ACUSTICUS TUMOURS.*

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THE appearance within the past five years of two important contributions to the subject of acusticus tumours (I refer to Harvey Cushing's book, and to Mr J. S. Fraser's paper, read before this Section) has left little to be said, and I have no original information to impart, but it may be of some use to lay stress on certain points in the clinical picture, and to make some observations on the functional examination of the labyrinths in the light of the work of Magnus and de Kleyn. To my mind this work has shown the inadequacy of what has hitherto been regarded as a complete investigation of labyrinthine functions, and has revealed in a remarkable manner, and for the first time, the complex character of labyrinthine activities.

In its most characteristic form the clinical course of a case of eighth nerve tumour is well known and easily recognisable. From the fact that the tumour arises on the nerve itself, and on that part of it which lies in the internal auditory meatus, it is not surprising that symptoms referable to the *eighth nerve* should usher in the malady. These symptoms are progressive deafness with or without tinnitus. It is usual to regard tinnitus as an irritative symptom, and, in respect of the cochlear division, we should expect stimulation of the nerve to produce some such symptom. It must be remembered, however, that, in general, neurological experience does not readily favour the idea of "irritation" as an explanation of a symptom which may persist continuously for years, and is associated with a very slowly progressive pathological process. Such symptoms are more commonly found with acute or rapidly progressive lesions, and tend to be intermittent and transient. In this connection we may note that tinnitus is not a constant symptom, and when it does occur it may be long delayed. While, therefore, I have no alternative explanation to provide, we should not assume as a matter of course that tinnitus indicates what is loosely called "irritation" of the auditory branch of the eighth nerve. Nor is it always easy to establish deafness as an initial symptom, for deafness, like unilateral blindness, may pass unnoticed by a patient until revealed by examination. With

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regard to the vestibular division of the nerve, I think vertigo is the only symptom we can definitely attribute to this branch. Only now, in the light of Magnus and de Kleyn's work, are we in a position to differentiate between cerebellar and labyrinthine defect-symptoms with any degree of accuracy, and the evidence indicates that ataxy in movement, nystagmus, and muscular atonia or hypotonia are cerebellar in origin and not labyrinthine. Magnus and de Kleyn have found that throughout the whole animal scale, from guinea-pig to ape, nystagmus is merely an immediate and very transient result of unilateral labyrinth extirpation. The sole permanent symptom is rotation of the head—so that the face points away from the side of the lesion—with some inclination of the head towards the affected side. In man this amounts to turning of the chin away from the lesion, and a lowering of the occiput towards the side of the lesion. This is a relatively common manifestation in cerebellar lesions, and is spoken of as a cerebellar symptom. When we recall the fact that the study of cases of tumour and gunshot wounds provides us with the bulk of our clinical material, it is apparent that in both instances involvement of the labyrinth or of the eighth nerve is to be anticipated, and therefore it seems probable that the so-called cerebellar position of the head is a sign of unilateral labyrinthine defect. Loss of muscle tone does not appear to be a direct result of labyrinthine extirpation, but when present it is due to the rotation of the head, which sets up what Magnus has called a "tonic neck reflex." This reflex, in turn, produces diminution of tone in the extensor muscles of the limbs on the side of the lesion. I would therefore say that deafness, tinnitus, vertigo, and inclination of the head are the symptoms definitely indicative of progressive paralysis of the two divisions of the eighth nerve. It may happen that these four symptoms long antedate the appearance of other focal symptoms, and when this is the case we may rightly speak of an initial or "otological stage" of the disease. So far, despite the exhaustive nature of the tests employed for the functional assessment of the eighth nerve, I have not met with a case of eighth nerve tumour recognised as such during this stage. I do not level this as a reproach at otologists or neurologists, for I believe that it would be very difficult to persuade the surgeon to act upon even the fullest information obtainable at this stage.

The next group of symptoms to appear, when the malady

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follows its usual course, are those referable to progressive paralysis of function in *cranial nerves adjacent to the eighth nerves*, and in the cerebellum. The first of the cranial nerves to show signs of involvement is usually the *fifth*. Here the symptoms may be grouped as irritative and paralytic. Among the symptoms actual pain is not common; the patients complain rather of abnormalities of sensation, such as numbness, creeping sensations in the skin, and so on. In one case under my observation, the patient first sought advice for a distressing feeling of numbness over the right cheek, and, among other diagnoses, that of an infected antrum was made. This may seem a trivial symptom to attract much attention, but anyone who has endured the application of cocaine by a dentist will know how obtrusive and annoying a small patch of anæsthesia on the face may be, and indeed this patient was found to have sensory loss and absent corneal reflex on the right side, physical signs which, had they been looked for, would have betrayed the nervous origin of the symptoms. It is commonly said that diminution or absence of the corneal reflex is the initial defect-symptom in progressive fifth-nerve lesions, and, in the case of which the specimens are shown, this was the only objective sign referable to the sensory division of the fifth nerve within a fortnight of death. Cushing records a similar case. The motor division of the fifth nerve is less commonly affected, though in the case just mentioned I was satisfied that there was slight but appreciable weakness of the masseter on the side of the corneal arreflexia.

Next in importance, when it is present, is involvement of the *facial nerve*. Facial paresis may be a long-delayed sign, and is rarely profound. Generally it consists in slight asymmetry of the lower part of the face on voluntary and expressional movements. Like sixth nerve palsies, which we shall consider next, it is apt to vary from day to day, as was strikingly shown in one case which came under my observation. The patient was a woman with right-sided eighth and fifth nerve lesions and slight right-sided cerebellar symptoms. It was not possible to say definitely whether there was or was not a slight paresis of the right side of the face. However, a diagnosis of acusticus tumour having been made, the question of operative treatment had to be set before the patient, who was extremely upset and wept copiously for the rest of the afternoon. I saw her again just after tea-time, and was

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surprised to observe a profound weakness of all the facial muscles on the right side. By the next morning this weakness had again disappeared. Undoubtedly, the violent muscular activity entailed in her crying had brought out, by a process of fatigue, a latent weakness of the face. Occasionally, irritative symptoms referable to the seventh nerve may be present in the form of clonic spasms of the face muscles, and Cushing records that a diagnosis of Jacksonian fits has been erroneously made and exploration of the crossed motor cortex undertaken to reveal the cause. The mistake may seem an absurd one, but it is by no means so. The twitching of the facial muscles may be so regular in force and rhythm as closely to resemble a Jacksonian fit. In a fatal case of lethargic encephalitis, which came under my observation some time ago, such a clonic spasm of the lower part of the face on the right side was the sole focal nervous symptom present throughout the course of the malady, and it was quite impossible to decide whether its point of origin was the cerebral cortex or the seventh nerve nucleus. Microscopic examination revealed an intact motor cortex, but showed that the seventh-nerve nucleus was the seat of characteristic and well-marked lesions. It must also be borne in mind that signs of a crossed hemiplegia may be present in eighth-nerve tumours, appearing first as slight paresis of the face of upper neurone type; I shall later describe such a case. It is scarcely surprising, therefore, that twitching of the face should be mistaken for a symptom of cortical *origin value in cases where the typical picture of eighth-*nerve tumour is not present.

Sixth-nerve Palsy.—Paralysis of the external rectus is relatively common, but, as it occurs in cases of the kind under discussion, it is almost certainly what Collier has called "a false localising sign." In other words, sixth-nerve palsy here, as in so many other varieties of cerebral tumour, is not the result of direct involvement of the nerve by the tumour, but is a general pressure effect due, according to Cushing, to strangulation of the nerve between the floor of the skull and the anterior inferior cerebellar artery. Its relatively late appearance, and its tendency to fluctuation and transient disappearance, both indicate this mode of origin.

Symptoms referable to the *ninth*, *tenth* and *eleventh nerves* certainly occur, but must be regarded as terminal symptoms, rather than as aids to diagnosis. They are manifested as

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dysphagia and dysarthria, and need not be further considered here.

Next in order of importance are symptoms dependent upon *compression of the anterior part of the lateral lobe of the cerebellum.* These vary in date of onset and in intensity from case to case, but they may be said to be a constant part of the typical clinical picture which we are now considering. They are referred to the side on which the tumour is situated and consist of muscular hypotonia, an inability to perform rapidly alternating movements, a tendency to spread of innervation to muscles not normally taking part in a particular movement and a tendency to error of projection, so that the patient overshoots the mark and may deviate above or below, to right or to left of the object aimed at. In the case of the lower limbs, this ataxy produces a staggering gait and an inability to co-ordinate the component elements in wide movements employing the musculature as a whole, so that the patient loses balance and tends to fall towards the side of the lesion. In the case of the muscles of the head and neck, the same disorder of co-ordination is manifested as nystagmus and defects in articulation. I do not propose to describe cerebellar ataxy in greater detail, or to recite the list of polysyllabic names of Greek derivation by which we have learned to replace a simple descriptive account. I cannot see that such words as "dysdiadochokinesis" have any informative value. On the contrary, they are apt to lead us to suppose that we have really got to the bottom of the nervous disorders which lead to the phenomenon in question, whereas we have not.

In short, the true focal symptoms of an eighth nerve tumour are referable to the eighth, fifth, and seventh nerves and to the cerebellum, and these generally usher in the clinical picture and dominate it throughout its course, and are the basis of clinical diagnosis.

We must, however, consider further the general signs of raised intracranial tension, not only because they form a part of the whole clinical picture, but because, as we shall see, they may dominate it so greatly as to obscure the focal symptoms. In this way they may give rise to a clinical picture widely different from that which we have been considering. In cases of eighth-nerve tumours, general pressure symptoms result from compression and distortion of the brain-stem, which block the exit of cerebro-spinal fluid from the ventricular

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system and thus lead to secondary internal hydrocephalus The symptoms produced in this manner are headache, vomiting and progressive impairment of vision from choked discs. In addition, there may be marked impairment of intelligence, leading to the not unknown error of diagnosing a frontal lobe lesion in cases of cerebellar tumour. As a rule, the true focal symptoms have developed before the effects of general increase of intracranial tension have reached any high degree, but, from time to time, general pressure symptoms dominate the clinical picture from the beginning, and the focal symptoms may never appear to their full extent. Gordon has recently recorded a series of cases of cerebellar tumour, including three of eighth-nerve tumour (so far as can be gathered from his report) in which focal symptoms were conspicuously absent throughout the whole course of the illness, and the impression obtained is that the apparent uniformity of the clinical course of an eighth-nerve tumour, as described by Cushing and others, depends in part on the fact that cases of this lesion are not recognised as such, unless this typical clinical picture is presented. In other words, a somewhat false and deceptive precision in our notions on the subject is apt to develop.

The case from which the specimens exhibited this evening were obtained was of such a nature, and a brief summary of the patient's history of the symptomatology may serve as a corrective to the simple cut-and-dried symptom-complex we have described. The patient was a young woman who was admitted to hospital for a uterine lesion. On admission she made no complaint of symptoms referable to the nervous system, but it was observed that she had considerable headache, appeared to have defective visual acuity, and was occasionally sick. In addition, she was apathetic and distinctly stupid. Examination revealed a chronic otitis media with free discharge in the left ear, which was completely deaf. There was bilateral papillœdema and gross impairment of visual acuity. Repeated examination from day to day revealed two other physical signs, the significance of which I freely admit I had not the courage to face, though their possible meaning did occur to me. These were: absence of the left corneal reflex and slight paresis of the left masseter muscle. The facial movements were normal on the two sides at first, and there was no defect of cutaneous sensation on the left half of the face. Arm movements, articulation, and deglutition were normal. She was not

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taken out of bed to test her gait. During the week following admission, there developed a distinct and constant paresis of the lower part of the right side of the face. On the appearance of this sign a left supra-tentorial decompression was performed, but it gave no relief to the rapidly increasing general pressure signs, and she died comatose.

At autopsy the tumour which is now before us was found. In situation, relations, and appearance, and on microscopic examination, it is clearly a typical eighth-nerve tumour. In addition, there is seen to be some dilatation of the ventricles and secondary internal hydrocephalus.

In this case, the presence of middle-ear disease on the left side blinded us as to the significance of the deafness, while the absence of sensory changes in the face induced me to hesitate to place much reliance on the loss of the left corneal reflex, or on the slight paresis of the left masseter. It is now clear that both were true localising symptoms. Nevertheless, the clinical picture is very different from that described as typical of this lesion and was dominated throughout by the signs of hydrocephalus. The pons is seen to be grossly distorted, and this no doubt was the source of the hemiplegic type of facial paresis observed on the side opposite to the tumour. The distortion of the brain-stem present in this specimen indicates also the high degree of compression that the ascending and descending paths may undergo without giving rise to disturbances of function, and, in general, we may say that variations in the reflexes, in power and in sensation in the trunk and limbs, develop late if they develop at all, and give no reliable information as to the side of the lesion.

There is one other point to be mentioned. I do not propose to describe the characters of the deafness found in these cases. Many of you have had a wide experience of eighth-nerve tumours and can state whatever generalisation may be possible in the matter far better than I can, but the vestibular division of the nerve raises some questions of physiological interest which may be worth brief mention. We may consider the eighth nerve as composed of sensory and non-sensory parts, namely, the cochlear and vestibular divisions. The vestibular nerve belongs to the non-sensory afferent proprioceptive system, and, as the work of Magnus and de Kleyn has revealed, is itself physiologically dual. It has two end-organs, subserving

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separable and distinct functions; these we may speak of as the otolith organs and the semicircular canals. These receptors react to stimuli of different quality and give rise to reflex reactions in the musculature of totally distinct character. The otoliths are stimulated not by movement, but by variations in the position of the head in relation to the horizontal plane. The reflex reactions they evoke are variations in muscle tone, and therefore in attitude, and these variations persist as long as the posture of the head which gives rise to them is maintained. Thus a tonic reflex arising in the otolith organ may persist for months, as Magnus has shown. The semicircular canals, on the other hand, are stimulated by rotation or by movement in a straight line, either vertically or horizontally. The muscular reactions resulting are not postures, but movements which cease when the stimulus evoking them fails. Mr Alexander Tweedie has described these different types of reflex reaction before the Section, and I shall not recapitulate his description. The point I wish to make, and I shall discreetly leave the subject having made it, is that what we commonly accept as a complete examination *of the vestibular division of the eighth nerve is merely an examination of the semicircular canals and cannot be considered as throwing any light on the otolith organs*, the functions of which are not less important. In other words, the work of Magnus has revealed that the labyrinth is a dual *organ physiologically, and that hitherto we have investigated but one aspect of its functions. It seems possible that a careful* investigation of the tonic labyrinthine reflexes might in these cases provide signs of distorted function in the labyrinth at a time when the present repertoire of semicircular canal tests gives us negative results. The elaboration of such an examination technique is in itself a piece of work which would be well worth doing, if for no other purpose than that it would make otologists familiar with the series of delightfully lucid papers, in which Magnus and his collaborators record observations, which have rendered obsolete much of the lore which still passes muster amongst us as the physiology of the vestibular nerve and the labyrinth.

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