TUMORS of the CENTRAL NERVOUS SYSTEM

by

LUCIEN J. RUBINSTEIN, M.D.

AFIP

ATLAS OF TUMOR PATHOLOGY

Second Series

Fascicle 6

TUMORS OF THE CENTRAL NERVOUS SYSTEM

by

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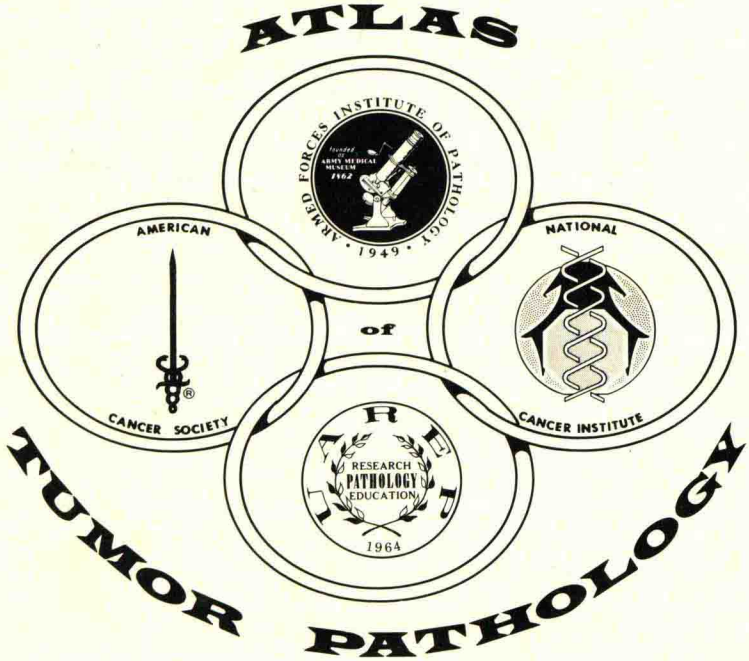

EDITOR'S NOTE

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 brainchild 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 received worldwide acclaim. Private contributions by almost 600 pathologists 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 of the new 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 term chosen by the Committee on Tumor Nomenclature of the International Union Against Cancer is shown by an asterisk if it corresponds to the author's heading, or as the first synonym in bold if it differs from the author's first choice.* 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 biologic 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 author feels that the present International Nomenclature for tumors of the central nervous system is unsatisfactory. The International Union Against Cancer has decided not to reappoint the Committee on Tumor Nomenclature, and the responsibility for drafting an international classification of neoplasms is now in the hands of the World Health Organization. We understand that an effort to develop a more widely acceptable classification for CNS tumors is proceeding at this time. Consequently the Editor chose in this Fascicle to depart from the past practice of highlighting the present IUCC Nomenclature. Hopefully a new, more comprehensive international classification for CNS tumors will be forthcoming in the near future.

FOREWORD

This fascicle of the second series of the Atlas of Tumor Pathology could well be considered a textbook on tumors of the central nervous system.

As in the entire series of fascicles, the illustrations in this one are numerous and excellent; there are more than 400, including 15 color plates. The nomenclature is elaborate, incorporating many of the subtypes of rare neoplasms that have been described. In the almost 30 years since we produced the earlier fascicle on this subject, there have been advances in techniques and methods of study that have helped clarify some of the problems in this field. Some of them have been of aid to the neurosurgeons and irradiation therapists, but still more help is needed for patients harboring many of the gliomas—especially the highly malignant ones.

It has been a pleasure to write the Foreword for this fascicle by L. J. Rubinstein, whose qualifications in the field of neuro-oncology are well known.

James W. Kernohan, M.D.
George P. Sayre, M.D.

Mayo Clinic and Mayo Foundation
Rochester, Minnesota

PREFACE AND ACKNOWLEDGMENTS

The invitation by the Editor and the Editorial Advisory Committee of the Atlas of Tumor Pathology to undertake the Second Series of the fascicle on Tumors of the Central Nervous System presented the writer with a challenging task. The previous fascicle by Dr. J. W. Kernohan and Dr. G. P. Sayre had long established for itself a position of authority in neuro-oncology. Aside from its intrinsic excellence, it embodied the unrivaled experience of the Mayo Clinic workers and represented a school of thought that had gained a considerable measure of acceptance both in this Country and abroad. It would have been presumptuous on the part of the writer to attempt to improve or modify the fascicle of the first series, and he therefore decided to plan the present fascicle as a new work that, in concept and execution, is not designed to supplant the monograph by Dr. Kernohan and Dr. Sayre. The views expressed in this fascicle differ in a number of minor points from those in the previous work, but these differences of opinion are probably more a matter of terminology than concept.

The classification and nomenclature used in this text are those widely adhered to in English-speaking countries. In an attempt to provide some continuity with the previous fascicle, attention has been paid where relevant to the system of grading advocated by the Mayo Clinic workers; this aspect is further discussed in the introduction. This is not to deny that a number of conceptual differences do exist between neuro-oncologists in English-speaking countries and those on the European Continent; attempts to reconcile these divergences by the adoption of various terminologic compromises are premature at this stage, and little seems to be gained by them.

The diagnostic recognition of tumor entities being the primary purpose of this Atlas, a relatively minor role has been assigned to modern tools of investigative oncology, such as electron microscopy, tissue culture, and enzyme histochemistry. Nevertheless, recent advances in the field have made it desirable to outline in the introduction some of the general principles that determine the biologic character of tumors of the central nervous system in man and some of the results of their experimental production as a pointer toward further research. The fascicle is primarily addressed to the resident in pathology, neurology, and neurosurgery, and to the general pathologist who may be confronted by the practical diagnostic problems that are frequently raised by these neoplasms. In this spirit it was thought desirable to include a chapter on the effects of radiation on cerebral tumors and the adjacent brain, and a brief account of the special staining and impregnation technics the writer has found useful in the diagnosis of cerebral tumors; a note on rapid diagnostic methods has also been appended, including an assessment of the brain smear technic. Although this method is sometimes regarded as less reliable than other rapid diagnostic technics, this has not been the experience of the writer, and since it is still employed in a number of neurosurgical centers it cannot be considered obsolete.

The writer is indebted to many colleagues for their guidance. To his teacher, Professor Dorothy S. Russell, he owes most of all.

For their counsels in the planning of this fascicle, he would like to thank the members of the Editorial Advisory Committee, especially Dr. Harlan I. Firminger, and, for their suggestions, Dr. Amico Bignami, Dr. Kenneth M. Earle, Dr. Mary M. Herman, Dr. John J. Kepes, and Dr. William C. Schoene.

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Most of the material used to illustrate this fascicle was studied by the writer at the London Hospital, England, the Montefiore Hospital and Medical Center, New York, and the Stanford University School of Medicine. He wishes to record his thanks to Dr. Harry M. Zimmerman for the use of the Montefiore Hospital material. Dr. John K. Frost of The Johns Hopkins Hospital has generously provided the material and illustrations on spinal fluid cytology. He is also much indebted to Dr. Kenneth M. Earle, Chief, Neuropathology Branch, Armed Forces Institute of Pathology for liberal access to gross and microscopic neuropathologic material from the Armed Forces Institute of Pathology, as well as for the kind hospitality bestowed by him and his department during the completion of this work.

The writer is grateful to the many colleagues who have submitted cases to him for consultation or interest in the past decade. It is impossible for him to name them all, but he would like to record his appreciation of his colleagues at Stanford University for the use in this fascicle of material studied, and in several instances published, by them; they include Dr. Jan Belza, Dr. Adam Borit, Dr. Michael M. Brand, Dr. John H. N. Deck, Dr. Lysia S. Forno, Dr. Jack Griffin, Dr. Mary M. Herman, Dr. Marshall E. Kadin, Dr. William J. Logan, and Dr. William C. Schoene. He would like to thank his neurosurgical colleagues for their cooperation and for making available to him the clinical data on the material he has examined: they include Mr. D. W. C. Northfield, London, England; Dr. Leo M. Davidoff and Dr. Emanuel H. Feiring, New York, N. Y.; and Dr. John W. Hanbery, Dr. James B. Golden, Dr. Daniel Meub, and Dr. Lawrence Arnstein, from Stanford. Outside cases referred for consultation, of which use has been made in this fascicle, were sent to the author by Dr. S. Ry Andersen, Copenhagen, Denmark; Dr. M. Sidney Anderson, Houston, Tex.; Dr. P. V. Best, Aberdeen, Scotland; Dr. Richard L. Davis, Los Angeles, Calif.; Dr. Clarissa L. Dolman, Vancouver, British Columbia; Dr. S. M.

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Finally, the writer would like to express his deep appreciation for the unfailing help and cooperation he has received from Miss Gertrude Fehrlin, Mrs. Audrey Cyr, and Mrs. Margaret Mumford of the editorial staff of the Atlas of Tumor Pathology, and from the technical personnel of the Armed Forces Institute of Pathology.

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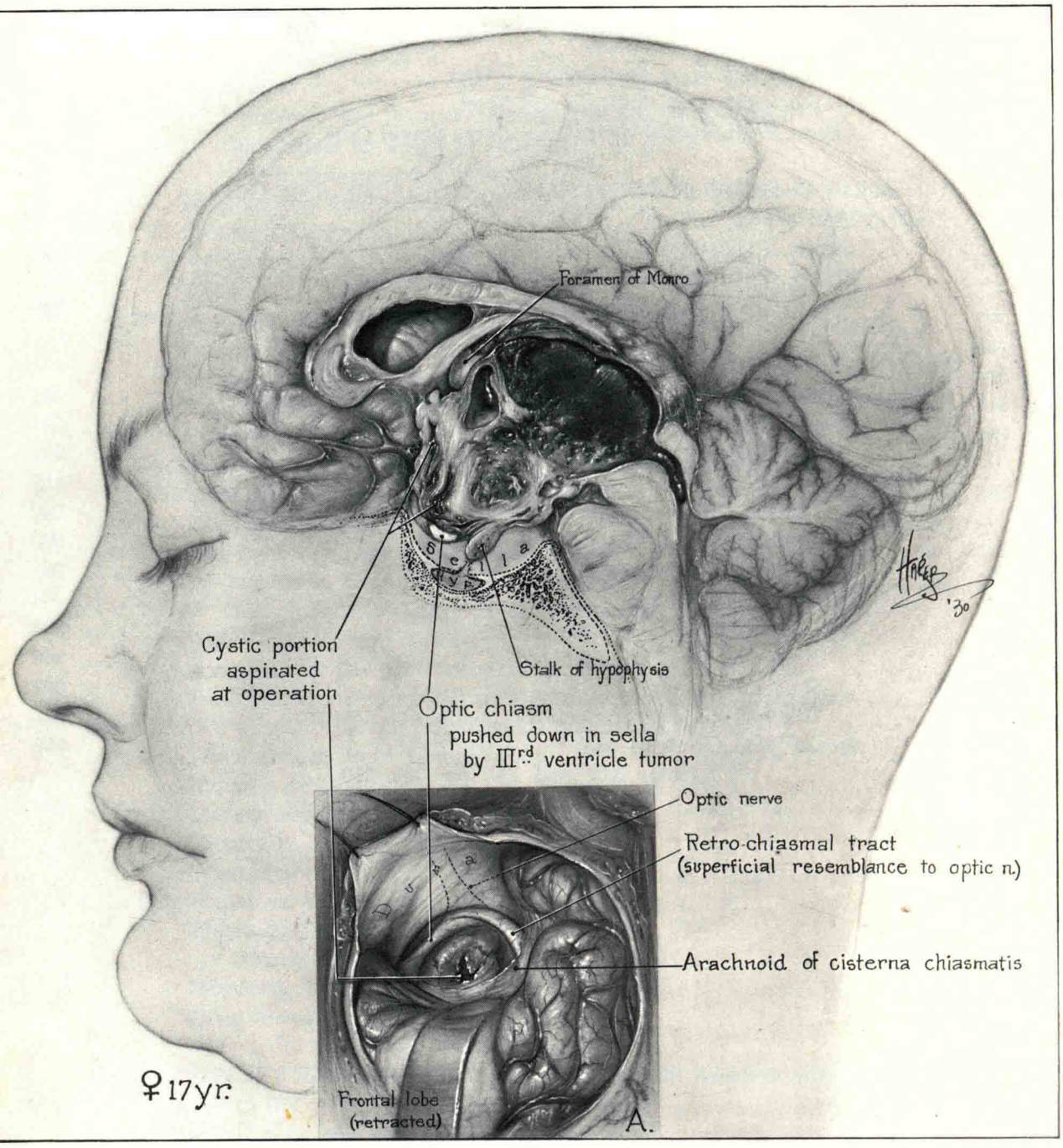
Lucien J. Rubinstein, M.D.



ASTROCYTOMA OF THE THIRD VENTRICLE

This astrocytoma arose in the third ventricle of a 17-year-old girl and caused obesity and obstructive hydrocephalus.

Drawing by Dorcas H. Padget
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The Johns Hopkins School of Medicine

ATLAS OF TUMOR PATHOLOGY

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TUMORS OF THE CENTRAL NERVOUS SYSTEM

CONTENTS

TUMORS OF THE CENTRAL NERVOUS SYSTEM

INTRODUCTION

INCIDENCE AND STATISTICAL DATA

Primary tumors of the central nervous system and its coverings account for about 1.2 percent of all autopsied deaths and for approximately 9 percent of all primary neoplasms. Eighty-five percent of them are found within the cranial cavity.

Among the **intracranial** tumors, those of central neurogenic origin claim priority in number and complexity. These are the tumors derived from the intrinsic, or parenchymatous elements of the central nervous system, excluding the microglia; they are widely credited to account for 40 to 50 percent of all intracranial tumors both primary and metastatic encountered at all ages of life. Following Bailey and Cushing, they have been generally termed tumors of the **glioma** group; in fact, they encompass not only those which are derived from neuroglial elements but also those from neuronal cells and their primitive bipotential precursors.

Second to the gliomas in neurosurgical importance in the adult are the meningiomas (13 to 18 percent), followed by the acoustic nerve schwannomas (8 percent). Pituitary adenomas account for 3.4 to 17.8 percent of intracranial tumors, depending on the neurosurgical centers which attract them. Also the incidence of metastatic carcinomas differs widely: it may be as high as 37 percent in some series and as low as 4.2 percent in others, according to whether the figures emanate from general autopsy material or from neurologic and neurosurgical sources. The high estimate of 32 percent has been computed by Kurland for the resident population of Rochester, Minnesota. In pathologic practice a figure of 15 to 25 percent is probably realistic (Courville).

The central nervous system is the second most common site of primary tumor formation in **children**, who show in this regard a much higher relative incidence than **adults**. This is because neoplasms in adults most often involve the respiratory, mammary, gastrointestinal, and genital tracts, whereas up to 75 percent of the neoplasms in children arise from the hematopoietic, urinary, and nervous (central and peripheral) systems. Primary neurogenic neoplasms comprise from 80 to almost 90 percent of all intracranial tumors below the age of 15 years. On the other hand, those that originate from the sheath elements covering the brain and nerves—meningiomas and schwannomas—with very rare exceptions are tumors of adults.

Another important incidental difference between adults and children lies in the localization of these neoplasms within the cranial cavity. In children 70 percent of all intracranial tumors are **infratentorial;** in adults 70 percent are **supratentorial.**