Observations ON OCULAR PALSIES*

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We are accustomed in dealing with loss of the power of movement of any part of the body to distinguish those varieties due to structural lesions of, or functional disturbances in, the upper motor neurones which carry impulses from higher centres to the motor nuclei and their nerves, and those the result of lesions of the nuclei and nerves which innervate the muscles directly. The latter are the lower motor neurones or the final common paths, for by them only can nervous impulses reach the muscles.

This distinction of two types of paralysis can be profitably applied to the ocular movements, too, though here, owing to the inaccessibility of the muscles to direct examination, some of the characteristic features of each type of paralysis are not readily recognizable; we cannot, for instance, by the ordinary means of examination detect wasting of the ocular muscles, or test easily their electrical excitability, but the essential differences are obvious, for in lesions of the upper motor neurones movements as such are lost, and sometimes those that normally are excited by certain impulses only, while in disease of the lower motor neurones the muscles themselves are paralysed and react to no nervous message. This can readily be seen by contrasting a left internal rectus paralysis with a palsy of conjugate movement of the eyes to the right, for in the former the left eye is not adducted whether the patient attempts to look to the right or to converge, while in the latter case the left eye comes into the inner canthus as readily and fully as the right when the patient tries to converge; the internal rectus muscle is therefore not paralysed, but fails to contract in a certain movement only, that is, looking to the right. This corresponds to the condition we so frequently meet in an upper neurone paralysis of the limbs, in which no movements may occur in response to voluntary effort, but they can be excited reflexly or synkinetically.

Owing partly to the necessity of an extremely accurate adjustment of the movements of the two eyes in response to numerous stimuli, and partly, perhaps, to the considerable distance apart of the nuclei from which the ocular nerves arise, the nervous mechanisms which control the ocular movements are more complicated, and according to the part of it which is involved two types of supranuclear palsy can be distinguished—the so-called conjugate or associated paralysis in which no movement of the eyes in one or more directions can be obtained by any means, and the true supranuclear palsies in which movement of the eyes in certain directions is lost to certain stimuli only, but can be affected by others.

Whether the regulation and co-ordination of the ocular movements depend on special centres in the brain stem which receive and distribute to the motor cells all impulses that bring about contraction and relaxation of all the muscles concerned in the movement, or whether the distribution of the impulses is determined merely by the anatomical arrangement of the fibres that conduct them, need not be discussed here. There are certainly arguments

in favour of each hypothesis. We can, however, divide ocular palsies into three main and distinct classes:

1. *Paralysis of the ocular muscles*, due to disease of their motor nerves or nuclei.
2. *Conjugate palsies*, in which the movements of both eyes in one or more directions are lost to all adequate stimuli.
3. *Supranuclear palsies*, in which movements of the eyes can be excited by certain stimuli only, but not by other impulses which elicit them under normal conditions.

PARALYSIS OF THE OCULAR MUSCLES

These, owing to their frequency and their importance in general medicine as well as in ophthalmology, deserve first consideration. The muscles which are affected depend on which nuclei or nerves are involved by the lesion. The essential feature is that the muscle is weak or paralysed in all movements in which it may be concerned. As its palsy leads to loss of parallelism in the visual axes, strabismus results, and if the faculty of projection of the image formed on the retina of the squinting eye is preserved, diplopia occurs.

The wide distribution of the ocular nuclei in the brain-stem, and the long and exposed courses of the ocular nerves on the base of the skull, are the chief reasons why they are so frequently damaged by intracranial disease. The nerves are particularly liable to suffer when the membranes on the base of the brain are involved. According to the anatomical site of the lesion these ocular palsies may be divided into: (a) nuclear palsies, in which the nucleus is involved, (b) stem palsies, due to interruption of the motor fibres as they pass through the brain stem, and (c) root palsies, in which the extra-medullary portions of the nerves are affected. It is not, however, always possible to distinguish clinically to which of these three types any case belongs.

Nuclear Palsies

These are most commonly due to local lesions of an inflammatory, vascular, or neoplastic nature involving one or more of the ocular nuclei. It may be a haemorrhage or a softening due to vascular occlusion, or to a tumour growing in or compressing the brain stem. The mode of onset and development of the palsy, and more particularly the other signs of injury of the nervous system which are almost invariably associated with these lesions, usually make it possible to distinguish them. Isolated ocular palsies following on an injury to the head are, however, often not accompanied by other symptoms; in many of these the nerve is torn or bruised in the vicinity of a fracture through the base of the skull, or compressed by a meningeal haemorrhage, but small haemorrhages into the central grey matter of the brain stem not infrequently involve the ocular nuclei, especially that of the third nerve. The explanation of this is probably that the momentum of the blow sets up a wave in the cerebro-spinal fluid of the third ventricle, which breaks with its greatest force against the grey matter around the anterior end of the aqueduct of Sylvius, where the nucleus of the third nerve lies. In such cases the ocular palsy may be the only symptom.

Inflammatory lesions are often more difficult to recognize with certainty. Paralysis of one or more ocular muscles is sometimes due to acute poliomyelitis, and unless it occurs during an epidemic may be difficult to diagnose. Encephalitis lethargica has been, since the recent outbreak of this disease, such a common and well-recognized cause that further reference to it is scarcely necessary, but as during the last few years few, if any, acute cases of this form of encephalitis have occurred in England, at least, a warning may be entered against using this diagnosis as

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a convenient explanation of acute ocular palsies. We still, however, meet occasionally paralysis of the ocular muscles recurring or appearing for the first time in chronic cases of this disease. Acute bilateral ophthalmoplegia due to inflammatory nuclear lesions is an important, though not a common, type. Wernicke originally described it in cases of chronic alcoholism as acute superior haemorrhagic polio-encephalitis, since haemorrhages into the grey matter are the most striking anatomical change, but in two cases which I examined there was a considerable inflammatory reaction as well as small extravasations in the ocular nuclei. It has been described in other intoxications, as in poisoning by lead, sulphurous acid, ergot, phosphorus, and particularly in botulism. In most of these the intrinsic as well as the external ocular muscles are affected, but in food poisoning there may be an ophthalmoplegia interna only.

In contrast to these acute nuclear affections, there is a group of cases of chronic nuclear ophthalmoplegia in which a progressive degeneration of the nerve cells is assumed to be the essential lesion, but few of these cases have been examined anatomically. It generally develops in middle life, and in some of the recorded cases, and in two of my own, there was a definite familial or hereditary tendency. The course is slowly progressive: at first a few muscles only, and frequently the levatores palpebrarum, are paretic, but all the external muscles are gradually involved, but in many cases the intrinsic muscles escape.

Stem Palsies

Stem palsies, due to lesions of the fibres of the roots as they pass through the brain stem, can be usually recognized by the accompanying symptoms produced by involvement of other nervous structures by the causal lesion. In these cases one often meets the various types of crossed hemiplegia, in which there are ocular palsies on the one side, and a hemiplegia, a hemianaesthesia, or disturbances of co-ordination on the other, as in the syndromes described by Benedikt, Weber, Gubler, Millard and others. The chief clinical interest of this type is that it includes the ocular palsies of disseminated sclerosis, for in this disease the root fibres are involved before they leave the brain stem, though possibly in some cases the palsies may be the result of nuclear lesions.

Root Palsies

There can be no doubt, however, that in the great majority of ocular palsies the lesion involves the extra-medullary portions of the roots in their intracranial course. It is here the fibres are damaged in acute and chronic meningitis, in superficial or subarachnoid haemorrhage, in fractures of the base of the skull, by tumours, and by a toxic or infective neuritis.

Lesions of the roots are also responsible for most of the ocular palsies met with so frequently in cerebral syphilis, and probably for the majority of those occurring in tabes dorsalis, though in this disease a primary degeneration of the motor cells, especially of those of the third nerve, has been described. It is well known that while in some cases the tabetic palsies recover under specific treatment, in others they are permanent; the first group may be due to compression or gummatous infiltration of the roots as they pass through the membranes, the second to atrophy of the nuclear cells.

The other more important groups of root palsies are those due to infections or intoxications producing a degenerative or parenchymatous neuritis of the nerves; those the result of raised intracranial pressure; and those associated with cerebral arterio-sclerosis.

Ocular palsies are not infrequent in generalized peripheral neuritis, as in that due to alcohol and diphtheria; in these there is no doubt the ocular nerves, like those of the limbs, suffer directly from the action of a circulating poison or toxin that produces disintegration of the myelin sheaths and other characteristic changes. Similar palsies, usually isolated and transient, have been described in many other infectious diseases, including influenza. They are not uncommon in diabetes, but their pathology in this disease has not been yet definitely established. Whether a true rheumatic paralysis of the ocular nerves exists is open to doubt; not infrequently signs of palsy of one or more of the ocular nerves, or even a complete ophthalmoplegia, follows exposure to cold without other cause being apparent, but the sheltered course of the nerves, far from the surface of the body, makes it improbable that cold can affect them directly. The more likely explanation is that cold merely lowers resistance to some infection or other pre-existing pathological condition. Finally, the palsy of the external rectus muscle, which occasionally follows the use of stovaine and similar drugs for spinal anaesthesia, deserves mention; it seems that the fibres of the sixth nerve are particularly vulnerable to the small amounts of these drugs which diffuse into the intracranial cavity.

An infective or interstitial neuritis of the nerves, apart from meningitis and syphilis, is rarer, but it is occasionally found in association with suppurative disease of the cranial sinuses. The most common form is the abducent palsy in otitis media, due probably, as Gradenigo suggests, to the infection spreading along the air cells and involving the nerve where it passes the apex of the petrous bone in its extradural course. In cases of ethmoiditis in which one of the ocular nerves, in my experience most frequently the third, is paralysed, the lesion is probably a similar infective neuritis. The ocular palsies which have been repeatedly observed in herpes, involving the trigeminal and upper cervical root areas, are probably due to direct infection and infiltration of the nerves.

Various ocular palsies are common in intracranial tumours and aneurysms; they may be due to direct compression of the nerve, as is likely to happen when the tumour lies near the base of the brain, or they may be caused indirectly by compression or by displacement of the brain. The external rectus palsy, which so frequently occurs in cerebral tumours, no matter what their localization may be, is due to displacement backwards of the brain stem and the consequent stretching of the nerve, which is held in position at its exit from the pons by the middle cerebellar artery around which it bends.

Another cause of ocular palsies which is usually overlooked, though in my opinion it is relatively common, is compression of one of the nerves by an arterio-sclerotic vessel. All the three ocular motor nerves lie in close contact with large arteries on the base of the brain; the sixth bends, sharply round the middle cerebellar, and it lies so close to the basilar artery in its forward course that it may be compressed when this vessel is thick and tortuous; the fourth nerve crosses the posterior cerebral, and the third lies in the narrow space between the superior cerebellar and the posterior cerebral arteries as the latter leave the basilar. Further, all three nerves run relatively close to the internal carotid artery in the cavernous sinus. It might be therefore expected that when any of these arteries are dilated or tortuous they might press on one of the nerves sufficiently to interfere with the conduction of impulses through it. This occurs chiefly in elderly subjects with signs of cerebral arterio-sclerosis and raised blood pressure. The palsy usually develops rapidly, is rarely complete, and generally disappears within a few weeks

if the patient is kept at complete rest and iodides or drugs that lower blood pressure are administered.

Ocular palsies associated with pain and sensory disturbances in the area of the ophthalmic division of the trigeminal nerve constitute the characteristic clinical picture produced by disease in the region of the sphenoidal fissure. A squint with diplopia may be the earliest symptom, but frequently the patient first complains of pain in the forehead and behind the eye. The order and the relative severity in which the nerves are involved naturally depend on the exact site of the lesion in the fissure, but usually a complete ophthalmoplegia results, and cutaneous sensation in the area of the ophthalmic nerve is lost or greatly diminished. A syphilitic periostitis is one of the most common causes of this syndrome, but it occurs also with tumours and inflammatory lesions in the region of the sphenoidal fissure. In many cases, however, the nature of the lesion cannot be determined. Except when the lesion is syphilitic and the patient can be treated early, the prognosis as regards recovery of the ocular movements is usually bad.

CONJUGATE OCULAR PALSIES

This is the term usually applied to loss of the power of movement of the two eyes in any direction, provided this is not due to the chance association of paralysis of the prime movers of each eye. It is characterized by the facts that the loss of movement develops equally and simultaneously in the two eyes and squint and diplopia consequently do not occur or are subsidiary features, and that though in attempting the movement the palsied muscles do not contract they may act normally in some other movement; the left internal rectus, for instance, may fail to move the eye inwards on attempting to look to the right, but functions normally on convergence. It is therefore a palsy of movement rather than of muscle.

The most common form is perhaps paralysis of the lateral movements of the eyes to the one side. This frequently results from disease in the lower and dorsal portion of the pons varolii in the region of the nucleus of the sixth nerve. When unilateral the patient is unable to look towards the side of the lesion. In such a case the internal rectus of the one eye and the external rectus of the other fail to contract, and as these muscles are innervated from two separate nuclei about 3 cm. apart, it is necessary to assume that they receive branches from the same fibres, or are connected by association fibres which distribute the same impulses to the cells of both. As in conjugate palsies the muscles fail to respond to all impulses, to those from the vestibular and other mechanisms in the brain stem and from subcortical centres, as well as to cortical messages, the first hypothesis necessitates the assumption of a very complicated arrangement of fibres. On other grounds, too, it seems more probable that conjugate lateral deviation depends on association fibres between the sixth and third nerve nuclei, and we know such exist in the dorsal longitudinal bundles. These tracts have undoubtedly an important part in the co-ordinated movements of the eyes. It is beyond doubt that the impulses which can effect conjugate lateral movements converge towards the region of the sixth nucleus, those from the forebrain passing by the nucleus of the third nerve in their downward course. From here part of them are shunted back to the nucleus of the opposite internal rectus along another path in the dorsal longitudinal bundles.

The exact position which a lesion must occupy to cause a conjugate palsy of lateral movement of the eyes to the same side has not been definitely determined; according to some it must involve the sixth nucleus, but it is more

probable that the controlling mechanism lies, not in the abducens nucleus, but oral, and slightly ventral to it. It is here that all the paths conveying impulses for lateral movement to the same side converge, and a lesion in this position, therefore, abolishes reflex as well as voluntary deviation of the eyes in this direction. As this mechanism lies near the middle line of the brain stem, a single focus of disease often affects lateral movements to the two sides, but the patient can still deviate his eyes upwards and downwards, and converge normally.

Occasionally, a lesion involves the associational fibres connecting the sixth with the third nerve nucleus; then, on attempting to look to the same side, the ipsilateral eye deviates outwards, but the contralateral eye does not pass the middle line, though it rotates inwards in convergence.

The conjugate vertical movements may be similarly abolished, either together or separately, or convergence may at the same time be impossible. The exact positions lesions must occupy to produce these conjugate palsies of vertical movement have not been definitely ascertained; according to some authors upward and downward movements are lost when the anterior quadrigeminal bodies are injured, but others attribute these palsies to lesions in the tegmentum of the anterior part of the midbrain. There is, however, considerable evidence that the mechanism for elevation of the eyes lies more anteriorly than that for depression. As the power of convergence is frequently lost in association with palsy of downward deviation, we may assume that the nervous apparatus that control these two acts lie close together.

SUPRANUCLEAR OCULAR PALSIES

While in conjugate palsies no movement of the eyes in one or more directions can be obtained by any means, in the so-called supranuclear palsies the eyes fail to move in response to certain stimuli only. It is often assumed that the movements of our eyes are largely volitional or under control of our will, but these represent a small part only of the ocular motor activities; impulses which determine ocular postures and movements are constantly flowing in from the vestibular nuclei in the brain stem, from the lower visual and auditory centres in the midbrain, from the occipital cortex and from other sources. It happens not infrequently that movements in response to one or more sets of these impulses only are lost, while those excited by other impulses are preserved. In the most common type the patient is unable to deviate his eyes in one or more directions when he is ordered to do so or asked to look towards an object held in these directions, but these movements can be elicited by labyrinthine stimulation and other means. As a rule, when the power of voluntary movement is abolished the eyes can follow a slowly moving object which the patient has fixed, and if while he fixes his eyes on an object his head is moved actively or passively towards one side, his eyes still remain on the object—that is, they rotate in the direction opposite to the movement of his head. Following objects with the eyes, and the compensatory deviations that occur on movement of the head, are largely reflex acts, and dependent on other paths than those which effect voluntary movement. We must therefore assume that in the supranuclear palsies the lesions interrupt some of the afferent paths to the conjugate mechanisms, but leave the latter unaffected, and the available pathological evidence points to this conclusion.

Other forms of dissociation are occasionally met with, but, owing to rarity and their small clinical importance, further reference to them here is unnecessary.