

Some neuro-ophthalmological observations¹

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This paper records some neuro-ophthalmological observations made on the Stroke Service of the Massachusetts General Hospital. For the most part they are not mentioned in textbooks of neurology. Although they are but minor additions to the clinician's fund of knowledge, their recognition may aid in accurate diagnosis. Some are possibly of interest in their own right as illustrations of the integrated action of the central nervous system. Since the abnormalities are generally unrelated, they will be discussed individually.

DILUTED PUPIL IN OCCLUSION OF THE INTERNAL CAROTID ARTERY

Several times, in occlusion of the internal carotid artery we have found the ipsilateral pupil enlarged and fixed or poorly reactive to light.

A man, aged 69, with bilateral internal carotid artery occlusion had two spells of unconsciousness six months and five months before admission. The first lasted five minutes, the second, 20 seconds. There were no residual ill effects. A short time later it was found that the right pupil was larger than the left (right 7 mm., left 3 mm.) and that the right pupil reacted sluggishly to light directly and consensually. The extraocular movements were full. There was no ptosis. Sweating on the forehead was not different on the two sides.

The central retinal artery diastolic pressure was approximately 20 units in each eye. Arteriography showed bilateral internal carotid artery occlusion with extensive filling of the carotid territory intracranially via the ophthalmic arteries. A vertebral angiogram showed filling of the carotid system from the basilar artery.

Extensive neovascularization of the retina was present bilaterally. Gonioscopy demonstrated a confluent dense mesh of fine vessels involving the iris of the right eye over a 360° extent and a slight involvement of the left iris. A diagnosis of rubeosis iridis was made. Glaucoma was not present and there was no cataract formation. Atrophy of the iris was not described.

A glucose tolerance curve showed a fasting blood sugar

of 76, half hour 150, one hour 180, two hours 147 mg.%. The blood cholesterol was 383 mg.%.

A diabetic man, age 49, had noted three months before admission that vision in the right eye was 'broken into a jig-saw puzzle'. Everything looked gray and he saw only fragments of objects here and there; *e.g.*, in looking at a face he saw part of an eye and a little of the mouth. One month before admission vision had become worse and he could see only from the corner of the right eye. Four months before admission at a time when vision was intact, it had been noted that the right pupil was dilated and unreactive.

Pulsation was absent on the right side in the upper extremity and in the subclavian, common carotid, external carotid, and temporal arteries, *i.e.*, distal to the innominate artery. The right pupil was 6 mm., slightly irregular and fixed. The left pupil was 3 mm. and reactive. The retina of the right eye presented a most unusual picture, a slow movement of blood being visible in almost all veins. The arteries were attenuated and the optic disc was pale. The central retinal artery pressure on the right was virtually zero, on the left, 65, diastolic. The extraocular movements were full and there was no ptosis. Vision was limited to a small sector of the inferior temporal visual field. There was early neovascularization of the iris, especially at the angle. A diagnosis of early rubeosis iridis was made. Dr. J. B. Whitney, who performed the ophthalmological examination, attributed the mydriasis to atrophy of the iris.

In another case, a man aged 50 with a mild right hemiparesis and dysarthria, bilateral occlusion of the internal carotid arteries was found. The left pupil was 6 mm. and reacted poorly, the right was 4 mm. and reacted well.

In these cases the lack of associated neurological symptoms suggests that the mydriasis is the result of a local disorder of the iris, probably ischaemic atrophy rather than a primary nervous derangement. The abnormality has been described in the past in Takayasu's disease in which the carotid circulation is obliterated by an arteritic process (Walsh, 1957).

OCULAR SIGNS IN LARGE HYPERTENSIVE INTRACEREBRAL HAEMORRHAGE

Hypertensive haemorrhage has four main sites of predilection: putamen 60%, thalamus 10%, pons

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10%, and cerebellum 10%. Ocular signs are all-important in identifying the site of the bleeding and helping to determine the feasibility of surgery. These signs may be pithily summed up as follows:

In putaminal haemorrhage there is a hemiparesis and the eyes are deviated conjugately to the opposite side. In thalamic haemorrhage there may be a hemiparesis and the eyes are deviated downwards as if peering at the nose. Vertical eye movements are impaired and the pupils are about 2 mm. in size and unreactive. In pontine haemorrhage the eyes are in the central position, the pupils are pin point and reactive, reflex eye movements are absent, and ocular bobbing may be present. In cerebellar haemorrhage there is a forced conjugate lateral deviation of the eyes, a paralysis of conjugate lateral gaze, or a sixth nerve palsy without paralysis of the limbs. The pupils are of average size and reactive.

PRESERVATION OF THE PUPILLARY REACTION IN PONTINE MIOSIS

One of the classical diagnostic signs of intrapontine haemorrhage or massive pontine infarction has been small (1 mm.) fixed pupils. The cause of the miosis is customarily held to be interruption of the descending sympathetic pupillo-dilator fibres, but the mechanism of failure of the pupils to react to light has remained unexplained.

We have carefully tested the reaction of pinpoint pupils in four patients with pontine haemorrhage and two with massive pontine infarction. Using a bright light and a hand lens, it was possible in each case to observe a definite reaction. Extreme miosis has probably caused the reaction to be overlooked in the past. This finding relieves the neurologist of trying to explain how a pontine lesion paralyzes the pupillary reaction.

While on this subject it may be added that pontine miosis has not been encountered as a unilateral phenomenon and probably occurs only with bilateral interruption of the descending sympathetic system.

CONJUGATE DEVIATION OF THE EYES TO THE 'WRONG' SIDE IN SUPRATENTORIAL HAEMORRHAGE

It is axiomatic that a large supratentorial haemorrhage causes a contralateral hemiplegia and deviation of the eyes away from the paralyzed side, towards the side of the lesion. In occasional cases, however, most signs point to a supratentorial haemorrhage when the eyes are tonically deviated conjugately to the side of the hemiplegia mimicking the formula of a pontine lesion.

In three successive such cases the haemorrhage on pathological examination has proved to be located

in the medial part of the thalamus on one side with a relatively massive collection of blood within the third ventricle.

In one case an 89-year-old hypertensive woman was admitted with a right hemiparesis and forced conjugate deviation of the eyes to the right and downwards. The pupils were 4 mm. and unreactive. Necropsy showed a 2 cm. haemorrhage lying in the left mid-thalamus extending into and distending the third ventricle. There were scattered ancient softenings in the right occipital lobe due to an old occlusion of the right posterior cerebral artery. There was also a cavity 2 by 5 mm. in the anterior limb of the internal capsule on the right side and a 1 mm. 'lacune' in the mid-pons.

In another case a hypertensive man aged 49 was brought to the emergency ward having collapsed 30 minutes before. He was restless, moving the right side aimlessly. The left arm was tonically flexed, the left leg tonically extended. The pupils were 2 mm. in diameter and unreactive to light. The head and eyes turned spasmodically to the left. The eyes did not move to the right on head rotation. The patient died 12 hours later. Pathological examination showed a large haemorrhage into the thalamus on the right side with blood ballooning the third ventricle, and, in addition, blood had dissected downwards into the midbrain and pons. The absence of such a dissection in the first case indicates that this downward tracking is not a necessary part of the process. The third patient had a left hemiplegia, non-reactive pupils and roving movements of the eyes from the central position fully to the left but not to the right. Pathological examination was much the same as in the case just described.

PUPILLARY SIZE IN BRAIN STEM LESIONS

Studies of the effect of brain stem lesions at various levels on pupillary size have led to the following conclusions: (1) lesions located in the subthalamus produce a moderate miosis (2-3 mm. approximately); (2) subtotal damage to the midbrain as in tentorial herniation produces huge pupils (7-10 mm.) presumably due to paralysis of the parasympathetic outflow; (3) with more extensive midbrain damage the pupils are less widely dilated (4-6 mm.) presumably because the sympathetic fibres passing downwards are also interrupted; (4) a large lesion of the tegmentum of the pons produces an intense pinpoint miosis (1 mm.) which is usually attributed to sympathetic paralysis and sparing of the parasympathetics; (5) combined pontine-midbrain damage results in a pupil of the same size as in (3), *i.e.*, intermediate in size between (2) and (4), parasympathetic interruption being added to pontine sympathetic paralysis. The failure of the pupil to be of pinpoint size with total cervical cord lesions and in Horner's syndrome has not yet been explained.

REFLEX EYE MOVEMENTS IN COMA

It is now routine neurological practice in cases of coma to determine the reflex eye movements on head rotation and on ice water irrigation of the ear canals. When necessary, these two manoeuvres should also be combined.

During the last 12 years we have assessed these reactions in over 100 cases studied clinically and pathologically and have found that full reflex horizontal movements indicate that the lesion or lesions causing the coma lie in the cerebral hemispheres and not in the infratentorial brain stem. This rule appears to be infallible. Incomplete horizontal movements signify a brain stem lesion which may or may not be the sole cause of the coma. If eye movements become impaired early in the illness, a site in the brain stem is favoured as the cause of the coma; if impairment appears late a supratentorial lesion with a secondary effect on the brain stem (tentorial herniation) is more likely.

On this basis, it is useful clinically to divide all coma cases into two groups, those with full reflex eye movements and those with restricted movements. It must be remembered that intoxication with barbiturates or other hypnotics very readily paralyzes reflex eye movements, the only metabolic disorder to do so.

REFLEX EYE MOVEMENTS ON HEAD TILTING

In assessing cases of stupor and coma, we have found that in addition to head rotation the response of the eyes to head tilting (flexing the neck) is also useful, giving information about the integrity of the brain stem from the vestibular system to the conjugate vertical gaze centre in the region of the posterior commissure and nucleus of Darkschewitsch and of the peripheral outflow of the third and fourth nerves as well. Thus the manoeuvre probes or tests the brain stem to a slightly higher level than does head rotation.

The test is analogous to that of head rotation: on tilting the head forward the eyeballs are elevated and on extending the neck the eyeballs are depressed. The excursion of the eyes is less than the corresponding lateral movement on head rotation. Upward excursion is especially limited. For better observation during head tilting, the patient's eyelids may be taped open with adhesive rather than being held with the fingers.

In hemispherical lesions when full reflex horizontal movements of the eyes are difficult to elicit, full symmetrical vertical excursions on head tilting may be used as evidence that the upper brain stem is relatively intact.

EYE CLOSURE AND ABSENCE OF BLINKING IN STUPOR

In the neurological examination of the stuporous patient, the state of the eyelids is usually not recorded or observed. For example, closure of the lids is so obviously a part of the picture that it is apt to go unnoted. Yet it strongly suggests a parallel with natural sleep and thereby provides a framework for analyzing and classifying comatose states. In both stupor and sleep, closure of the lids reflects a diminished or unsustained responsiveness to external stimuli and probably a dearth of internal ideation. The degree of narrowing of the palpebral fissures in the undisturbed as well as in the stimulated subject parallels the depth of stupor and the severity of drowsiness. The length of time the lids remain open after arousal is also a measure of the depth of the stupor. The onset of sleep is associated with an active tonic contraction of the orbicularis oculi, and in stupor if the lids are only partly closed the superimposition of full sleep results in a more complete closing of the lids; or the slightly open eye on the side of a hemiplegia closes further. The degree of tone in the closed lid is variable and not infrequently trying to lift the lid evokes a brisk blepharospasm. Not always are these changes obvious on casual observation.

When a person is falling asleep, one of the first events is cessation of blinking. Thus in the unresponsive patient if blinking persists it may be concluded regardless of the electroencephalographic pattern that the brain stem reticular system, at least in part, is 'awake'. Observations of blinking may therefore be of value to the neurophysiologist interested in signs of alertness in the 'nerve net'. Blinking varies in rate, speed of contraction and amplitude, the feeblest action being a slight medial movement of the lower eyelid. Blinking continues in the presence of ptosis or Bell's palsy and may occur when the eyelids are closed as in drowsiness.

Although closing the lids and cessation of blinking often occur together as indicators of a sleep-like state, it is not uncommon for the lids to be closed, yet blinking continue, suggesting that lid closure is a better index of reduced reticular activity. After an epileptic seizure the lids may be closed and the patient almost totally unresponsive yet blinking is very active. At other times the lids may be closed and unblinking, yet the patient, although appearing asleep, is almost awake judging from his pulling up the covers or scratching his nose or opening his eyes when he is gently tapped or his name is quietly called. This state, not uncommon in large hemispherical lesions, is akin to 'going to sleep' or 'being on the verge of sleep'. Other apparent dissociations between

the depth of stupor and the state of the lids may be seen in barbiturate intoxication where the lids may be closed and unblinking, when a pinch on one arm brings an appropriate response from the other arm, indicating that some parts of the brain can carry on rather 'high level' activity when other parts appear deeply asleep. In the apnoeic phase of Cheyne-Stokes respiration, the eyes may close and blinking stop or become greatly reduced.

Usually when a stuporous patient is 'roused', the eyelids open slowly and only partially in accord with the degree of drowsiness. There is a condition, however, in which a slight stimulus produces a sudden and full, wide-awake opening of the eyes as if staring or startled. There may be little or no other somatic reaction. Unless stimulated, the patient keeps the eyelids closed. There is here a discrepancy between the fully awake lid response and the appearance of sleep as judged by the lack of other responses and the persistent closure of the lids before stimulation. In one of our cases it accompanied failure of central reflex respiration at a time when the limbs were still mobile and the ocular movements were preserved to some extent.

THE 'ONE-AND-A-HALF SYNDROME'—COMBINED
LATERAL GAZE PALSY AND UNILATERAL
INTERNUCLEAR OPHTHALMOPLÉGIA

In lesions of the brain stem, vascular particularly, demyelinating or neoplastic more rarely, there occasionally occurs a paralysis of eye movements in which one eye lies centrally and fails completely to move horizontally while the other eye lies in an abducted position and cannot be adducted past the midline. Vertical eye movements are relatively preserved. Although the explanation is straightforward, the condition is included here because it is almost invariably misinterpreted.

A woman, aged 79, awakened with a right hemiparesis. She was alert and gave her own history. Examination showed moderate weakness of the right limbs and dysarthria. The left eye did not move to the right past the midline. The right eye was in abduction, *i.e.*, deviated to the side of the hemiplegia. It could be moved toward the left voluntarily. Two days later the patient's condition had deteriorated. The left eye was now fixed in the central position and did not move or develop nystagmus with ice water irrigation of either ear canal. The right eye moved only from the central to the lateral position. On head rotation and ice water irrigation of the right ear canal, the right eye abducted but no other ocular motion occurred. The patient could elevate the eyes satisfactorily. The pupils were 2 mm., equal and reactive. There was no ptosis. At necropsy there was basilar artery thrombosis with softening of the lower three-quarters of the left side of the pons.

A 78-year-old woman, after complaining of headache, dizziness, vomiting, and unsteadiness on her feet for one week, became severely dysarthric, somnolent, and confused. On examination she was cooperative. Speech was slurred beyond comprehension. There was a mild right facial weakness. The tongue moved weakly. The limbs moved normally. The pupils were equal and reactive. There was no ptosis.

The right eye was in the primary position and could not be moved voluntarily. It abducted on head rotation and on ice water irrigation of the right ear. The left eye was deviated outwards at rest, and voluntarily could not be moved past the midline to the right.

We have attributed the syndrome to a lesion in or near the pontine conjugate lateral gaze centre on one side (causing the conjugate gaze palsy) plus an internuclear ophthalmoplegia due to the interruption of the ipsilateral medial longitudinal fasciculus after it has crossed the midline from its site of origin in the contralateral vestibular nucleus (causing failure of adduction of the ipsilateral eye). The ocular response on head rotation varies, depending on the integrity of the vestibular-conjugate gaze centre connexions.

This syndrome which we have colloquially termed the 'one and a half syndrome' because it consists of a conjugate lateral gaze palsy in one direction, plus one half of a gaze palsy in the other, provides evidence that the medial longitudinal fasciculus fibres running to the opposite ocular adductors cross to the other side caudally in the pons near their origin in the vestibular nucleus rather than running cephalad for some distance before crossing.

DILATED, FIXED, OR HYPOREACTIVE PUPILS IN
TUBERCULOUS MENINGITIS

Recently our attention has been drawn to the state of the pupils in tuberculous meningitis.

A 17-year-old boy was admitted with a 10-day history of headache and fever. The spinal fluid was under a pressure of 410 mm. and contained 375 lymphocytes and 8 polymorphs per c.mm., 153 mg. % of protein, 25 mg. % of sugar, and on smear 2 tubercle bacilli. The patient was drowsy. The pupils measured 7 mm., the right reacting well, the left only slightly. The extraocular movements were full. Six hours later the pupils were 8 mm. and fixed to light. Yet the eye movements were unimpaired. The patient was restless but cooperated in the examination. A left occipital burr hole was placed and during the anaesthesia the pupils narrowed to 3 mm. Next day the patient was more alert but the pupils were still widely dilated and slow to react, particularly the left. A remarkable hippus was observed. Ophthalmological examination showed a fourth nerve paresis. There was no ptosis or other impairment of the muscles supplied by the third nerve. No clear explanation for the ocular disturbance was forthcoming and when the patient made an excellent

recovery on antibiotic therapy, the finding was not pursued further.

More recently we were consulted concerning a woman aged 45 with fever, headache, confusion, 190 lymphocytes in the cerebrospinal fluid, and raised intracranial pressure. In the third week of the illness the pupils were noted to have become wider (8 mm.) and only feebly reactive to light. The reaction on convergence was less impaired. The pupils were equal. Visual acuity was excellent. The extraocular movements were full and there was no ptosis. The pupillary response was reminiscent of the case described above and a diagnosis of tuberculous meningitis was suggested. When therapy was changed to streptomycin, isoniazide, and aminosalicylate, the previously steady downhill course was reversed. Culture of the spinal fluid later showed *M. tuberculosis*. As the patient improved, the right pupil returned to a diameter of 4 mm. and was reactive at the time when the left pupil remained large and almost fixed. In a third case of tuberculous meningitis pathologically proved, the pupils were barely reactive at an early stage of the illness. A fourth case showed a remarkable hippus.

While there are no modern descriptions of the pupillary changes in tuberculous meningitis, in an older series (Uthoff, 1915) abnormalities were reported in 40% of cases. The pupils were unreactive in 15%, the reaction was impaired in 15%, and the pupils were unequal in another 10%. Although many of the patients were stuporous and light perception could not be adequately tested, the author felt that lesions of the optic nerves were not the cause of the failure of the light reaction. In addition, optic neuritis was reported in 25%, papilloedema in 5%, third-nerve disturbance in 18% (partial ocular palsy eight, ptosis 10), sixth nerve paresis in 12%, nystagmus in 10%, conjugate deviation in 8%, and other movement disorders in 1%. Hippus is apparently an uncommon event in tuberculous meningitis and is more frequently recorded in meningococcal meningitis.

Recognition of these ocular disturbances not only assists in the management of the illness but nowadays when tuberculous meningitis is frequently a difficult diagnostic problem, it may even suggest the true nature of the meningitis. The mechanism by which pupillary dilatation occurs has never been the subject of inquiry. Exudate over the base of the brain is probably responsible but whether the change in reaction is anatomically or biochemically determined can only be speculated upon at present. The abnormality seen in tuberculous meningitis is no doubt related to several other sometimes obscure pupillary changes, for example, in syphilitic meningitis, diabetic oculomotor neuropathy, and tentorial herniation.

It is appropriate here to refer briefly to the ocular abnormalities seen in meningococcal meningitis. In one of the most thorough studies, pupillary

disturbances were recorded in 63 of 69 cases (Ballantyne, 1907).

In a recent case on our Neurology Service a young woman aged 21 with fever, headache, and vomiting for four days, showed 1,200 leucocytes/c.mm. in the cerebrospinal fluid (81% polymorphs, 19% lymphocytes). The right pupil was 5 mm. and fixed, the left pupil 3 mm. and reactive. A striking finding was an intense injection of the bulbar conjunctiva of the right eye. Culture of the spinal fluid disclosed meningococci.

Ballantyne reported that conjunctival hyperaemia was frequent, a finding not confirmed by Uthoff.

CENTRAL PHOTOPHOBIA (CENTRAL GLARE, CENTRAL DAZZLE)

This not uncommon complaint takes the form of an excessive visual brightness, glare, or dazzle in patients with a homonymous hemianopia. Although often mild, it may be the patient's main distress after an otherwise satisfactory recovery.

A patient with a massive demyelinating lesion of the right hemisphere causing hemiplegia, sensory loss, homonymous hemianopia, and apractagnosia of the minor hemisphere recovered well except for the complaint, 'It's my eyes. It's the amount of light coming in, it's either too much or too bright. I don't see clearly across the room. I don't pick up details. It's not too light and it's not too dim. The surround is affected. When it's darker I see all right. Twilight is better. Oncoming headlights are bad. On a bright day I am dazzled as if I can't focus. It creates a glare and yet there is a dimness.' Examination showed a dense left upper quadrant-anopia and in addition a mild left hemiparesis.

Four patients with a partial left homonymous hemianopia, presumably the result of thrombosis of the posterior cerebral artery, had a similar complaint. One said that it bothered her if there was too much light around. Yet paradoxically she said that her vision 'just doesn't look like daylight, it's like twilight'. The patient kept the blinds in her home drawn. Here again there was a combination of dazzle and dimness. The second patient complained of the 'excessive brightness of things' which gave him a feeling of unreality and prevented him from focusing sharply.

The third stated that for three years after his stroke there was an oversensitiveness to bright light. 'The sunlight is too bright. There is so much glare from the ceiling it partially blinds me.' Looking out of a window was unpleasant because of the brightness. He blamed the phenomenon for his poor performance on the non-verbal part of his cortical function tests. The fourth patient noted a 'tremendous glare' when light came from the left, but not when it came from the right.

In the following case a visual field defect could not be demonstrated.

A woman, aged 69, after having had recurrent attacks of visual blurring for two weeks, suddenly developed clumsiness of the hands, dysarthria, and confusion. Immediate examination showed impaired memory and nystagmus. The signs cleared in a few days but the patient then noted on going outdoors 'an unpleasant blurring of vision' due to the brightness. 'I don't seem to see well. The strain is terrific. I'm living as in a nightmare.' The sun shining on water was particularly unpleasant. On cloudy days she could see better, and at twilight or dusk, the moon, stars, trees, silhouettes, and sharp corners of buildings became very plain. Neurological examination was normal except that on looking through a pinhole one side seemed to be obscured, 'as if there was a thread there'. The visual fields were full to routine testing. The pupils were equal and reacted normally. The diagnosis was basilar artery ischaemia with damage to the visual system in the occipital region.

Central photophobia appears during the healing phase of the pathological process. It is probably related to a lesion of the calcarine region, for it has not been associated with homonymous visual field defects resulting from infarction in the middle cerebral territory. It is probably the visual counterpart of dysaesthesias and hyperpathia arising in the damaged somatosensory system, and of the related phenomena in the olfactory and other special sensory systems.

THE LOCALIZING VALUE OF SCINTILLATIONS

In cerebrovascular disease, infarction as well as haemorrhage, the patient may experience visual scintillations akin to those of migraine. Sometimes these are fleeting, sometimes persistent. It is our impression that scintillations always indicate involvement of the occipital lobe, probably the calcarine visual cortex, and never arise from involvement of the optic radiation as it passes through the temporal lobe. They point to posterior cerebral artery disease and have not been observed in visual field defects of middle cerebral origin. Since at times the posterior cerebral artery is a branch of the internal carotid artery, scintillations may be associated with internal artery occlusion, but this is a rare event. In most instances the scintillations have occurred along the edges of a visual field defect.

A woman, aged 65, four days before admission while standing at work, lost consciousness and was found lying on the floor. The spell lasted about five minutes. After this episode she constantly saw zigzag white lights in the left field of vision. Many years before, the patient had had migraine with an aura of similar zigzag lights. Neurological examination was normal. The blood pressure was 170/130 mm. Hg. During the next month the patient developed a left homonymous hemianopia, dysarthria, nystagmus, cerebellar ataxia, and finally a quadriplegia. Necropsy showed thrombosis of the left

vertebral and basilar arteries with infarction of the pons and both occipital lobes.

A woman aged 58 was recuperating after myocardial infarction when suddenly the left field of vision became filled with dancing lights, yellow above, white below. At the onset bilateral frontal headache and nausea were present. The initial dazzle subsided to a zigzag line of lights which outlined the medial edge of a dense left lower homonymous quadrantanopia. The lights persisted when the eyes were closed but they were less bright. The diagnosis was embolism to the right posterior cerebral artery.

A patient, aged 56, suddenly developed a headache in the right temple associated with flickering lights in the extreme left visual field. Examination showed a left upper homonymous field defect. The scintillations persisted for five weeks. Surgical exploration revealed a haemorrhage in the posterior extremity of the right occipital lobe.

THE LOCALIZING VALUE OF VERTICAL NYSTAGMUS

It is still widely held and taught that vertical nystagmus may result from upper (as well as lower) brain stem lesions. It is our experience that this is wrong. In studying the recovery of oculomotor function in cases of thalamic-subthalamic haemorrhage in which paralysis or paresis of conjugate vertical gaze, both upwards and downwards, was present, in no case was a stage of vertical nystagmus observed. Vertical nystagmus therefore does not reflect weakness of vertical gaze. Cases with upper brain stem lesions of other types were especially examined for vertical nystagmus and they did not show it. On the other hand, cases with vertical nystagmus have all had evidence of lower pontine or upper medullary mischief.

It is our conclusion, therefore, that vertical nystagmus reflects disease localized in the region of the ponto-medullary junction and not the upper brain stem.

TRANSIENT DEVIATION OF ONE OR BOTH EYES

Transient conjugate deviation of the eyes associated with focal epilepsy and oculogyric crises is a commonplace event. Episodic deviation of one or both eyes may occur under other circumstances, however, particularly in transient ischaemic attacks associated with cerebrovascular disease in the vertebral-basilar territory.

(a) A man aged 59 had innumerable 30-second attacks of a giddy feeling in the head, at times associated with vertical diplopia. Twice the right side of the tongue was numb. In one of the spells while he was seated it was noted that the right eye elevated to the extent of almost disappearing behind the upper lid and deviated slightly to the

right. This lasted 20 seconds. Severe stenosis in the vertebral system bilaterally was shown on arteriography.

(b) A man aged 54, for three months had recurrent spells of diplopia, sweating, and dizziness lasting five to 10 minutes. In one spell his wife noted that one eye was deviated far laterally. The patient went on to develop a stroke marked by dysarthria, weakness of the tongue, left arm and left leg, and persistent nystagmus.

(c) A woman, aged 67, had brief attacks of confusion, dizziness, and diplopia. The patient's daughter observed that in one spell upon sitting up the patient's eyes darted far laterally to the right side and could not be brought back to the midline. Upon lying down the eyes returned to the central position. In the next two weeks the patient developed an extensive basilar artery infarction including a right hemiplegia.

(d) A hypertensive man, aged 58, for four weeks had several spells each day lasting five to 30 minutes, characterized by sweating, dizziness, diplopia, staggering, loss of equilibrium, occipital headache, pain in the right nostril, icy coldness of the left side of the nose, and the walls appeared to break into sections. His wife described elevation of the left eye during the spell.

(e) A hypertensive man, aged 69, had six attacks lasting a few minutes of lightheadedness, blurred vision, and deviation of the eyes to one side with inability to bring them back to the midline position. Later spells were associated with diplopia, dysarthria, and weakness of the left arm. In one attack which was witnessed, the eyes deviated to the left and could not be brought to the right past the midline.

(f) A diabetic man, aged 48, in a period of one month had several attacks in which the right eye elevated to 11 o'clock and the left eye to 1 o'clock following which he immediately fell 'asleep' sitting up. After five minutes he 'awakened' and continued his scheduled activity. Hypoglycaemia was suspected, but finally in one of the spells dysarthria appeared and during the next week the patient developed a severe brain stem infarction from which he died. Necropsy showed basilar artery thrombosis.

The above cases are examples of cerebrovascular disease with lateral or upward deviation of one or both eyes. In the next three cases, the patients felt that the eyes were askew.

(g) A man, aged 63, had spells three times a week of spots before the eyes, imbalance, lightheadedness, and a sensation that the eyes were crossed. 'The left eye goes over to meet the right one and seems to move around interfering with vision in the right eye.' The patient later developed a brain stem infarct.

(h) A man, aged 58, had brief attacks of numbness of the left face and hand, a hot feeling rising from the waist, a puff of smoke before the eyes, and a turning in of the right eye as if it were going across the bridge of the nose.

(i) Another patient, aged 75, had a single two-hour spell of diplopia in which she felt that the left eye was being pulled out of shape toward the nose.

It is a good rule that dizziness associated with a

feeling that the eyes are 'crossing' or being pulled medially or laterally indicates major vertebral-basilar artery ischaemia and not a more benign labyrinthine disorder such as Ménière's syndrome.

(j) In this case, there were bouts of deviation of one eye associated with a glioma. A man, aged 28, for 18 months experienced episodes in which the left eye deviated laterally for a few seconds without other symptoms. Later a persistent third nerve palsy appeared. Operation disclosed a glioma involving the region of the third nerve in the cavernous sinus region. Periodic lateral deviation of the eye had clearly preceded other symptoms.

(k) In this case no cause for the disorder was found. The patient, aged 41, for two months had spells of vertical diplopia lasting a few seconds up to eight hours. There was no associated dizziness, numbness, or headache. Examination during a 30-second spell showed 50% limitation of downward gaze in the left eye. All other ocular functions were normal.

UNWITTING CLOSURE OF ONE EYE

In this disorder the patient closes one eye without being aware that he is doing so. Not only may he deny that he is closing the lid, he may even claim that it is not closed. Yet on command he can readily hold both eyes open. The patient may erroneously be thought to have ptosis.

The condition has been observed several times, usually in brain stem disease, and it is our interpretation that the closure is an 'automatic' reaction of the central nervous system to the presence of diplopia. Although diplopia may actually be a complaint, or the eyes may move disconjugately, there are cases in which the patient disclaims double vision.

A boy, 5 years of age, with increased intracranial pressure, papilloedema, and ataxia of the left leg after a closed head injury, developed a left sixth nerve palsy. While being examined he kept the left eye closed, the right eye remaining open. This was interpreted as ptosis. However, when the right eye was covered the left eye opened widely. When questioned in simple language about diplopia, the boy denied it. From our observations it appeared that he automatically held the left eye closed. The combination of a sixth nerve palsy and isolated ptosis had posed a difficult problem.

A patient, aged 64, had an intracerebral haemorrhage associated with a right hemiparesis, dysphasia, impaired vertical eye movements, and paresis of abduction of the left eye. When addressed, he opened the right eye but kept the left eye closed, unaware that he was doing so. On command, he kept both eyes open.

A woman, aged 63, with basilar artery thrombosis, had a 'one and a half syndrome' and in addition always kept the left eye closed. She said she was unaware of actively or voluntarily closing it. Yet when the right eye was covered the left eye promptly opened.

A woman, aged 36, with a ponto-medullary tumour complained of dizziness, vomiting, unsteadiness of walking, and 'imbalance in the eyes'. Examination showed horizontal and vertical nystagmus and a cerebellar gait. In addition she persistently kept the left eye shut but was unaware of this. 'My family says, "Why do you squint out of one eye?" But I don't notice it.'

A patient, aged 45, with a left hemiplegia and anosognosia, kept the right eye squinted shut without realizing it. On looking towards the left side she closed both eyes. When asked to open her eyes she said, 'Aren't they open?'

It would be of no little interest if the nervous system should, without being aware of its action, obliterate the confusing information arising from a disconjugate position of the eyes. Almost always it was the left eye that was affected.

BRAIN STEM PTOSIS

A rare event in basilar artery infarction or ischaemia is the relatively isolated finding of bilateral ptosis. Eye movements and the pupils may be little affected. The patient remains alert. The lid droop is relatively complete and much greater than that of a bilateral peripheral Horner's syndrome. The mechanism of its production is unknown although one might speculate that it is the result of bilateral central sympathetic paralysis. However, the pupils have not been pinpoint in our cases.

A man of 47 suddenly developed headache, dizziness, vomiting, diplopia, and both lids drooped until 'the eyes were like slits'. Improvement occurred in six days when examination showed horizontal nystagmus on gaze to the left, vertical nystagmus on gaze downwards. The extraocular movements, including convergence, were intact. The right pupil was 3 mm., the left pupil 3.5 mm. and both reacted. There was still ptosis bilaterally, marked on the right, moderate on the left. The remainder of the neurological examination was normal. The cerebrospinal fluid contained 116 mg. % of protein. A diagnosis of brain stem ischaemia due to basilar artery thrombosis was made. In another case a 73-year-old hypertensive woman was admitted because of diplopia and dizziness for 12 hours. Examination showed dysarthria, left hemiparesis, right facial weakness, and right sixth nerve palsy. Bilateral ptosis was present, the palpebral fissures being narrowed to about 5 mm. The pupils were equal and reactive. Third nerve function was intact. In a third case the patient was subject to transient spells of dysarthria and ptosis in which the pupils were covered and vision was obscured. Basilar artery thrombosis occurred after three months.

ASYNCHRONOUS BLINKING

Occasionally while observing a patient it is one's impression, usually from a sideways glance, that

the patient does not blink the two eyelids synchronously *i.e.*, one lid starts to move a fraction of a second before the other. Our observations indicate that this is always a sign of a very mild peripheral or nuclear seventh nerve palsy. In upper motor neurone lesions, Parkinsonism, Horner's syndrome, and ptosis, blinking is synchronous. In more advanced defects the facial weakness will be obvious.

OCULAR AGITATION (RESTLESS EYES)

Roving movements of the eyes are a common finding in many obtunded states—strokes, sleep, anaesthesia, and hepatic coma. Occasionally the movements are so brisk and exaggerated as to attract special attention.

A woman, aged 65, suffered a right-sided stroke with aphasia three years before. Recovery was not perfect although speech was surprisingly good. The fingers remained weak and a brace was worn on the right leg. On the day of the present admission the patient gradually became stuporous. Examination showed an obtunded woman whose left limbs were flaccid and immobile. She moved the right fingers and toes on command and mumbled a few words when addressed. The eyes were closed and blinking. The right limbs were spastic. Both plantar responses were flexor. The blood pressure was 200/120 mm. Hg. The cerebrospinal fluid was grossly bloody.

The head was turned to the right. The pupils were 1.5 mm. and unreactive. Both eyes tended to be deviated to the right but were subject to rapid, abrupt, violent oscillations at 75 to 100 per minute in which the eyes moved from the left lateral position towards the centre and then darted back to the full lateral position. There was a slight downward motion as well. Ice-water irrigation of the ear canals evoked tumultuous motion of the eyes but momentarily brought them fully to the right lateral position.

In this case the ocular agitation or restlessness probably had its basis in bilateral hemispherical lesions, old in the left hemisphere, recent in the right. This is the most frequent cause and is illustrated further in the following two cases.

A man aged 86 was walking across the room when he suddenly fell unconscious. Examination showed atrial fibrillation, deep coma, bilateral decorticate postures, and equal reactive pupils. The extraocular movements were extraordinary in that the eyes constantly roved from one extreme lateral position to the other, every two seconds. The patient lived two days. Pathological examination showed bilateral cerebral infarction. There was an old silent occlusion of the right internal carotid artery and a recent embolic occlusion on the left side which had produced bilateral infarction of the hemispheres.

A woman, aged 72, in the past had had a right hemi-

plegia and aphasia from which she recovered satisfactorily. She then developed a complete left hemiplegia and anosognosia. The eyes were deviated to the right but showed a constant drift toward the left and nystagmoid beating towards the right at 2 to 3 per second. The motion also had a slight downward component.

The origin of the impulse for roving eye movements is at present unknown but probably lies in the upper brain stem. When hemispherical control is in abeyance bilaterally, the 'lower' mechanism appears to operate more freely. The occurrence of spontaneous nystagmus of this degree might be related to the finding that when successive hemiplegias occur, the eyes tend to remain deviated for a longer time after the second hemiplegia. One might have anticipated the opposite, for if one cortical conjugate gaze centre has been put out of action, the tendency to drive the eyes to the opposite side when the second hemiplegia occurs should be diminished.

COMBINED WEAKNESS OF THE EXTRAOCULAR MUSCLES AND ORBICULARIS OCULI IN MYASTHENIA GRAVIS AND PROGRESSIVE OCULAR DYSTROPHY

The selective combined weakness of the extraocular muscles and the corresponding orbicularis oculi with relatively preserved power in the lower facial muscle is a distinctive sign of myasthenia gravis and of progressive ophthalmoplegic dystrophy. It is thus particularly important in examining cases suspected of ocular myasthenia to assess carefully the force of eye closure and to compare it with the strength of the orbicularis oris. Speaking generally, weakness of eye closure is at all times a tell-tale of facial myasthenia.

TRANSIENT MONOCULAR BLINDNESS IN GLAUCOMA

A man, aged 63, reported that for approximately two months he had been having attacks of dim vision in the left eye once or twice weekly. Everything went black for 30 seconds. On the day before he came for consultation, upon arising in the morning, blurred vision had lasted for one half-hour. The patient had been struck in the right eye as a child following which he had no useful vision in that eye although he could see vaguely in the temporal field. Ophthalmological examination had shown the central retinal artery pressure to be 40 systolic, 6 diastolic.

On examination, the right eye, which lay in the medial position, had no useful vision. Visual acuity in the left eye was 20/30. The left pupil was 4 mm. and reacted sluggishly. The fundus was normal. The extraocular movements on the left were full. The central nervous system was intact. The blood pressure was 120/70 mm. Hg. There were no bruits in the neck or over the eyes. The central retinal artery pressure was 70/45 on the left. It could not be determined on the right. The common carotid pulses were judged to be normal.

Spells of transient monocular blindness continued to occur about three times weekly during the next three months. In some spells he could see only the bottom part of things, not the top. Bending over was liable to bring on an attack. One episode lasted an hour during which vision was blurred rather than blacked out entirely. On the day after cyclopentolate hydrochloride (Cyclogyl) was used to dilate the pupil, the patient reported that he had awakened with dim vision in the left eye which persisted until 2 p.m. On examination at 5 p.m. the pupil was still large (5 mm.). Visual acuity was 20/30. The fundus was not remarkable, and the fields were full. The central retinal artery pressure in the left eye was extremely low. The sclera was injected, but there was no pain.

Acute glaucoma was suspected and the patient was referred to Dr. David Donaldson. Tonometry showed a pressure of 35 mm. The angle was narrow everywhere. On Pilocarpine therapy the pressure was maintained at 15 mm. for a week. While he was on this regimen the transient attacks of blindness stopped. Narrow angle glaucoma was diagnosed and a peripheral iridectomy was carried out. There were no further episodes in the following year. Two other similar cases have been studied.

OCULAR BRUIT DUE TO BASILAR-CAROTID ANASTOMOTIC FLOW

A bruit heard over the eyeball originates customarily in the internal carotid artery and is strong evidence that the vessel is patent intracranially. There are occasional exceptions, however, in which a distinct ocular bruit is present despite bilateral occlusion of the internal carotid arteries.

A woman, aged 55, was lying on a sofa when the telephone rang. She jumped up to answer it and fell to the floor because of weak legs. She crawled to the telephone on her hands and knees and found she was unable to speak clearly. All symptoms cleared in 15 minutes and on examination the only abnormalities were mild right facial weakness and slight dysarthria. The blood pressure was 210/110 mm. Hg. The diastolic central retinal artery pressure was 16 on the right, 32 on the left. A moderately loud systolic bruit was easily heard over the right eyeball. The bruit persisted despite compression of the common carotid arteries in the neck separately and together. There were no bruits in the neck. The radial pulses were equal.

Carotid arteriography showed occlusion of both internal carotid arteries. On the left side, intracranial filling occurred via the ophthalmic artery. A right vertebral angiogram through a brachial catheter showed a large vertebral artery, the column of dye narrowing in the basilar artery as a sign of good flow along the left vertebral artery. The right posterior communicating artery was 3 mm. wide and dye passed along this vessel to fill the middle cerebral and anterior cerebral arteries on both sides of the brain. The left posterior communicating artery was small.

In this case the bruit probably represented flow through the large right posterior communicating artery. In some cases there has been an augmentation bruit in the vertebral system, *i.e.*, a bruit resulting from enhanced collateral flow along the vertebral arteries in the neck. Collateral flow via the ophthalmic artery from the external carotid artery, no matter how rich the supply, has never in our experience produced an ocular bruit.

PTOSIS ASSOCIATED WITH SUBDURAL HAEMATOMA

A man aged 75 was admitted because of dysphasia and clumsiness of the right limbs, all of uncertain development. The cerebrospinal fluid was normal and initially a mild stroke was suspected. An unexplained finding was a left ptosis of moderate degree, the upper half of the pupil being covered on forward gaze. A carotid arteriogram showed a massive subdural haematoma on the left side. This was surgically removed by Dr. Robert Ojemann and the patient recovered completely.

On the day after operation the left ptosis had cleared and did not reappear. The palpebral fissures had become equal. The possibility was considered that the wider palpebral fissure on the right was the result of a right facial weakness and that the apparent ptosis on the left was actually the normal lid level. However, the palpebral fissure on the right side did not change postoperatively and this, combined with the dramatic disappearance of the drooping lid favoured ptosis as the interpretation.

The degree of ptosis was consonant with a Horner's syndrome but sweating and pupillary size were not involved. The ptosis is tentatively attributed to a mechanical effect on elements of the third nerve, although there was no other evidence of involvement of the nerve.

DOLL'S EYELIDS

This term designates a remarkable reflex opening of the eyelids when, in the comatose patient, the neck is briskly flexed. A truly doll's eyelids phenomenon! The eye-opening mechanism appears to function fully in the presence of a 'sleeping' brain. The reaction has been observed most often in the stupor of subarachnoid haemorrhage.

CESSEATION OF BLINKING ON HOLDING THE BREATH

During auscultation over the eyeball and also during ophthalmodynamometry, excessive blinking by the patient may pose a difficulty. It has been observed that blinking temporarily ceases when the patient is asked to hold his breath. This simple manoeuvre should eliminate most of the noise from the lid musculature. At the same time the eyeballs stop moving. It might also be mentioned here that in order to promote relaxation of the orbicularis oculi during these tests, having the patient let the mouth hang open loosely, will usually inhibit the tendency to squeeze the eyes shut.

XANTHOPSIA IN DISEASE OF THE GLOBE OR OPTIC NERVE

Yellow vision is a well-known accompaniment of digitalis poisoning. It has also been reported after the ingestion of santonin, trinitrophenol, and amyl nitrate and in association with jaundice and eclampsia (Giesler and Wolff, 1927). It has been described with sulphonamide drugs, after metrazol, and in hysteria (Walsh, 1957). We have recently examined three cases in which yellow vision was a feature.

A woman, age 36, had a subtotal thyroidectomy for hyperthyroidism. Seven months later everything began to appear either yellow or blue. 'Things did not have a natural colour. I was colour blind.' When she read, there appeared to be a flashlight shining in the centre of her vision. In the next seven months malignant bilateral exophthalmos with ophthalmoplegia developed and the patient became blind in the right eye and almost so in the left eye. Supraorbital decompression resulted in a remarkable return of vision.

A man, aged 52, became blind in the right eye, over a period of 12 hours. On the following day everything seen with the left eye appeared yellowish and 24 hours later the left eye became blind. A full restoration of visual acuity took place in the next two months and a diagnosis of bilateral retrobulbar neuritis was made.

A man, aged 72, on sitting up in bed at night before going to the bathroom experienced a momentary streak of orange-yellow lightning in the left eye. It was as if the light in the next room had been turned on and was shining through a slightly opened door. Similar flashes then began to appear on other occasions such as when leaning over, turning quickly, or shaking the head. A series of three to eight streaks occurred each time. The light was always distinctly yellowish. A diagnosis of Moore's lightning streaks was made.

The argument whether or not the yellow vision of digitalis intoxication is a peripheral or central phenomenon remains unsettled. These three cases indicate that yellow vision at least in some instances is a peripheral manifestation.

SUMMARY

A number of neuro-ophthalmological observations covering 25 separate topics have been presented and briefly discussed.

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