hair, heavy eyebrows, grey-blue eyes, a retroussé nose and full mouth." He then said: "Everything is more easy with the eyes open without exception." He was then asked to close his eyes again and describe his own foot. He said: "I can't see a foot: I can't visualize anything at all when my eyes are shut." He was then asked to open his eyes and promptly described a naked foot in detail. He had no aphasia or agnosia. His answers in the 100–7 test were correct. He was able to repeat 7 digits forwards and 4 backwards, and he succeeded in repeating a Babcock sentence accurately at the second attempt.

Routine examination revealed no abnormality in the fundi, visual acuity, visual fields or central nervous system elsewhere.

The patient was seen again six years later and his condition in respect of the symptoms described was unchanged. Six months previously, however, he had been admitted to a mental hospital on account of an attack of depression for which he had been treated with electrical convulsant therapy and modified insulin treatment. When I saw him on the second occasion he was again depressed and I admitted him to hospital with a view to treatment and in the hope of carrying out further investigations, including electro-encephalography. Unfortunately, however, he became worse and had to be certified. He was sent to a mental hospital, where he was subsequently regraded as a voluntary patient, and one day he walked out of the hospital without giving notice and was later found dead on the railway line.

The two patients whom I have described present more features of theoretical interest than it is possible to discuss fully now. Loss of visualization is usually found in association with other forms of higher visual disability; Charcot's patient, for example, suffered from word-blindness—but only when it is encountered in relative isolation can its nature be investigated. The suggestion was made in the case of my first patient that it might be a neurotic symptom, and my second patient suffered also from psychotic depression. But my first patient had in no way a neurotic personality and I do not believe that so clear-cut a syndrome, characterized by symptoms possessing no conscious relationship to each other, and occurring in every case after organic lesions of the brain, is other than organic in origin.

If this be accepted, the main interest of the syndrome is psychophysiological. The loss of visual imagery was more complete in Case I than in Case II, in whom visual images sometimes occurred spontaneously though in an impoverished form, and not under the control of the will. The patient exhibited a striking enhancement of visualization when his eyes were open, as though stimulation of the visual cortex strengthened visual imagery. It is unfortunate that this could not be tested electro-encephalographically. In Case I there was an asymmetry in the response to photic stimulation.

Perhaps the most surprising feature is how little the loss of voluntary visualization impaired functions in which visual imagery might have been expected to play some part. Thus, patient No. 1 had a normal memory span for visual objects and could draw designs and describe pictures from memory. Similarly, though he could not visualize parts of his body, he could chart a point touched upon an outline drawing of the body. Charcot's patient showed the same features. It would seem, therefore, that a patient who has no power of voluntary visualization can, nevertheless, recognize objects and persons, accurately propositionize about them, as Hughlings Jackson might have put it, and also reproduce objects graphically. It follows that visual imagery is not essential to these processes, which therefore must depend upon neurophysiological schemas which do not themselves enter consciousness. And the same is true of dreaming, which in patients who have lost the power of visualization continues without visual images.

What, then, is the value of visual images? The image, whether visual or otherwise, in so far as it is a representation of an object reproducible at will, has the great value of enabling thought to deal with the object in its absence, and the visual image being spatially extended has the special advantage of facilitating imagined action in space. Its function, in fact, is well illustrated by the hampering effect of loss of visualization upon the builder designing a house.

My patients throw no light upon the situation of the lesion responsible for the loss of visualization, but there is some evidence suggesting that it is probably in the parastriate region.

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Basilar Artery Stenosis and Thrombosis

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INTRODUCTION

INTEREST in arteriosclerotic brain disease has been increased, or revived, in recent years for several reasons. These include the common use of arteriography, newer physiological observations of cerebral circulation, and the increased recognition of carotid artery occlusions. Verification by angiography of arteriosclerotic stenosis and thrombosis of the internal carotid artery has been

frequent enough to give fairly clear understanding of the related clinical symptoms. We have come to appreciate the occurrence of quite focal signs in this condition, signs formerly thought indicative of small vessel occlusion on the basis of thrombosis, vessel spasm, or the rupture of small miliary aneurysms. The symptoms of arteriosclerotic basilar artery stenosis, comparable to carotid stenosis, are much less clearly worked out. It is unlikely that arteriographic confirmation will ever be common enough in this area to answer our questions. It would seem reasonable to approach this problem by considering the antecedent cerebral symptoms in cases of basilar thrombosis.

Method and Material

It has been our plan to study basilar stenosis by surveying in detail the symptoms antedating thrombosis of the basilar artery and its main branches. This preliminary report is limited to consideration of confirmed cases of basilar artery thrombosis. 22 such cases are here reported. 16 cases occurred in Sunnybrook Hospital, Toronto, between the years 1946 and 1952; 6 cases are from the Toronto Western Hospital between the periods of May 1951 and May 1953. 13 of the 22 cases presented the fairly typical clinical picture of basilar artery thrombosis. The remaining 9 cases were of special interest regarding their pre-thrombotic symptoms. These symptoms included disturbances of consciousness, dysarthria, mutism, hemiplegic manifestations, vertigo, tinnitus, &c.

Description

General (22 cases).—19 patients were male, 3 were female. The age range was 47–81 years, with an average of 66 years. 14 patients were hypertensive; 6 had normal blood pressure prior to their terminal illness; the remaining two cases showed marked hypotension with no record of previous average blood pressure.

Group I (13 cases).—These showed the more typical clinical picture of basilar artery thrombosis. There was sudden onset of loss of consciousness, either as the presenting symptom or closely following brief initial symptoms such as severe generalized weakness, frontal headache, vertigo, dysarthria, vomiting, or hemiplegia. Coma usually developed rapidly and persisted, and death followed in from one to eight days. Flaccid quadriplegia was the initial finding in 4 of the 13 cases. In 7 cases early examination revealed signs of hemiplegia but this only preceded quadriplegia for periods up to twenty-four hours. One patient had 3 transient episodes of right hemiplegia in the nine days prior to his terminal sudden loss of consciousness and quadriplegia. The remaining patient was admitted in coma but a history was obtained of forty-eight hours' duration. This patient began with hemianæsthesia for twelve hours, hemiplegia in addition for the next twelve hours, and on the day before coma developed he was also incontinent and dysarthric but quite conscious. Accurate history of recurrent premonitory symptoms was not obtained in this group as they were admitted to hospital in coma. Pathological examination in this group revealed thrombus formation in the proximal portion of the basilar artery in 6 cases and in these oculomotor palsies or ophthalmoplegia were noted in all. In 7 cases the distal portion of the artery and the vertebral arteries were involved in the thrombus formation.

Group II (9 cases).—These cases had recurrent premonitory symptoms prior to terminal basilar artery thrombosis that suggested periodic insufficiency of blood flow to the brain-stem. Stenosis of the basilar artery occurred so frequently in the whole series that it suggested that it was a causal factor in the terminal thrombus formation. With basilar stenosis as the underlying cause, the immediate cause could well be temporary lowering of blood pressure occurring in hypertensive, normotensive, or even hypotensive individuals. Brief comment of some premonitory symptoms is recorded.

(*a*) *Disturbances of Consciousness*

A 59-year-old man was perfectly well until ten days prior to death when he developed brief episodes of loss of consciousness, associated with transient paralysis of the left arm and leg. Between these episodes the patient was alert, cheerful, well oriented with no specific complaints. He walked into hospital on the day prior to death but the following morning was discovered to be mute and extremely weak, although aware of his surroundings and responding to commands. Eye movements were normal. Shortly afterwards he suddenly became unconscious; a right facial weakness was evident and quadriplegia was present with spasticity a little more marked on the left. Bilateral lower limb clonus and Babinski responses were also present. He died the same evening, but four hours prior to death he developed status epilepticus that could not be controlled. Post-mortem revealed marked atherosclerotic stenosis with a recent thrombus occluding the whole length of the basilar artery.

Derangements of consciousness and their relationship to brain-stem lesions have evoked considerable attention in recent years. It has been shown that lesions anywhere in the brain-stem can cause disturbed consciousness. Tumours, hæmorrhages, concussion and other factors have been widely studied. From case reports it is apparent that the liability to unconsciousness in brain-stem lesions varies with, among other things, the rapidity with which the lesion develops. Temporary ischæmia as one of the causes of intermittent attacks of unconsciousness appears a logical concept. We feel that atheromatous stenosis of a vessel lumen sets the stage for these intermittent episodes to occur.

The concept of a waking centre and the introduction of the term parasomnia to denote damage resulting in a "mixture of sleep and illness" is well known. Anatomical localization of such a centre is still unsure but the reticular formation in the medial tegmentum of the mid-brain is attracting most attention at the present time. Extensive areas of softening in the brain-stem, occurring without alteration of consciousness, have been evident to pathologists for many years. However, when a large area is temporarily deficient in blood supply and collateral circulation is poorly developed, the situation is apparently not comparable. The degree to which consciousness is altered is extremely variable and may persist for long periods.

Six months prior to death a 62-year-old housewife noted frequent transient attacks of blurring of vision of both eyes, generalized weakness, and brief paræsthesiæ in the left hand. One month prior to death, left frontal headache, vertigo, and tinnitus occurred which was persistent and increasing in degree. Teichopsia was an occasional complaint. Fourteen days prior to death she became bedridden and exhibited a pathological desire for sleep, and terminally was sleeping most of the day and night and difficult to arouse. A few days later, difficulty in swallowing was apparent and she became increasingly unresponsive. On examination her blood pressure was 130/75, she responded to painful stimuli only, the left corneal reflex was absent and there was left facial weakness and a right Babinski response. Terminally there was flaccid quadriplegia, contracted pupils, and deep coma. At post-mortem there was extreme atherosclerosis and stenosis of the basilar artery with recent thrombus formation. The right vertebral artery showed both old and recent thrombi and the left vertebral artery contained a partially occluding recent thrombus.

A 69-year-old hypertensive male had had three episodes of right hemiplegia in a five-year period prior to his death. All episodes had cleared with minimal residua. One month prior to death he developed incontinence and three days later developed a flaccid quadriplegia with bilateral Babinski responses. At this time he lapsed into a somnolent state from which he could be aroused with increasing difficulty. This state lasted for thirty-two days with terminal deep coma. Post-mortem revealed old and recent pontine softenings, extensive basilar artery stenosis and thrombosis. No cerebral pathology was evident.

Thus, while these patients invariably die in coma we may have a variety of experience in respect to their states of consciousness in the pre-coma stage of their illness.

(b) Dysarthria and Mutism

Dysarthria is one of the commonest symptoms in both severe classical cases and also in the intermittent episodes preceding basilar thrombosis which seem to be the result of transient anoxia of the brain-stem. Mutism can occur alone, develop on dysarthria, or it may be seen with hemiplegic symptoms and signs. We do not feel the mutism is necessarily anarthria or indeed that it is due to pseudobulbar palsy, and bilateral involvement of the cortico-bulbar tracts in each case would hardly seem a rational explanation. We do not feel it is transient motor aphasia. A relationship might be contemplated to that state called akinetic mutism because of the frequently associated intermittent complaints of profound weakness. We feel this symptom is not clear in terms of its pathophysiology and certainly merits reconsideration in cases coming to attention.

A 63-year-old widow developed episodes beginning with paræsthesiæ of her upper lip and right hand, and a sensation of heaviness in the right arm and leg. Almost immediately she experienced extreme coldness of the tip of her nose, her upper lip, her right hand and her right foot. She was quite mute and unable to utter any sound, she could still move her limbs and never lost consciousness. Months later these attacks became associated with uncontrollable weeping but pathological laughter was never present.

A woman patient developed episodes of vertigo, uncontrollable rolling conjugate movements of her eyes, and paræsthesiæ of her left arm and right leg. The paræsthesiæ were accompanied by weakness of these limbs. The attacks were brief, lasting 30–60 seconds but became progressively longer with the months. One year later they lasted up to fifteen minutes and at that time she developed complete mutism for the whole episode, followed by normal speech.

(c) Pontine Hemiplegia

Hemiplegia as an initial symptom occurs very frequently. This may be transient, or may be severe and quickly followed by quadriplegia. There may only be weakness and heaviness experienced in the arm and leg. Where crossed hemiplegia occurs a localization appears certain. Hemiplegia of an arm and leg, almost equally affected and without facial involvement, suggests possible thrombosis of a vertebral artery. Further, if on the dominant side and involving face, arm and leg, without sensory or speech disturbance, and with early and severe spasticity then the lesion is suspected as being in the anterior pons. Penetrating branches of the basilar artery are known to come off at right angles. The origin of these vessels may be affected by severe atheroma or thrombosis. Thrombus formation is probably lateralized early and this may account for the initial hemiplegic findings. Anatomically we might expect to see an occasional bilateral anterior pontine softening resulting in a paraplegia or quadriplegia.

A 76-year-old man with normal blood pressure had a sudden attack of vertigo and loss of consciousness three years prior to death. He was then apparently well for about one year when he developed a slowly progressive spasticity of both legs and a little more marked on the left. He became incontinent of urine. Over the next two years he had repeated transient attacks of acute vertigo associated with severe generalized weakness. Terminally signs of spasticity occurred in both arms. Three days prior to death he developed a series of "tonic fits" and death occurred with hyperpyrexia. At post-mortem there were old and recent softenings in the pons, most marked anteriorly, and the degree of stenosis of the basilar artery was remarkable. There was recent thrombus formation. No other pathology was found to account for the symptoms.

(*d*) *Vertigo and Tinnitus*

These symptoms occur so frequently that it suggests that Deiter's nucleus is the most sensitive mechanism in the pons to react to transient anoxic effects. We have not encountered decrease of acuity of hearing or deafness as a complaint. When vertigo and tinnitus occur suddenly, are persistent, or even increasing in severity they appear to constitute a bad prognostic feature. In one patient briefly mentioned in respect to pathological somnolence, she voluntarily retired to bed in her terminal illness because of persistent and increasing vertigo, buzzing tinnitus, and teichopsia. In typical basilar artery thrombosis a frequent history is of a sudden onset of profound weakness, acute vertigo, nausea and vomiting and then coma developing.

(*e*) *Miscellaneous*

Brief attention is drawn to several other features of interest.

(1) *Epilepsy.*—This was seen in 3 cases. All were in deep coma. 2 showed "tonic fits" that were mistakenly labelled decerebrate rigidity by the house staff. One already mentioned had terminal status epilepticus. Epilepsy only appears to occur in the terminal phases.

(2) *Paræsthesiæ.*—Lateralized sensory disturbance of lip, hand and foot occurred in 3 cases in early symptomatology. This has been previously recorded. Paræsthesiæ of the trunk did not occur, hemianæsthesia was noted in one case only. The lateralized sensory complaints, particularly if accompanied by visual symptoms, are almost indistinguishable from those seen in migrainous aura.

(3) *Weakness.*—The most frequent subjective sensation experienced by these patients was a feeling of profound weakness. While observations are inadequate this did not appear to parallel blood-pressure variation. A profound vasovagal mechanism suggests itself.

(4) *Behaviour.*—Abnormal behaviour occurred occasionally in these patients. This included amnesia, hysterical patterns, transient paranoid reactions, noisy confusion and personality regression. Some cases suggest a possible thalamic involvement from interference with blood supply but this remains speculative.

Conclusions

The usual clinical presentation of basilar artery occlusion by thrombosis and embolism is well known and allows ante-mortem diagnosis. The clinical variants dependent on the site of occlusion being proximal or distal in the vessel are also understood.

The usual assumption as to the cause of isolated or repeated vascular brain-stem lesions has been occlusion of penetrating pontine branches of the basilar artery. We would suggest that in certain of these cases, the symptoms occur as a result of stenosis of the basilar artery, in a similar fashion to that seen in carotid stenosis.

The symptoms preceding thrombosis of the vertebral and posterior inferior cerebellar arteries may have similar implications.

Arteriosclerotic stenosis of larger cerebral arteries is probably more important than has been shown in causing minor cerebral attacks.

Basilar artery stenosis as a cause of brain-stem symptomatology is probably more common than is generally appreciated.

Pure Word Blindness Considered as a Disturbance of Visual Space Perception [*Abridged*]

By J. Purdon Martin, M.A., M.D., F.R.C.P.

(1) Pure word blindness is a condition in which the patient is unable to read, though showing no signs of aphasia. The fact that he is able to write shows that he has not lost the memory of the word forms or of their symbolic meaning. What he has lost is the ability to recognize the word forms, and because of this he cannot read even his own writing.

(2) In cases of gross disturbance of visual space-perception of the kind described by Holmes and Horrax the patients are unable to read, but if the postural sense is normal in the patient's right hand he is able to write. He thus presents the essential features of pure word blindness. Such a patient is unable to locate objects in the field of vision or marks on a paper in front of him, he is unable to draw a line between two points, or to see the relations of lines to each other. He has lost what may be called his visual geometrical appreciation or visual geometric sense.

Since I became aware in 1938 of this variety of word blindness I have not seen any case of pure word blindness in which there were not also disturbances of visual space appreciation of geometric type.

(3) Letters and words are geometric formations and are arranged geometrically in lines and pages. For instance each of the letters A, H, K, N, F, Y consists of three short lines, in different geometrical relationship to each other. The child when he goes to school has to learn to recognize these forms and to discriminate between letters of similar form such as M and N, M and W. As he does so he is cultivating his visual geometrical appreciation. Later he learns short words and they become