INTRACRANIAL PATHOLOGY AND DIPLOPIA

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SUMMARY

Diplopia, as a common symptom of intracranial pathological processes is defined. Monocular diplopia, the least common variety, is often hysterical in origin but may be a sign of a cortical or peripheral visual pathway disturbance. Muscle paretic diplopia is the clinical entity commonly encountered and has great diagnostic value if one considers that ocular nerves may be involved directly by intracranial pathological processes or in a distant dislocation effect due to raised intracranial pressure. In the latter instance diplopia may have false localizing value. A brief description of the origin, course and relationships of the ocular nerves as well as their actions, is given. The common pathological processes involving these nerves are briefly described. The less common varieties of diplopia: that due to visual field loss and the type produced by failure of convergence are also considered.

When the same object gives rise to two images in consciousness with the result that a second similar image is seen to one side of the fixation object, we speak of diplopia. Stated simply: 'One object is seen as two'. In general, it may be said that diplopia is caused by the stimulation of non-corresponding retinal points by the same object. The fixing eye will have its image falling on the macula and the deviating eye will have its image falling on an eccentric point in the retina.[1] The image from the deviating eye is not as clear as that from the sound eye, as it is projected from the extrafoveal retina.

Three basic requirements have to be fulfilled before one can have any form of diplopia. Firstly, the patient must be conscious to be able to appreciate this subjective phenomenon. Secondly, he must have sufficient visual acuity in both eyes to have binocular diplopia. Obvious as this may seem, it tends to be forgotten at times in everyday practice. Thirdly, the patient must be able to communicate clearly his subjective disturbance of vision. A child or aphasic patient cannot do this. Diplopia is usually binocular but may be uniocular. When both are combined in the same patient, triplopia or even quadrilopia may result and at the same time many of the causes of monocular diplopia may produce multiple images (polyopia).

Monocular Diplopia

This condition is uncommon and present when diplopia appears or persists when only one eye is opened.[2] It may occur in a wide diversity of conditions of ophthalmological (peripheral) or neurological (central) nature. We will not concern ourselves with the peripheral causes here. One must utter a warning, however, that in the absence of a

peripheral lesion, monocular diplopia should not be labelled as hysterical. It often is a hysterical symptom but this is by no means absolute. The commonest central cause of monocular diplopia or polyopia is homonymous hemianopia due to cortical lesions. Monocular diplopia in a patient with homonymous hemianopia is almost diagnostic of a lesion of the calcarine cortex.[3] We have seen one such case following local cortical injury. Bender[4] also described this condition as well as monocular diplopia occurring with chiasmal involvement by pituitary tumours.

In these cases a new central functioning area or pseudofovea may be established in the surviving part of the field around which spatial perceptions are orientated. If this shift is incomplete, double vision may result. There may be a cerebral dissociation allowing the independent activity of areas of the cerebral cortex which normally act in association to produce a single sensory impression.

Long-standing convergence paresis in epidemic encephalitis as a cause of monocular diplopia was first described by Gerstmann and Kestenbaum.[5] Monocular diplopia in these cases was restricted to near objects, i.e. objects closer than the pathological near point of convergence. It occured, therefore, first in that region in which diplopia could be expected because of the convergence paresis. The authors explained this phenomenon on the basis that retinal points have been provided with new space values in order to adapt themselves to the convergence paresis. The simultaneous perception of these new space values and the old ones gives rise to monocular diplopia.

As indicated, monocular diplopia may be a hysterical symptom.

Muscle Paretic Diplopia

Paralysis of nerves of ocular motility may be the manifestation of two intracranial processes: A remote effect due to raised intracranial pressure resulting in brain herniation or distortion and as such may have no localizing value. Such paralysis of eye muscles which occur in later phases of the evolution of intracranial disease have to be interpreted with considerable caution as they may be false localizing signs (third and sixth nerve paralysis in particular). On the other hand ocular nerve palsies may occur with certain lesions situated basally and directly involving these nerves in the pathological process and may then become of extreme importance in the clinical localization of the site of the lesion.[6]

Almost all patients with paralytic squints have diplopia but in the milder cases it may be limited to a small portion of the binocular field corresponding to the field of action of the muscle involved. The fusional amplitude may be sufficiently great, however, to overcome the deviation and keep it latent, as is seen particularly in a horizontal divergent squint. When a squint develops before the binocular reflexes have become established, the diplopia is not obtrusive despite the almost invariable retention of good vision in both eyes. Several mechanisms are utilized for accomplishing this which need not be discussed here On the other hand in squints developing after the binocular reflexes have become established, diplopia is usually a very distressing symptom, and, indeed, is usually the symptom which initially makes the adult aware of his condition. This subjective disturbance is always greater at the onset of the squint but most patients succeed eventually in adapting themselves to some extent to it.[1] It is the detection and objective evaluation of disturbances of motility, even when latent, and closer analysis of this which may yield the only information on which a diagnosis can be based. Double vision may be detected in the initial tests of eye motility in the six cardinal positions. A latent diplopia may be missed and it is specially tested for with red/green glasses by the patient's bedside or in the outpatient clinic but merits full ophthalmological investigation.

APPLIED ANATOMY OF NERVES OF OCULAR MOTILITY

The six muscles which move the eye are supplied by three nerves: oculomotor, trochlear and abducens.

Oculomotor Nerve

The nuclei of the third nerve are situated in the ventral peri aqueductal grey matter at the level of the superior colliculus. From this nucleus the nerve fibres pass ventrally through the medial longitudinal fasciculus, red nucleus and substantia nigra to emerge on the medial aspect of the basis pedunculi. The nerve then extends forward through the interpeduncular cistern between the superior cerebellar and posterior cerebral arteries and maintains a close relationship to the posterior communicating artery. It passes through a small triangular area in the dural roof of the cavernous sinus lateral to the posterior clinoid process, to run in the lateral wall of the cavernous sinus which it leaves through the superior orbital fissure to enter the orbit. It supplies all the ocular muscles except the superior oblique and lateral rectus.

Trochlear Nerve

The nucleus of the fourth nerve is situated along the dorsal border of the medial longitudinal fasciculus at the level of the inferior colliculus. Its fibres run dorsolaterally round the edge of the peri-aqueductal grey matter at the level of the caudal end of the inferior colliculus. All the fibres decussate and emerge on the dorsal aspect of the midbrain just below the lower border of the inferior colliculus. The nerve passes ventrally round the lateral aspect of the midbrain between midbrain and temporal lobe, runs along the undersurface of the tentorium cerebelli, to which it is adherent, and crosses the free border of the tentorium cerebelli immediately lateral to the third nerve where it pierces the dura to gain a position in the lateral wall of the cavernous sinus just below the third nerve. From here it passes forward through the superior orbital fissure to enter the orbit on its way to the superior oblique muscle, which it supplies.

Abducens Nerve

The nucleus of the sixth nerve lies in the floor of the fourth ventricle near the midline, at a caudal pontile level. Its fibres pursue a strictly ipsilateral course and extend ventrally to emerge from the brainstem near the midline at the junction of pons and medulla in close relationship to the anterior inferior cerebellar artery. The nerve then passes forward upwards and laterally and pierces the dura to enter Dorello's canal. This canal is in close relation to the posterior wall of the sphenoid sinus and is bounded by the posterior clinoid process, the apex of the petrous bone, and the petroclinoid ligament. Here the nerve lies on the apex of the petrous bone before it enters the cavernous

sinus in closer relationship to the internal carotid artery than the other nerves and also leaves the sinus through the superior orbital fissure.

In summary of the anatomy, one may therefore say that the three nerves subserving ocular motility all arise in paramedian nuclei in the midbrain or pons, and from these nuclei nerve fibres run an intramedullary course before they leave the brainstem to enter the subarachnoid cisterns. They all pierce the dura and come to lie in the cavernous sinus where they have a close relationship with the carotid artery and the three divisions of the trigeminal nerve. They leave the cavernous sinus through the superior orbital fissure.

GENERAL CHARACTERISTICS OF PARALYSIS OF THE OCULAR NERVES

It stands to reason that an isolated paralysis of the fourth or the sixth nerve may occur, giving rise to isolated paralysis of the superior oblique and lateral rectus respectively. An isolated paralysis of an individual muscle supplied by the third nerve must be extremely uncommon, because a nuclear lesion must be very small and circumscribed to produce such an effect as an isolated phenomenon. When it does occur, it is usually due to a minute infarct.

Paralysis of the Third Nerve

When complete, there is very little difficulty in recognizing this characteristic entity. The ptosis on that side is of a severe degree and the resting position of the eye is one of lateral deviation due to unopposed action of the lateral rectus and slight downward deviation due to the action of the superior oblique, but this may be minimal due to the inefficiency of the superior oblique in abduction. The pupil is widely dilated and not responsive to light. Variations on the theme are, however, numerous due to different degrees of severity of the palsy and presence or absence of pupillary involvement.

A patient with a third nerve palsy usually turns his head away from the paralysed side in an effort to compensate for the abnormal position of the eye. Two practical points need be emphasized: a rapid paralysis of all the muscles supplied by the third nerve is almost always due to a lesion involving the nerve in its subarachnoid course. On the other hand rapid paralysis of a single extra-ocular muscle supplied by the third nerve is very rare and, when it does occur, it is due to destruction of part of the third nerve nucleus by an infarct. Progressive paralysis of extra-ocular muscles without intra-ocular muscle palsy may result from a peripheral third nerve lesion, for example an involvement of the nerve by a pituitary chromophobe adenoma.[7] Usually, however, it is due to a nuclear lesion. Walsh and Hoyt[8] state that in most cases of 'pupil-sparing' third nerve palsy, the cause is diabetes. In my own experience it has been impossible at times to assign a lesion to a definite level merely on the basis of intra-ocular and extra-ocular involvement. In testing for diplopia in a patient with a complete third nerve palsy, one has, of course, to elevate the eyelid to do so. The patient will therefore not subjectively be conscious of diplopia due to the fact that vision in the affected eye is obscured, bringing us back to our basic requirement of at least normal vision in both eyes to be able to have binocular diplopia. A practical point which need perhaps not be laboured, is that anything done to elevate this lid permanently would be pointless, because the patient then ends up with a troublesome diplopia.

Fourth Nerve Paralysis

The characteristic disturbance of motility is a slight deviation of the affected eye upwards and outwards. This becomes more marked in adduction and attempted downward gaze. The diplopia is vertical with maximum vertical divergence gazing with the affected eye inwards and downwards. There is a turning and tilting of the head to the opposite side. Isolated paralysis of this nerve is uncommon despite its long intracranial course probably due to its protected position under the edge of the tentorium.

Sixth Nerve Paralysis

Characteristic disturbance of motility of the eye is an inability to look laterally, in other words, failure of abduction of the affected eye and there is diplopia on attempted abduction of that eye with maximum divergence of the image on lateral gaze. If there is complete paralysis the eye has a resting position of convergence. In this position there may be over-action of the inferior oblique and elevation of the eye because of the maximal efficiency of this muscle with the eye in adduction.

PATHOLOGY OF CRANIAL NERVE PALSIES

The course taken by the ocular nerves can be divided into intramedullary, subarachnoid, cavernous and orbital. The cause of conditions affecting these nerves can be readily grouped together on this anatomical subdivision.

Intramedullary Palsies

Usually supranuclear lesions give rise to gaze paralysis and therefore the eyes move in unison and diplopia is not experienced, but in upper brainstem lesions such as pineal tumours individual eye movements may be affected and diplopia produced. Nuclear palsies and intramedullary fascicular palsies are very rarely isolated due to the compact nature of the brainstem where these nuclei and roots are situated, and they are therefore usually associated with other evidence of damage to associated cranial nerves or long tracts giving rise to the so-called crossed hemiplegias characteristic of brainstem lesions. Such lesions affecting the brainstem may be traumatic, of vascular occlusive nature, but angiomas of the brainstem are not uncommon and should be remembered when there is a progressive disorder of the brainstem with intermittent symptoms indicating nuclear and long tract involvement.[9] Gliomas of the brainstem affect the nuclear and long ascending and descending tracts in a slowly progressive manner but, contrary to what one might think, there may be no evidence of hydrocephalus until fairly late in the disease. Virus infection can affect the brainstem, particularly in the entity of brainstem encephalitis[10] but also in more diffuse forms of encephalitis, and produce ocular palsies as a prominent feature but in association with a decreased level of consciousness.

Subarachnoid Cistern Palsies

In the subarachnoid space nerves are liable to be affected by such diffuse processes as meningitis and in particular syphilitic basal meningitis and tuberculous meningitis. In

other forms of septic meningitis ocular palsies may occur due to the associated endarteritic process. The abducens nerve in its very long exposed course is particularly liable to distortion associated with displacement of the brain-stem secondary to raised intracranial pressure and isolated paralysis of this nerve has very little localizing value. The reason for this involvement of the sixth nerve has given rise to many speculations. Cushing[11] thought that the nerve was compressed by a taut anterior inferior cerebellar artery or internal auditory artery. Compression against the sharp petrous ridge has been blamed[12] or sharp angulation where it pierces the dura and consequent impairment of blood supply of the nerve.[13] Tumours arising from the clivus, or from the basilar part of the pons, and aneurysms of the basilar artery may produce bilateral sixth nerve palsies usually with bilateral spasticity.

The third nerve is in particularly close relationship to the posterior communicating artery and the terminal basilar where it emerges between the superior cerebellar and posterior cerebral arteries in the interpeduncular fossa. It is liable to be affected by aneurysms of the terminal carotid in its immediate supraclinoid division, the posterior communicating artery or the terminal basilar.

Terminal carotid aneurysms may expand medially to involve the optic nerve or chiasm or laterally to involve the third nerve. The third nerve is usually adherent to the apex of the aneurysm and stretched taut over it. An aneurysm here may be silent until it gives rise to a subarachnoid haemorrhage. The third nerve palsy then indicates the site of the aneurysm which has bled.

It may give rise to a third nerve palsy without producing subarachnoid haemorrhage. The onset is usually of sudden severe pain in the eye and forehead and this is followed by a third nerve palsy immediately or within a few hours or days. Clinical examination usually shows a complete third nerve palsy with no involvement of the fifth nerve and no paralysis of fourth or sixth nerves. These points serve to differentiate the supraclinoid from the infraclinoid carotid aneurysm. It is uncommon for fifth nerve involvement to occur with a supraclinoid aneurysm. If it does, it is rarely more than an impaired corneal reflex.

Painless third nerve paralysis due to a supraclinoid aneurysm is extremely uncommon.

Tumours situated at the level of the posterior clinoid such as meningiomas, chordomas, epidermoids and invasive pituitary adenomas may affect the ocular nerves singly or in combination in their subarachnoid course.

The sudden onset of the third nerve palsy and the severe pain which precedes it are fairly characteristic of an aneurysm at this site. Tumours rarely have such a sudden onset and the development of the clinical picture is less dramatic.

Cavernous Sinus Syndrome

The syndrome of the cavernous sinus is characterized by a greater or lesser degree of involvement of the third, fourth, fifth and sixth cranial nerves on the same side. All three divisions of the trigeminal nerve may be affected. This syndrome can be caused by a variety of conditions, of which an aneurysm of the infraclinoid carotid artery is the commonest if one excludes traumatic carotid cavernous fistula. Tumours involving this region are not common. Neuroma of the fifth nerve, middle fossa meningiomas, laterally extending pituitary adenomas, metastatic tumours, locally invasive tumours from the sphenoid sinus and nasopharynx are the more important ones.

It is differentiated from the syndrome of the superior orbital fissure by the fact that all three divisions of the fifth nerve may be involved and that vision is rarely affected in the cavernous sinus syndrome at an early stage.

In the differential diagnosis a few points are noteworthy. Intracavernous aneurysms occur predominantly in women over middle-age and the onset of symptoms is abrupt with unilateral pain, ocular palsy and diplopia and loss of function in one or more divisions of the fifth nerve depending on the anatomical site of the aneurysms.[14] When there is a third nerve paralysis due to a lesion within the cavernous sinus, the pupillary dilation is often of a mild degree due to the associated paralysis of the ocular sympathetic. Tumours usually have a less sudden and more gradual evolution except the sudden expansion of a pituitary adenoma in an episode of pituitary apoplexy. Pain in the side of the head and face with fifth nerve involvement and perhaps a seventh nerve palsy and later a sixth nerve palsy should, of course, lead one to suspect a suppurative otitis media with extension of infection to the petrous apex (Gradenigo's syndrome). A diabetic patient may present with double vision and extremely severe headache and pain in the distribution of the first and second divisions of the fifth nerve. An isolated ocular nerve palsy may be found but no arteriographic abnormality may be detected. The prognosis for full recovery is good.[15]

DIPLOPIA DUE TO DISTURBANCES OF THE VISUAL FIELDS: NON-PARETIC DIPLOPIA

Beckman and Kubie[16] described diplopia occurring in their series of patients with tumours of the pituitary and pituitary stalk unassociated with ocular palsy. The diplopia seemed to occur when normal extra-ocular muscles attempted to compensate for defects in the visual fields and depressed macular acuity by deliberately bringing into simultaneous use, portions of the retinae which had hitherto been unassociated. They stated that 3 conditions were necessary for its occurrence: (*i*) a heteronomous field defect, usually bitemporal; (*ii*) reduction of acuity at the maculae to less than that of the adjacent normal halves of the retina; and (*iii*) the power to project the images accurately in relation to the point at the retina on which the image falls. Under these conditions the patient attempts to improve his binocular vision by using his perimacular fields. Others, notably Chamlin *et al.*[17] have come to similar conclusions and found 4 cases in 156 patients with pituitary tumours and craniopharyngiomas. The validity of this theory is open to question according to Lyle and Clover[18] because the central visual acuity of each eye in their patients was practically normal. Clover[19] reports on 100 patients with pituitary tumours of whom 20 cases showed diplopia. This diplopia was thought to be non-paretic in 13. Two of the paretic cases occurred in attacks of pituitary apoplexy. Lyle describes this diplopia as being horizontal in nature and due to a slight divergence in the visual axes consequent to a loss of the area of field overlap common to the two eyes.

This diagnosis should be made with a great deal of caution, since the amplitude of convergence normally depends on numerous factors, including the effort of the patient, the presence of binocular vision, the visual acuity in each eye, and the refractive status of the eye, and the attention value of the object used to elicit convergence. An abnormality in any of these may produce an apparent decrease or absence of convergence. A failure of convergence on functional grounds should not be confused with a true paralysis of convergence caused by organic disease. Suggestive of a true paralysis is a history of a sudden onset of diplopia in the presence of a known intracranial lesion and a constancy of the findings on successive trials.

With simple paralysis of convergence, either eye may be rotated inward to the full extent with conjugate lateral movements showing that there is no true paralysis of the internal recti. While disturbance of convergence occurs with occipital lesions, it is with lesions from the region of the superior colliculi that paralysis of convergence occurs characteristically. Aside from discrete lesions in this region, such as tumours or aneurysms, diffuse lesions such as occur with encephalitis, multiple sclerosis, Parkinson's disease and vascular lesions also cause a paralysis of convergence.

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