation with which we are familiar, the reader need not expect to find recorded here all the tumor forms that can grow. There are many cases of undiagnosed tumors in the files of every laboratory of pathology awaiting future study and recognition. This is particularly true of many puzzling varieties which develop in infancy and childhood. Tissue culture and meticulous cytologic and histochemical technics no doubt will eventually lead to the clarification of those mysteries.

In order that the reader may have some conception of the relative numbers of the different varieties of growths to be discussed in this fascicle, it may be stated that there have been recorded in the Laboratory of Surgical Pathology of Columbia University during the 45½ years from February 1, 1906, to September 1, 1951, 8,686 tumors and tumor-like lesions of the soft tissues, of which 7,337 were benign and 1,349 were malignant. It must be emphasized that almost all the diagnoses have been made on surgical material, and that doubtless the malignant tumors are heavily overweighted because many of them come from other institutions and there is a much greater tendency to request consultations on malignant rather than on benign tumors. It is useful, however, to know something about the relative frequency of the different tumor types in a sequence such as this.