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MEDULLOBLASTOMA

This monograph, written mainly from a neurosurgical viewpoint, will interest those dealing with the clinical problem of medulloblastoma--the neurologist, the pediatrician, the neuropediatrician, as well as the neuropathologist and the neurosurgeon.

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MEDULLOBLASTOMA

By

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To

my wife, Beverly

PREFACE

THE PRESENT author's interest in medulloblastoma was aroused by Dr. Earl Walker in 1947. Dr. Walker made available the files and pathological material pertaining to medulloblastoma at the University of Chicago Clinics at that time.

Acknowledgment is also made to Dr. William German for permission to review the clinical records and microscopic slides of the cases of medulloblastoma at Yale University. Similar acknowledgment is made to Drs. Gilbert Horrax and James Poppen for making accessible the age and sex statistics of their cases of medulloblastoma at the Lahey Clinic. Dr. Ernest Sachs also graciously made available the age and sex statistics from his tumor file.

Thanks are also extended to Dr. Louise Eisenhardt for permission to review the records and microscopic slides of Dr. Cushing's cases in the Brain Tumor Registry at New Haven, Connecticut. Both Dr. Eisenhardt and Dr. German reviewed portions of the present manuscript and made many helpful suggestions and recommendations.

I am grateful to Donna Nelson of the Department of Pathology at Yale University for typing the first draft of this material; and to the Editorial Staff of the Lahey Clinic, headed by Miss Charlotte R. Thompson, for corrections and typing of the final draft. I am also indebted to Gordon F. Ellerbeck YN2 for checking proof and to Mrs. Mayre S. Swickard for verifying the bibliography and index.

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B. L. C.

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PART ONE

Chapter 1

INTRODUCTION

ANY CLASSIFICATION of disease that is not based on etiological factors will usually have a limited application. In the study of neoplasia there are only a few malignant tumors of which the causative agent is known; and, even in these cases, the basic mechanism, whereby the precipitating factor initiates neoplastic growth, can be described only in broad general terms. This lack of knowledge is nowhere more apparent than in the subject of primary intracranial tumors.

But it became increasingly obvious that intracranial neoplasms, like new growths elsewhere,[120] were made up of many distinct clinical and pathological entities, and that observations regarding one were not safely applicable to another. Therefore, some type of workable classification of intracranial tumors was found to be an absolute necessity. This need was accentuated by the developing specialty of neurological surgery. Although some apparently underestimated the advances of neuropathology,[105] most, if not all, neurosurgeons today would agree that the single most important underlying consideration in treating any patient with a brain tumor is the pathological type of the neoplasm in question. The histology, as well as the location, must be ascertained before accurate definitive treatment can be undertaken or any reliable prognosis given.

A classification of intracranial tumors could not be made based on etiology. Even a satisfactory pathogenic classification could not be made. In many instances, it was impos-

sible to tell from what tissue the neoplasm arose, or whether a "cell rest" mechanism was involved in them all. Different histological types seemed to arise from the same tissue. As an example, the leptomeninges seemed to give rise to endothelial (mesothelial), fibroblastic, hemangioblastic,[25] melanoblastic, osteoblastic, lipomatous, and even sarcoma-

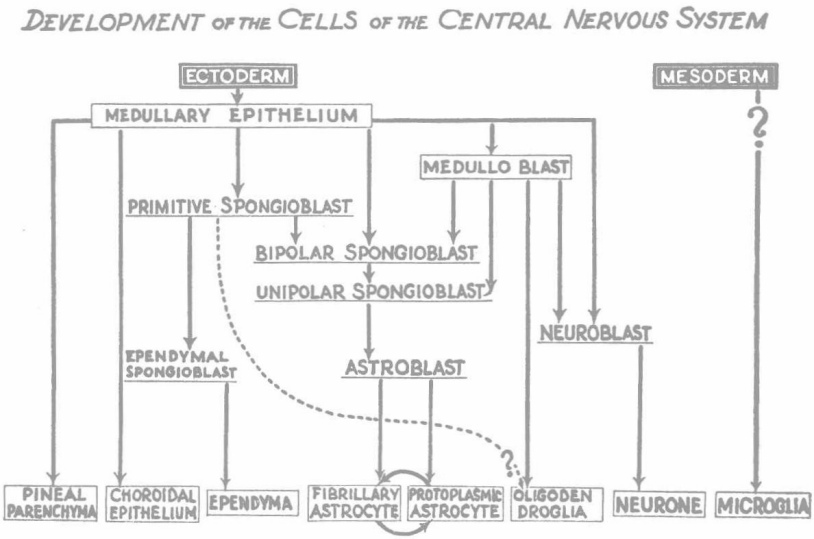

FIGURE 1

(ADAPTED FROM BAILEY)

tous tumors.[37, 101, 149] (This will be discussed further in Chapter V.)

However, in the glioma group of neoplasms, there appeared cells resembling those seen in the brain during normal embryonic formation. Thus in the classification of the gliomas an attempt was made to correlate the appearance of the predominant cells of the tumor with what was known of the cytogenesis of the nervous system (see Fig. 1). Bailey and Cushing proposed a classification of intracranial tumors based mainly on groupings according to microscopic appearance. This classification was found to be valid in that

there was a definite correlation in most instances with gross appearance, location, and the patient's clinical course.[22] But, while this type of classification has served its purpose well, it is based primarily on histological description and thus must have certain limitations.

Bailey summarized the problem[37] by stating that neoplasms have one factor in common—autonomous growth. But otherwise they cannot be treated profitably as a group, including the primary intracranial tumors. The origin, location, structure, symptoms, and treatment of each pathological entity must be studied separately.

The present monograph is concerned with one of these separate entities. The tumor is now most commonly known by the name *medulloblastoma,* although many other terms have been suggested (see Chapter III). This specific single pathological type seems, moreover, to merge with several other pathological varieties (see Chapters V and VI).

In considering any specific neoplasm, it must be treated as an over-all disease entity and not merely a collection of individual malignant cells. Thus in discussing the characteristics of the main cell type or types, one cannot forget the tumor framework—the *stroma* in which the cells live and are nourished. In glioblastoma multiforme, for example, perhaps as intriguing as the tumor cells themselves, is the effect on the blood vessels with characteristic endothelial proliferation. In medulloblastoma, a consideration of the stroma is of interest in the problems of metastasis (implantation) and of reticulin formation (to be discussed in Chapter V).

The *site of occurrence* of the tumor under study and its effect on the surrounding normal host tissues must also be remembered. With the medulloblastoma this is of importance in the theoretical pathogenesis (Chapter II) as well as in any discussion of symptomatology (Chapter IX) or treatment (Chapter X). To avoid confusion, it should be noted

at the beginning that the term posterior fossa is used interchangeably with the terms subtentorial and suboccipital fossa, and does not refer to the area of the occipital lobes.

The present author is a neurosurgeon and not a neuropathologist; but, it seemed that an initial approach from a pathological orientation might be the most fruitful. Hence, the unusual order of presentation.

There is little that is new to be found in this compilation, with the exception of some of the age and sex statistics (Chapter VIII); and there are several excellent previous works which contain chapters on medulloblastoma.[35, 37, 91, 189] However, in these books a larger subject is covered and, in the attempt to be all inclusive, the space afforded any one pathological entity must of necessity be limited. Furthermore, in the older articles, such as Bailey and Cushing's original presentation[21] and Cushing's classic follow-up,[97] much room was quite justifiably allowed for detailed case histories.

It is the present author's intent to take some of the lesser emphasized aspects of medulloblastoma and examine them, in the framework of a theoretical discussion, to see just how this one specific tumor "fits in" with our more modern, but still inadequate, concepts of neoplasia. For detailed case histories and pathological microphotographs, the reader is referred to previous articles.

The ever increasing number of neurological surgeons will mean in all probability that in the future more and more surgeons will be encountering relatively fewer cases of medulloblastoma. This seems likely in spite of the increasing population and earlier diagnosis. The exception will probably be those doing neurosurgery at the larger clinics and at children's hospitals in metropolitan centers. There has been a considerable amount of literature published on medulloblastoma. A relatively extensive bibliography of American writers, as well as some of the older foreign works of histori-