THE NOMENCLATURE OF INTRACRANIAL TUMOURS

TUMOURS of the brain are among the commoner afflictions with which the neurologist has to deal, but their frequency is scarcely realized either by the general practitioner or by the public. The death-rate of 1 in 400 from cerebral tumour given recently by the Registrar-General for England and Wales is a reflection of the opinion of the general practitioner, but there is reason to believe that deaths from primary intracranial neoplasm are at least four times as frequent as these statistics indicate. Thus Weil, in his recent book, quotes statistics gleaned from the post-mortem records of large general hospitals in Vienna and Heidelberg. In the former, during the period 1855-78, primary brain tumours were found in 1.14 per cent. of all post-mortem examinations, and in the latter during the period 1854-1931 in 1.39 per cent. Similar statistics have also been published from England and America, primary cerebral tumours being found in 1.1 per cent. of 18,000 autopsies at the Leeds General Infirmary (Garland and Armitage), and in 1 per cent. of 6,000 autopsies at the Los Angeles Hospital (Courville). These were all hospitals where no special claim was made for the diagnosis and treatment of diseases of the nervous system, and the remarkable accord of the statistics is strong evidence of their veracity.

These statistics explain the increase of interest in cerebral tumours which has taken place during the last few years. While this interest has added something to our knowledge, it has also added a multiplicity of new names coined by the whim or fancy of enthusiastic workers and retained more often in loyalty to their originator, or simply to avoid greater confusion, than for their scientific value. For much of this new terminology there is little if any justification. New names should either connote new ideas or be more appropriate than the old, but in the case of cerebral tumours the advances in knowledge have been few and these largely debatable, while many of the names are neither exact nor suitable.
As regards the most important group of cerebral tumours, that of the gliomas, there is general agreement that the classification of Bailey and Cushing has been of value. Their description for the first time of the oligodendrogliomas represents a distinct advance in knowledge, and for the rest their classification has stimulated research and interest. It has not, however, been immune from criticism. This was directed in the first place against the basis of classification as being too reminiscent of Cohnheim’s ‘cell-rest’ theory. When the defence was made that the names given did not connote any aetiological theory but referred merely to the cell type, the attack was transferred to the names themselves. That of ‘medulloblastoma’ in particular has come under fire, since orthodox views on the development of the nervous system have no place for the ‘medulloblast.’ While American authors as a whole have accepted this name as a convenient designation of a characteristic form of tumour, others have rejected it on the ground that most of the tumours included under this name are neuroblastomas, the remainder being very malignant glioblastomas. Undoubtedly the term ‘medulloblastoma’ and the (French) alternative ‘neurospongioma’ are confessions of ignorance on the point whether the tumour cells are neuroblasts, glioblasts, or both, and while such terminology may have its uses it is apt to lead the uninitiated to think that the origin and nature of the tumour have been finally determined.

Ependymal tumours are also at present the subject of controversy. To the two types in Bailey and Cushing’s classification, Roussy and Oberling add a third, the ‘ependymoglioma,’ whereas del Rio Hortega refuses to classify any tumours as of ependymal origin, naming tumours with similar histological character either ‘glioblastoma isomorpho’ or ‘glioeptihelioma.’ Neither of these authors recognizes the polar spongioblastoma, and only one of them the astroblastoma as a distinct entity. Obviously there is need for much careful work before final classification of the gliomas is reached.

As regards the other forms of primary intracranial tumour, there is less disagreement as to essentials, but much still as to nomenclature. The term ‘meningioma’ is in many respects misleading. If under this name we may include all tumours growing from the meninges, it will include not only the well-known ‘dural endotheliomas’ and psammomas, but also myxomas, meningeal sarcomas and some melanomas. Bailey and Bucy add to this list lipomas and osteomas. Mallory
proposed the name 'meningeal fibroblastoma' for the common solitary meningeal tumours, but it is clear that in spite of its tendency to form collagen this form of tumour arises from, and reproduces the structure of, the layer of special cells which covers the outer surface of the arachnoid, rather than simple fibroblastic tissue. Its origin is therefore not from the dural endothelium, as used to be thought, but from the arachnoidal 'exothelium.' Del Rio Hortega emphasizes this by using the name 'exothelioma,' which has the additional advantage of avoiding all confusion with the endotheliomas which arise from the walls of bloodvessels and lymphatic channels.

A greater divergence of opinion is evident in the naming of the tumours of nerves and nerve-roots. There is a tendency among many writers to restrict the use of von Recklinghausen's name 'neurofibroma' to cases of multiple tumour, and to distinguish between these and the isolated tumours by using for the latter such designations as 'neurinoma,' 'perineurial fibroblastoma,' or 'schwannoma.' The term 'neurinoma' was proposed by Verocay as a means of escape from the question whether tumours of nerves arose from the cells of Schwann or from the endo- or perineurial connective tissue. But it appears to be forgotten that this name was proposed for the multiple tumours of von Recklinghausen's disease and not for isolated tumours. Apart from this connotation, the name has no advantage, except brevity, over that proposed by von Recklinghausen, as it merely uses the Greek term 'inoma,' which Paget had suggested as an alternative to 'fibroma.' Verocay himself called them tumours of 'Nervenfaserzellen,' of which either name is an equally good translation.

The other terms proposed are definitely linked to a theory of origin of these tumours, a fact which tends to confusion among those to whom the exact position of the controversy is unfamiliar. For it is difficult at first sight to realize that the terms 'schwannoma' and 'perineurial fibroblastoma' refer to exactly the same kind of tumour. There appears therefore to be little justification for departing from the old-established name 'neurofibroma,' unless we hold that isolated tumours are essentially different from those of von Recklinghausen's disease. For this view there is scanty justification. Undoubtedly there is great variation in the type of tumours found in neurofibromatosis, some resembling plexiform neuromas full of myelinated fibres, and others isolated fibromas in which it is difficult or impossible to demonstrate nerve-fibres. In some cases one or other type predominates or occurs alone, but
intermediate forms of tumour or tumours of both types are often present. Nor does there appear to be any histological difference between the tumours on the roots of the cranial or spinal nerves in cases with multiple tumours and those which occur in the same situation as isolated tumours. To call such a tumour a 'neurofibroma' if it is multiple, and a 'neurinoma' or 'perineurial fibroblastoma' if it is single, appears illogical, and is certainly confusing.

To the name 'craniopharyngioma' there have been strong and well-grounded objections, and it is surprising that it still has a certain vogue. Frazier and Alpers have recently attacked this term, and have suggested that the suprapituitary epitheliomas should be called 'tumours of the hypophyseal stalk,' and should be clearly distinguished from the simple cysts in this region, for which they suggest the term 'tumour of Rathke's cleft.' Whether it is always possible to make so sharp a distinction seems doubtful, as a small area of epitheliomatous tissue is often present in what are otherwise simple cysts, but the general principle of the suggested nomenclature is sound. Although the name 'tumour of the hypophyseal stalk' carries no suggestion as to the histological structure of the tumour, i.e. whether epitheliomatous or gliomatous, in view of the fact that many such tumours contain both types of tissue this may not be a disadvantage.

These questions of nomenclature have more than an academic interest, since clarity of thought is bound up with language. It is probable that the classification of the future will lay greater emphasis on the malignancy and rate of growth of the tumours, and that it will indicate more clearly which tumours arise from congenital 'cell-rests' and which from normal adult cells. The occurrence of malignant types of such tumours as neurofibromas and exotheliomas, although rare, is well known, and it seems probable that almost any tissue within the cranium may give rise to tumours of varying degrees of malignancy. The proportion of intracranial tumours arising from congenital cell-rests, although perhaps larger than in any other system in the body except the genitourinary, is probably small, and in adult cases may be insignificant.
The Nomenclature of Intracranial Tumours

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