BASILAR ARTERY THROMBOSIS — A REPORT OF 2 CASES

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Basilar artery occlusion was previously thought to be rare, as Biemond in 1951 stated "Thrombosis of basilar artery is regarded as rare and its clinical diagnosis difficult." But since the publication of Kubik and Adams' (1946) important paper, the syndromes of basilar artery occlusion have been increasingly recognised.

Two cases of basilar artery thrombosis seen recently in Medical Unit III are reported below.

CASE 1

A 46 year old Chinese labourer was admitted to Medical Unit III on 31.5.65 with a history of sudden weakness of all limbs and loss of consciousness on the morning of admission. As the patient was unable to talk, the history was obtained from a friend. On that morning, he was on his way with some friends to a coffee shop for a drink when he suddenly felt weak and fainted. He regained consciousness after a few minutes and was able to walk with the aid of his friends. Ten minutes later his speech was noted to be slurred. No convulsions, frothing at mouth or incontinence of urine were seen.

He was said to have had several similar fainting attacks during the two months prior to admission. After the fainting attacks, he would complain of transient hemiparesis and numbness of his left limbs, lasting for about half an hour. Between the attacks, he was apparently in good health, able to perform his duties without any difficulty.

He was an opium addict for 15 years. His past history and family history were non-contributory.

On the day of admission, his general condition was poor. He was very drowsy but could be aroused. He was afebrile, temperature being 98°F, and aphasic. No external injuries were detected. Both pupils were constricted with poor response to light. There was marked arcus senilis. Fundoscopy did not reveal any abnormality.

Both carotid arteries were palpable and the radial arteries felt thickened. His blood pressure was 130/90. No abnormalities were detected in the heart, lungs and abdomen.

He had a right infranuclear VII nerve palsy. All his limbs were spastic, the right side being slightly more than the left. There seemed to be no response to pain at all. The tendon reflexes, except for the superficial abdominal reflexes which were absent, were brisk and equal. The plantar response on both sides was flexor.

On the following day, he developed a fever of 102°F. His blood pressure was 185/100. He was still in a state of drowsiness and aphasic. He was noted to yawn occasionally. The spasticity of the limbs was found to be alternating with flaccidity on a few occasions. No clonus could be elicited. The plantar response became bilaterally extensor.

There was no change in his general condition and neurological disability until the fifth day when he developed a very high fever of 104·8°F and his coma deepened. He had Cheynes-Stokes respiration and his limbs became more spastic. His general condition showed rapid deterioration and on the same day he died soon after a bout of vomiting.

The following investigations were done. Haemoglobin was 94% and the total white cell count was 14,800 (Polyp. 92%, Lymph. 5%, Mono. 3%, Eos. 0%). Blood film was negative for malarial parasites. Blood urea was 33 mg.%. Blood sugar (random specimen) was 95 mg.%. The pressure of cerebro-spinal fluid was 30 mm. with no evidence of block. The fluid was clear with normal constituents. The culture and smear were both negative for organisms and Cryptococcus. Kahn test was negative both for blood and cerebro-spinal fluid.

At autopsy, the following findings were noted. Sections of the coronary arteries showed an advanced stage of atherosclerotic narrowing of the lumen. In the anterior descending branch of the left coronary artery, there was evidence of previous thrombosis and recanalisation. Exten-

sive fibrosis of the wall of the left ventricle was noted, corresponding to the blood supply of this artery. There was evidence of bronchitis and aspiration pneumonia in the lungs. The left adrenal gland appeared enlarged and histological section showed hyperplasia of the cortical cells forming nodular areas in the cortical layer. In the brain, sections from the pons showed evidence of a recent infarction. Serial sections of the basilar artery showed marked intimal thickening. A recent thrombus was seen in its lumen. The vertebral arteries were patent.

CASE 2

The next patient was a 64 year old Chinese businessman who was said to have been apparently in good health until the morning of admission on 17.10.65 when he failed to wake up at the usual time and was found to be unconscious. There was no history of any head injury prior to loss of consciousness. He had no previous history of headache, hypertension, diabetes or convulsions. His past history and family history were not significant.

At the time of admission, he was in a state of coma, responding only to painful stimulation. His temperature was 98°F. His blood pressure was 110/70. The pupils were equal and reactive to light. Fundoscopy did not reveal any abnormality.

There was no neck rigidity. All his limbs were flaccid and sensation to pain was intact. The reflexes in the limbs were exaggerated and equal. The superficial abdominal reflexes were absent. The plantar response was bilaterally extensor. The other systems were found to be normal.

The cerebro-spinal fluid was clear with normal pressure. The constituents were all within normal. Blood urea was 49 mg.%. Blood sugar (random specimen) was 138 mg.%. The skull X-rays were normal.

A few hours after admission, he developed a fever of 105·5°F and his coma deepened. He demonstrated alternating spasticity and flaccidity of all the limbs. On the following day, he remained in deep coma with constricted pupils, which were non-reactive to light. His limbs were spastic and there was no change in the tendon reflexes. His temperature remained high and just before death it reached 107·4°F. He died on the second day of admission.

Consent for only partial autopsy of the skull was obtained. This showed that the pons was swollen with multiple petechial haemorrhages. There was softening of the cerebellum. The basilar artery was atheromatous with a fairly extensive thrombus.

In both patients the treatment was essentially the management of coma. No anticoagulants were administered.

DISCUSSION

The vertebro-basilar system supplies a far more extensive territory than is generally realised. Awareness of the cephalad distribution of its branches is essential to the understanding of symptoms which arise. Three sets of branches supply the medulla, pons, mid-brain and cerebellum:-

1. Paramedian arteries, three to four in number, which are short and have a small lumen;
2. Circumferential arteries, numbering three to four, which stretch far out in the lateral pons. They have few side branches, and are long and tenuous.
3. Long circumferential arteries with a wide lumen and thick walls. These are the superior cerebellar, anterior and posterior inferior cerebellar arteries; they supply the cerebellar lobes as well as the brain stem.

The diencephalon medial to the territories of the anterior choroidal artery and striatal branches of the middle cerebral arteries, is supplied by the basilar system and its terminations in the two posterior cerebral arteries. Other areas that depend upon the basilar system include the cerebral peduncles, substantia nigra, red nucleus, and medial and ventral parts of the thalamus and the subthalamus. The posterior cerebral arteries, supplying the occipital lobe and all the visual cortex also supply the posterior, inferior and anterior parts of the temporal cortex through its parieto-occipital and anterior temporal branches. Finally, its branch the internal auditory artery supplies the middle ear.

Brain (1957), Wells (1960) and Hutchinson (1962) have made a review of the very important and variable role of the circle of Willis and other collateral mechanism within the head and neck in maintaining blood-flow when one or more of the large vessels is stenosed or occluded, thus

making possible the maintenance of function in brain-stem and hemispheres. It has been demonstrated by Alpers et al. (1959) and Alpers and Berry (1963) that while there are "normal" circles of Willis in 52% of subjects with normal brains, such a state is found in only 33% of the subjects with cerebral softening. Riggs and Rupp (1963) found that in 79% of 944 subjects with "neural dysfunction", there was evidence of hypoplasia of one or more components of the circle.

Basilar artery occlusion may present without significant warning symptoms, as seen in the second patient, or with fluctuating symptoms advancing to severe disability or death, as exemplified by the first patient. The latter may closely resemble the fluctuating downhill course resulting from a brain-stem tumour. In their series of patients studied over a period of five years, Williams and Wilson (1962) had not encountered one fatality without preceding symptoms, often minor, for days or weeks before death, except in the case of embolism of the basilar artery. The transient symptoms included white or black spots in the field of vision, which were easily overlooked or dismissed as unimportant by the patient and vertigo often mistaken for "Mènière's". Several of Kubik and Adams' (1946) patients, who eventually died from total occlusion of the basilar artery also had noted premonitory symptoms including blurred vision, diplopia, facial paraethesiae, vertigo, ataxia and dysarthria, and others had had attacks of disturbed consciousness or of transient hemiparesis or tetraparesis before their final illness.

The first patient had a history of frequent fainting spells associated with transient hemiparesis and numbness of the left limbs before his final fatal illness. It was rather unusual for the second patient to have no premonitory symptoms. As the history was obtained from a relative, the patient might have had minor preceding neurological disability without his knowledge.

The initial symptoms most commonly encountered are vertigo, visual hallucinations, lowering of consciousness, nausea, ataxia, diplopia and drop attacks, in that order. Sensory, hemiplegic, trigeminal or auditory symptoms are rather uncommon. Occipital headache is often noted. There my be sensory change in an arm at the same time, if the vertebral artery is also involved.

These premonitory symptoms were attributed to atherosclerotic narrowing of the vertebral and basilar arteries and intermittent ischaemia of structures in the brain stem was due to the resulting diminished blood flow. These symptoms, which were characterised by their complete reversibility with normal interludes, were related to inefficiency of the basilar system and could be caused by narrowing of the artery, structural anomaly or arterial hypotension. The term basilar insufficiency was suggested (Denny-Brown, 1953; Williams, 1958) to indicate the relationship between symptoms and reduced blood flow.

Other features that suggest the diagnosis of basilar insufficiency include the multiplicity of symptoms, the variability of their intensity in time, the localisation of these disturbances to the territory of the vertebro-basilar system and the absence or poverty of signs of neural damage or of gross lesions.

Both patients described in this paper presented with coma and spastic tetraplegia, indicative of brain stem infarction. Their sudden onset and the rapid deteriorating course were very suggestive of a cerebro-vascular catastrophe. These patients also showed a rather interesting feature which was not previously reported, *i.e.*, alternating spasticity and flaccidity in all the limbs, simulating stiff-man syndrome. The plantar response was also variable in the first patient. But the ultimate clinical picture was a syndrome of coma and spastic tetraplegia when the clinical diagnosis in both patients became evident.

Other predisposing causes of vertebro-basilar ischaemia which must be considered include the effect of neck movements (Sheehan et al., 1960) particularly at the atlanto-occipital joint (Brain, 1963) and extreme rotations, as in backing a car, therapeutic manipulations of the neck (Hallberg, 1957), especially with cervical arthritis (Sheehan et al. 1960) and the effects of arterial hypotension (relative to the needs of that particular patient) from any normal or abnormal cause, including ischaemic heart disease and fatigue. Anaemia as a precipitating cause is often overlooked. Subclavian steal syndrome must also be borne in mind.

Thrombotic occlusion of the basilar artery is almost fatal, as pointed out by Kubik and Admas (1946) in a classical review of 18 cases, confirming the earlier observations of Lhermitte

and Trelles (1934). The prognosis seems to be better when there are known predisposing or precipitating causes of vertebro-basilar ischaemia, such as mentioned earlier.

The clinical diagnosis could be confirmed by vertebral angiography. But before this diagnostic procedure is undertaken, a distinction must be made between the young generally healthy patient experiencing distressing and ominous attacks due to gross local stenosis for which reparative vascular surgery might be indicated, and the elderly and arteriopathic patient or hypotensive subject in whom arteriography is hazardous, as serious complications, sometimes followed by permanent field defects, at the time of the procedure have been reported. Dobrak, Beck and Murphy (1960) have drawn attention to the dangers inherent in the techniques of cerebral angiography in patients over the age of 50 and in their opinion, the procedure had contributed to the fatal outcome of the illness in 4 of 16 patients who had died from a group of 39% of 58 patients in whom angiography demonstrated abnormality of large vessels.

It is apparent that vertebral angiography by direct puncture is not justified in obvious cases, since it is unlikely to lead to any appropriate treatment. In most cases, in view of the hazards and complications that may be encountered in this procedure, we have to rely upon unaided clinical diagnosis in the management of these patients. On the other hand, Haugsted (1956) and Meyer, Sheehan and Bauer (1960) felt that angiographic investigations must be pursued in view of the advance in surgery of the vertebral arteries, particularly in their origin from the aortic arch. For this purpose, the whole territory of the vertebro-basilar system may be outlined including the origin of these vessels in the thorax, either by retrograde subclavian arteriography, or by improved techniques of intravenous arteriography.

In the study of the electroencephalogram of their patients, Williams and Wilson (1962) noted that about a third of them showed deviations from the normal in the records made from electrodes placed along the length of the Sylvian fissure, in the form of low voltage waves or rhythms of 2 or 3 a second. Occasionally faster waves were seen, but gross changes or focal spikes were uncommonly encountered. The disturbances were bilateral and often asymmetrical. As neurological investigations showed no structural disturbance within the hemispheres, two possible explanations for these electroencephalographic abnormalities were suggested. The first was that ischaemia of the upper brainstem might affect the eletrical activity of the temporal lobes through the mid-brain reticular structures which have close connections with the hippocampus and other deep temporal structures or that ischaemic changes were being reflected in the electroencephalogram through reduction in flow in the territory of the anterior temporal artery which takes origin from the posterior cerebral branches of the basilar artery.

In the management, the first medical decision is whether or not to use anticoagulants. Shaw (1962) in his review of the subject has come to the conclusion "their use should be limited to the treatment and prevention of cerebral embolism, actively progressing neurological deficit, particularly in the vertebro-basilar territory, and transient ischaemic attacks involving either the carotid or vertebro-basilar system." However, this view has not received wide acceptance. Some feel that intermittent disturbances due to stenosis proximal to the circle of Willis constitute a lesion like that in the coronary circulation, in which, given time, a collateral circulation may become established and anticoagulants may prevent the extension of thrombus during that time.

Anticoagulant therapy has been shown, notably by Milikan and his colleagues (1956), to be of great value in cases of recurrent vertebro-basilar insufficiency. They found that the attacks were abolished or reduced in 90% of patients given long-term treatment, and in 107 treated patients the mortality rate was only 8%. In a smaller but comparable series of patients who were not treated the mortality rate was 50%. On the other hand, the experience of Bradshaw and McQuaid (1963) was substantially different. Only 6 of their patients were treated with anticoagulant drugs and one died from thrombosis of the basilar artery, 4 of the patients did well and one continued to have attacks as before. Of the remaining 48 cases, 1 died nine months later from a stroke and 2 others could not be traced; 14 have neither symptoms nor signs, 15 are free from attacks but have minor neurological signs and 7 have occasional attacks and show only minor neurological disability. 9 patients only were unchanged or had got worse and of these 3 showed evidence of progressive neurological disturbance. Thus 36 patients had done very well without specific

treatment and were able to lead normal lives at the time of follow-up six months to five years after diagnosis. Thus it is clear that the present treatment consists mainly of general medical management, and the value of anticoagulant therapy remains to be assessed.

SUMMARY

Two cases of basilar artery occlusion are described. An interesting feature of alternating spasticity and flaccidity shown by these two patients in the early phase of their illness was noted. A brief review of this disorder is made, including a short discussion on the mode of presentation, investigations and management of vertebro-basilar ischaemia.

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