PATHOLOGY OF THE SPINAL CORD

HUGHES

Volume 6 in the Series
MAJOR PROBLEMS IN PATHOLOGY

Pathology of the Spinal Cord

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FOREWORD

THE purpose of this book is to present a comprehensive and yet concise description of the pathological changes of the spinal cord resulting from the various processes affecting the vertebral column and the spinal cord itself. The need for such a book, which covers practically the whole field of spinal cord pathology and correlates the author's wide personal experience with that of other workers in this field, has been urgently felt for some time.

In the course of his career as a pathologist, Dr. Trevor Hughes came to Stoke Mandeville Hospital in Aylesbury, where he became closely associated with the work of the National Spinal Injuries Centre and devoted much time and thought to the pathology of the spinal cord, especially following traumatic lesions. He had the good fortune to continue his studies and enlarge his experience in this complex subject of neuropathology at the Radcliffe Infirmary, Oxford, and has become an expert in this field.

For the clinician who is engaged in the management and rehabilitation of paraplegics and tetraplegics due to injuries of the spine as well as spondylosis, Dr. Hughes' own research on vascular disorders will be of particular interest. One can only hope that these chapters will be widely read and will disseminate knowledge about the hazards and dangers of hasty operative procedures in the acute stages of traumatic lesions of the vertebral column involving the spinal cord.

Apart from the author's personal experience, this book gives an excellent survey of the old and modern literature of the neuropathology of the spinal cord.

After reading through the pages, I warmly recommend this book as a valuable and reliable guide to all colleagues who are engaged in clinical pathology as well as research in spinal cord afflictions.

SIR LUDWIG GUTTMANN, C.B.E., M.D., F.R.C.P., F.R.C.S., F.R.S. *Director, Stoke Mandeville Sports Stadium for the Paralysed and Other Disabled.*

EDITOR'S FOREWORD

THE "Major Problems in Pathology" series is intended to bring to practising pathologists and clinicians up-to-date and critical reviews on the current status of knowledge in a wide range of specific subjects in anatomic pathology. This volume provides pathologists, neurologists and neurosurgeons with a much-needed authoritative reference devoted exclusively to the pathology of the spinal cord.

Pathology of the Spinal Cord, by Dr. Trevor Hughes, was widely acclaimed as a valuable reference when it was published in its original edition in 1965. It is a pleasure to include the second edition, which has been updated by the addition of much new material, in our series. Dr. Hughes who is also author of *Pathology of Muscle* published earlier in this series, is eminently qualified to deal with the broad scope and complexity of this subject. In addition to a vast personal experience in the field of neuropathology, he has a long-standing special interest in the pathology of diseases and injuries of the spinal cord. Dr. Hughes manages to convey his considerable interest and knowledge to the reader with a clear, concise discussion of the subject.

Among the diseases of the central nervous system, those of the spinal cord are particularly troublesome in their propensity for producing diagnostic difficulties for pathologists and clinicians. All those who must deal with the various disorders affecting the spinal cord will welcome this beautifully written practical and helpful diagnostic guide.

JAMES L. BENNINGTON, M.D.

PREFACE TO THE SECOND EDITION

DURING the eleven years which have elapsed since the production of the first edition of this book, the subject of neuropathology has shared in the general expansion of all biological sciences. The enlargement of the subject is reflected in the extensive revision made in the majority of the chapters. However, although much new material has been added, the original object of the book has been retained—that of providing a concise textbook of diseases and traumas affecting the human spinal cord.

Oxford
June 1977

J. TREVOR HUGHES

PREFACE TO THE FIRST EDITION

In writing this monograph on the pathology of the spinal cord I had in mind the needs of two groups of readers. My first aim was to provide a concise descriptive account for the use of neurologists, neurosurgeons, and in particular those clinical practitioners who have specialised with such notable success in the care of paraplegic and quadriplegic patients. But it is hoped that the work will also be of value to a wider medical public including general physicians, and possibly paediatric and geriatric specialists. The second group of readers for whom I have striven to cater are those morbid anatomists who, although skilled in necropsy and biopsy work, find the examination of the spinal cord technically exacting and their histological preparations difficult to interpret. I judge this to be so from the frequency with which cases of this kind are submitted to me for examination. For these readers, I have included a chapter with practical advice in obtaining and handling the necropsy tissues. The spinal cord, when correctly prepared and examined, is the easiest part of the central nervous system to interpret at necropsy. I hope to convince morbid anatomists without special training in neuropathology that, with care and attention to detail, but with only minor variations of their customary techniques, they can obtain excellent results in the examination of the spinal cord.

My list of acknowledgements to those who have assisted me in this work is necessarily a long one. Mr. J. Pennybacker and Mr. J. M. Potter of the department of Neurosurgery, and Dr. W. Ritchie Russell, Dr. C. W. M. Whitty, Dr. J. M. K. Spalding, and Dr. Honor M. V. Smith of the department of Neurology regularly participate in our clinico-pathological meetings at the Radcliffe Infirmary, where many of the points dealt with in this book have been cogently discussed. I am indebted to Dr. D. B. Brownell, Mr. J. Pennybacker, Mr. J. M. Potter, Dr. Honor M. V. Smith and Dr. D. R. Oppenheimer who have all read the manuscript. I have especial reason to thank my colleague Dr. D. B. Brownell who has assisted at all stages of the preparation of the book and has compiled the index. Miss B. Newton, librarian of the postgraduate medical library of the Radcliffe Infirmary, has checked all the

references and procured many journals and in this respect the staff of the Radcliffe Science Library, Oxford have also been extremely helpful. For secretarial assistance I wish to thank Miss Elisabeth Wilson and Mrs. Joan Smith who together have typed the manuscript. For many years Mr. R. Beesley and Miss Eileen Stanley have prepared innumerable spinal-cord sections which have been the foundation of the observations detailed here. For photographic assistance I am indebted to the departments of photography of Mr. E. L. Tugwell and Dr. Parry, my use of the latter being through the kindness of Dr. A. H. T. Robb-Smith, Director of Pathology, United Oxford Hospitals. I am grateful to Dr. L. Guttmann who, during my first steps in Neuropathology at Stoke Mandeville Hospital in 1956, actively encouraged my interest in the pathology of the spinal cord.

Finally I should like to record the courtesy and consideration that I have received from Mr. Douglas Luke during the publication of this book.

J. TREVOR HUGHES

October 1965

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IN the preparation of the second edition, I am again grateful to my clinical and neuropathological colleagues, and have drawn heavily on the resources of my laboratory technical staff, ably led by Mr. Ron Beesley. The Cairns Library of the Radcliffe Infirmary, and the Radcliffe Science Library have constantly traced obscure references and obtained rare journals. All the references cited have been checked by Mr. Ian Kennedy of the Cairns Library. For secretarial assistance I am indebted to Mrs. Joan Smith and to Mrs. Carmel Spicer. Dr. Betty Brownell has kindly revised the index that she prepared for the first edition.

Finally I am grateful again for the continued kindness and help of my publishers Lloyd-Luke in bringing the second edition to press.

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CONTENTS

CHAPTER I

INTRODUCTION

The structure of the human spinal cord is unique in many ways which are reflected in the particular features encountered in its diseases. Although of extraordinary functional diversity the component cells of the spinal cord are morphologically of few fundamental types. The neurones have their cell bodies in the grey matter and send out long axons, which form the white matter. The supporting tissue consists of the three types of glia to which may be added the few ependymal cells forming the central canal. Connective tissue is present in the form of fibroblasts with collagen or reticulin fibres. The connective tissue accompanies the vascular endothelial cells forming the blood vessels in the lumen of which are the variety of cells transported in the blood. This simplicity of component cells means that the variety of reaction permitted to the spinal cord, whether as a sequel of injury or disease, is limited. For an exhaustive treatment of the behaviour of neurones and glia the reader is referred to the larger texts listed among the references given at the end of the chapter. Of particular value are the reviews of Penfield (1932), Cajal (1952, 1959), Glees (1961), Hydén (1967), and Bourne (1968–72); and also the introductory chapters of Vol. XIII of Lubarsch *et al.* (1955–58), of *Greenfield's Neuropathology* (Blackwood *et al.*, 1976), and of Minckler (1968–72). In this chapter I shall mention only those features of recurrent importance in the pathological descriptions that follow.

THE NEURONES AND ITS CHANGES (Fig. 1)

The neurones vary in size, form and function, and all are exceedingly complex cells of whose working we are still largely ignorant. The work edited by Hydén (1967) reviews current knowledge of the neurones and traces the historical development of our understanding of this complicated and specialised cell. Investigation of the metabolism of neurones is clearly of great importance. We can look forward to the application of new techniques, e.g. those of histochemistry (Campa and Engel, 1970), which will elucidate the biochemical processes within the neurone

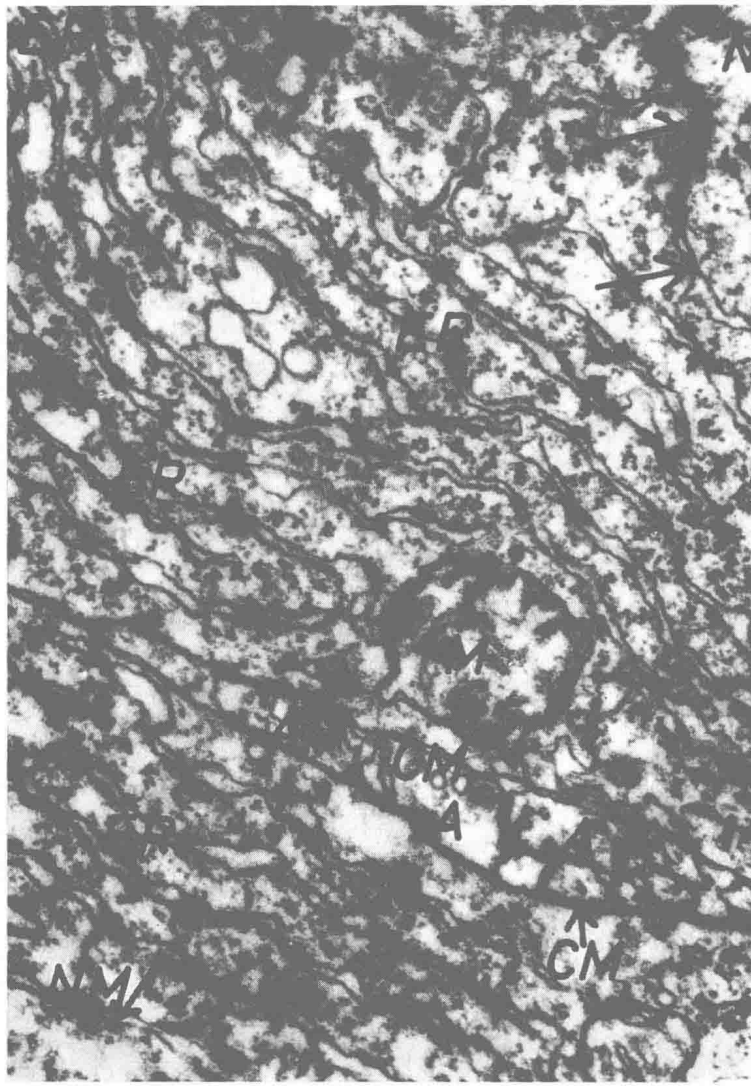

FIG. 1.—Electron microscope picture showing part of two large spinal neurones separated by cell membrane (CM). Between these two cells very fine axons pass (A). Note the laminated construction of the Nissl bodies separated here by a mitochondrion (M). E.R. = endoplasmatic reticulum, covered by ribosome granules which form the subunits of a Nissl body. N = nucleus, N.M. = nuclear membrane. The upper right arrows indicate the tortuous course of a nuclear membrane.

and which may show biochemical differences underlying differences in neuronal function.

Neurones have intense metabolic activity and are unusually susceptible to lack of glucose and oxygen, yet many must endure the life span of a human being, since, as far as we know, they are not formed after the neonatal period. When destroyed the cell body or perikaryon is not replaced and any injury is usually irreversible. When the axonic process is damaged the important change of central chromatolysis occurs in the cell body whilst the distal part of the axon undergoes the degeneration known as Wallerian degeneration. There are also changes in the central part of the axon, i.e. that part between the site of injury and the cell soma, but these changes are much less conspicuous (Cole, 1968).

Central Chromatolysis (Axonal Reaction) (Fig. 2)

This is the most important and distinctive variety of chromatolysis, a term which means the complete or partial loss of stainable Nissl substance from the cytoplasm of the cell body. In the spinal cord it is seen commonly in the motoneurones of the anterior horn and results from injury to the axon within the spinal cord, in the anterior spinal nerve root or in the peripheral nerve. It may also be seen in the large neurone cell bodies of the *nucleus dorsalis* (Clarke's column) when the appropriate axons of the posterior spinocerebellar tract are damaged, and in the smaller neurone cell bodies of the intermediolateral cell column when the preganglionic sympathetic nerve fibres are damaged either when traversing the white rami communicantes or elsewhere. The occurrence of central chromatolysis has provided a well-used method of tracing the origin of nerve tracts both in experimental animals and to a lesser extent in suitable human neuropathological material. The phenomenon itself has also attracted attention and the numerous light microscopical studies (Brodal, 1939; Cole, 1968; Cammermeyer, 1963) have now been supplemented by many ultrastructural observations (Torvik, 1972) and reports on the use of histochemical technique (Means and Barron, 1972). This body of experimental work must be used with caution in considering the cellular process of central chromatolysis in the human neurone. It must be emphasised that the phenomenon differs in detail according to the species, age of the animal, and the particular neurones concerned in the experiment. There is also

a difference between those experiments which use crushing of a peripheral nerve trunk, when regrowth of the axon and recovery of the cell body is possible, and those experiments transecting the peripheral nerve trunk which usually leads to irreversible degeneration of the axon and cell body.

In the experimental animal central chromatolysis begins well within 12 hours, is well developed by the second day and has reached its maximum change in from 2–3 weeks. The most

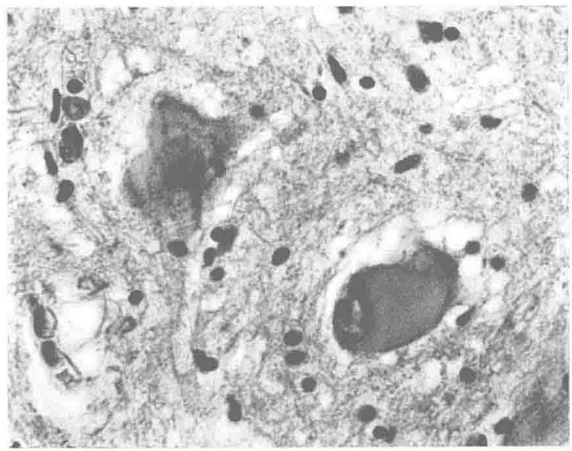

FIG. 2.—Anterior horn motoneurones in a case of peripheral neuritis. The cell body in the right lower quadrant shows central chromatolysis (axonal reaction). Note the disappearance of Nissl substance and eccentric position of the nucleus. Haematoxylin and eosin ×305.

obvious change is that the Nissl bodies disappear from the centre of the cell which swells and becomes more rounded whilst the nucleus is displaced away from the axon hillock to the margin of the cell and may change its shape, becoming oval. Cammermeyer (1963) has produced convincing evidence that some of these changes, notably the rounding of the cell and the displacement of the nucleus, are artefacts which can be avoided by meticulous control of tissue fixation. However the picture described above is that seen in human material and in most tissues of experimental animals.

The sequel to central chromatolysis in experimental animals is either the degeneration of the neurone or the gradual reversal

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