Tumors of the Soft Somatic Tissues

A CLINICAL TREATISE

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A Clinical Treatise

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Preface

This book describes in detail the clinical management of patients bearing tumors of the soft somatic tissues. It is based on the results of twenty-five years of study of the clinical behavior of such tumors. The soft somatic tissues, comprising the organs of locomotion, support, and lubrication and consisting of connective tissue, fat, smooth and striated muscle, fascia, synovial structures, blood and lymph vessels, and nerve tissue, give rise to a large and varied group of neoplasms. The book explains step-by-step how each type of tumor should be handled, from the initial clinical evaluation of the patient to the performance of the biopsy and the institution of the therapeutic plan, and discusses the anticipated prognosis. Each tumor type is analyzed, and criteria are presented for differentiating benign tumors and tumorlike growths from malignant neoplasms arising from the structures under discussion.

The monograph offers an evaluation of the results of treatment of the largest number of neoplasms of the soft somatic tissues ever reported from a single source. The patients in this report have come, in the main, from the Mixed Tumor Service of the Memorial Center for Cancer and Allied Diseases and the Pack Medical Group. Perhaps its greatest value lies in the fact that all were diagnosed and treated by a single group, with treatment policies being subjected to group judgment.

A quarter of a century has afforded adequate time for critical judgment of the effects of different methods of treating sarcomas and to evolve forms of therapy that by statistical analysis have proved to be best for a given patient with a given histologic type of sarcoma in a given anatomic location. Treatment methods have been retained or abandoned, depending upon their respective successes or failures.

A gratifying observation resulting from an analysis of these cases is the definitive curability. The over-all five-year survival rate without recurrence of sarcomas of the soft somatic tissues is 39 per cent.

In our opinion, the choice of treatment should not rest on the pathologist's report alone. Rather, proper therapy depends not only on the histologic criteria but, more importantly, upon the location of the tumor, the extent of its invasion, its mobility, its fixation or adherence to joint capsules, and many other factors that only clinical examination can ascertain. We have found these clinical factors of greater importance, for example, in making the choice between radical local dissection and amputation than is the correct diagnosis of the histogenesis of the sarcoma in question.

The clinician who has treated patients and followed them for many years thereby acquires a fund of information concerning the natural history of a tumor and its response to treatment which, when taken together with the local setting of the cancer and its histologic classification, offers the proper basis for a decision concerning therapy.

As this is the first book on the subject to contain extensive data on the clinical features of these tumors and the results of therapy, we have attempted to make the book all-embracing, discussing the various disciplines that contribute to the cure of the patient. The text is divided into six sections. Section I is devoted to a consideration of the natural history of tumors of the soft somatic tissues, their classification, incidence, hereditary and congenital occurrence, the roles played by trauma and other etiologic factors in their production, the question of malignant degeneration of benign somatic tumors, et cetera. Section II discusses the technics of wide local excision, resection of the tumor and its lymph-node metastasis as a monobloc dissection, and the various types of amputations. In Section III are described at length the different tumors which comprise this neoplastic group, as well as certain tumor-like or preneoplastic proliferations. Section IV discusses sarcomas of infants and children and points out the good results that can be obtained in the treatment of these neoplasms of childhood. Section V describes the principles of treating these neoplasms when they occur in certain anatomic locations such as the neck, the abdominal wall, the buttocks, and the retroperitoneum. Section VI is an over-all summary of the end results of treatment of malignant tumors of the soft tissues, with a critical analysis to evaluate those factors that influence prognosis.

The text is accompanied by 109 tables and more than 600 illustrations. Representative bibliographies at the ends of the chapters provide guidance for those who may seek further information and indicate the works of authors referred to in the text.

We recognize many hiatuses in knowledge pertaining to this cancer group—for example, the inability to classify some soft part sarcomas according to their cells of origin, and the reason why certain apparently benign neoplasms may after many years generate growth momentum and become clinically malignant.

We confess to a strong interest in this subject and have attempted to present in a concise and readable manner this complex branch of oncology. We hope thereby to contribute toward a better understanding of this form of cancer and to a lowered mortality therefrom.

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Acknowledgments

No monographic treatise can ignore the many previous contributions to the subject by the labors and ideas of earlier workers. In this light, one remembers the reply of Lord Moynihan to praise for a speech he had just delivered: "I have gathered a posie of other men's flowers and nothing but the thread that binds them is mine own." Our acknowledgments to our predecessors are best annotated in the selected bibliography that accompanies each chapter.

A complete review of the literature on any medical topic, with critical analysis and summarical presentation, has value as a compendium of information, but only too often it lacks the perspective of original thought and experience. Most of the chapters in this book present original experience and some were written with the collaboration of doctors who were or are now associated with the authors. The names of these collaborators are listed facing the title page. To each and all of them we express our appreciation for their interest and assistance.

Dr. Gordon McNeer and the late Dr. Isabel Scharnagel, senior colleagues on the Mixed Tumor Service of the Memorial Center for Cancer and Allied Diseases, although not writing specific chapters as collaborators, have contributed greatly by the wealth of their clinical experience in the treatment of many of the patients comprising the source material for this book.

We wish to express our grateful appreciation to Drs. Leslie R. Taber, James S. Gallo, and Abram Vermeulen, of the Lendrim Tumor Clinic of the Paterson General Hospital, Paterson, New Jersey, for their cooperation during that period when the authors were more actively associated with the Lendrim Tumor Clinic. Some of the most interesting case reports contributing to this volume were of patients treated at the Lendrim Tumor Clinic.

The medical profession is indebted to Dr. Arthur Purdy Stout for his detailed analyses of neoplasms arising within the soft somatic tissues. It is largely on the basis of his original and critical studies that this group of neoplasms, heretofore often catalogued into a heterogeneous group usually called spindle-cell sarcomas, has been classified into histogenetic divisions on the basis of histologic criteria. We are sure that this book could not have been so specific in its recital of the natural history of these tumors and the correlation of end results with treatment were it not for the classifications so accurately established by Doctor Stout.

The microscopic diagnoses of these soft somatic tumors were made in the Memorial Hospital series by the late Dr. James Ewing and by Drs. Fred W.

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SECTION I

Classification and Natural History of Tumors of Soft Somatic Tissues

Introduction

THE SOFT SOMATIC TISSUES constitute the greatest amount of tissue within the human organism. This mass of flesh, situated between the epidermis and the parenchymal organs, comprises over 50 per cent of the body weight. The muscles alone, of which there are over 400, constitute 40–45 per cent of body weight of the adult and about 25 per cent of the child's weight. A large number of specific tissue types comprise the soft somatic organs. These consist of connective tissue, blood and lymphatic vessels, smooth and striated muscle, fat, fascia, synovial structures, the reticuloendothelium, and others. Offshoots of these tissues penetrate into every portion of the human anatomy. No organ is exempt from their presence.

The soft somatic tissues comprise the form and substance of the body. The height, the weight, and the body conformation are largely determined by the distribution of these tissues. The delineaments of the shape, such as the facial features, are welded in a hereditary pattern by the functional equilibrium of intergrowth of these tissues.

The organs composed of the soft somatic tissues are subjected to injury and disease, as are any other tissues. It is truly remarkable that until recently very few efforts have been made to investigate those diseases to which the soft somatic tissues are subjected. Infectious processes, hyperplastic phenomena, metabolic alterations, degenerative changes, and neoplastic growths may each occur within the tissues which comprise the soft somatic organs of the body.

Great credit belongs to Klemperer, who served to classify and offer a nosologic scheme to such apparently diversified pathologic entities of the soft somatic tissues as rheumatic fever, rheumatoid arthritis, polyarteritis, acute lupus erythematosus, scleroderma, and others.

Certain afflictions of muscles (muscular dystrophy) and involvement of peripheral nerves (poliomyelitis) have focused attention upon the necessity for a better understanding of diseases which afflict the soft somatic organs of the body.

Neoplasms of the soft somatic tissues have, for some unknown reason, received scant attention regarding their nosology, natural history, and methods of treatment. This is truly remarkable inasmuch as this tissue which makes up the bulk of the human body is subjected to a host of neoplastic afflictions which vary from very benign growths to some of the most malignant. Such tumors have frequently been described by the nondescript generic heading of sarcoma and have been lumped into this nosologic repository without effort to identify, further subdivide, and thereby to learn and conquer the various subdivisions.

Certain of the sarcomas have been subjected to careful and critical study because of the interest of certain groups.

Thus, lymphosarcoma, the malignant proliferation of the lymphoid tissue, has been carefully studied because of the interest of the internists in that disease and because of the close relationship of the lymphosarcomas to the leukemias. The recent impetus in the development of chemotherapeutic agents and the initial success attendant upon the discovery of certain agents which have at least a temporary inhibitory effect upon tumors of the lymphoid tissues (the antimetabolites such as folic acid antagonists, 6-mercaptopurine, nitrogen mustard, and others) have produced a resurgence in the interest in these neoplasms.

Great interest has also been manifested in the tumors arising within bone. This has largely been the result of the investigations of certain schools (such as those of Bloodgood, Codman, Coley, Phemister, Jaffe, and others) into this disease process. Trauma to bone and the treatment of fractures have dominated medical thought for many centuries and have probably been factors contributing to sustained investigations of the neoplastic proliferation of bone.

It is indeed regrettable that no such interest has been generated in the tumors which arise within the soft somatic tissues. This dearth of application has resulted in haphazard classifications, in many faulty technics of treatment, and in confusion in the literature pertaining to the natural history of the various subdivisions which comprise this group of tumors. The physician today who is called upon to treat patients bearing neoplasms of the soft somatic tissues is faced with a dearth of information and a lack of proper publications which describe in detail the conduct of establishing the diagnosis, the exact therapeutic modalities to be instituted, and the expected prognosis. No monographs have been published on tumors of the soft somatic tissues. This is surprising in that these tumors are even more frequent than tumors of bone, for which there are a number of good-sized classic volumes. At the Johns Hopkins Hospital, where Geschickter and Copeland have published two editions of their classic book, *Tumors of Bone,* malignant primary bone tumors comprise 1.0 per cent of all cancers; malignant tumors of the soft somatic tissues comprise 0.7 per cent of all cancers. At the Memorial Hospital, where the Coleys have maintained a strong interest in bone tumors and from which institution Bradley L. Coley recently published his volume on bone tumors, there have been 912 patients bearing primary tumors of bone, of which 592 were malignant and 320 were benign. At the same institution, during a 25-year span, there have been 717 sarcomas arising within the soft somatic tissues of the body, a number in excess of the actual malignant tumors of bone.

In addition to the malignant neoplasms of the soft somatic tissues, there have been thousands of patients bearing benign tumors and tumorlike proliferations arising from the soft somatic tissues.

DEFINITION OF SOFT SOMATIC TISSUE TUMORS

The soft somatic tissues are those mesodermal structures which comprise the bulk of the organism and which are usually considered the organs of locomotion and support. They cover the exterior of the body within the skin casement and also form the coverings of the internal viscera, such as the retroperitoneum, the mesentery, and the mediastinum. The parenchymal organs are excluded, as are the bony structures. Inasmuch as previous neoplastic practice has subdivided the diseases of lymph nodes into a separate category, the diseases of these organs, such as lymphosarcoma, giant follicular lymphoblas-

toma, leukemic infiltrations, etc., will also be omitted from this presentation. Diseases of bone marrow, although this is a soft structure within the bony framework, will be excluded, as will tumors of the epithelial structures (skin) except for certain tumors of the mesoblastic structures of skin, such as leiomyoma, dermofibroma, dermatofibrosarcoma protuberans, Kaposi's sarcoma, etc. The neoplasms of nerves will be presented in this discussion, for although the nerves are of ectodermal origin, their anatomy and types of neoplasms, most of which arise from the mesodermal component of the nervous tissues, present characteristics akin to those of the other soft somatic tissues. Certain tumors of nerve endings, such as the glomus tumor, fit into this category. However, the problem of nevi and malignant melanomas, although possibly of either mesodermal or nerve-ending origin, will not be presented here; as this subject requires such a vast coverage, it will form the basis for a subsequent and separate publication.

An understanding of the tumors which arise from the soft somatic structures can be helpful in understanding tumors which arise from the cells of the supporting tissues contained within the parenchymal organs. Thus, leiomyosarcomas are not infrequent within the esophagus, and hemangiomas occur within the liver and kidneys. Any tumor which arises from the soft somatic tissues may be present in any of the parenchymal organs, as each organ will have certain supporting structures as well as neural elements and blood vessel elements within it. For the most part, those tumors within the parenchyma will manifest a natural history similar to those within the bulk of the soft somatic tissues, with certain exceptions such as the fibroma of the ovary producing a hydrothorax (Meigs's syndrome). However, certain neoplasms of the soft somatic tissues which arise in certain of the parenchymal organs occur with such frequency that they have been extensively investigated and will be included in this presentation. Included in this last group are the carcinosarcomas of the uterus, the adenomyosarcoma of the kidney (Wilms's tumor), and the giant follicular myxoma (cystosarcoma phylloides) of the breast.

This volume will discuss both the benign and the malignant tumors which arise from soft somatic tissues. It is necessary also to include certain tumor-like proliferations which are not truly neoplastic entities but either simulate tumors clinically or sometimes develop into tumors (plantar fibromatoses). Therefore, the clinical conduct of patients bearing these entities would be the same as for those with certain benign tumors. The diagnosis of these tumor-like proliferations is mandatory, for reports have been presented of needless amputation of extremities because of the mistaken diagnosis of these entities as sarcoma.

NOSOLOGY OF SOFT TISSUE TUMORS

The interest of James Ewing served to clarify some of the confusion which had existed concerning tumors of the soft tissues. Great credit also belongs to Dr. Arthur Purdy Stout for his painstaking and critical analyses of the numerous neoplasms which comprise this group.

At the combined symposium of the National Cancer Institute and the American Cancer Society, held in Memphis, Tennessee, in 1949, the panel discussions on tumors of the soft somatic tissues were a milestone in efforts to understand better this oncologic entity. This meeting, at which Dr. Stout presided and in which some of the leading students of tumors of the soft somatic tissues participated, made an effort to subdivide vari-