The foot that spoke - Rehabilitation in Midbrain locked-in syndrome, a case report

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Abstract: Posterior circulation strokes account for approximately 20% of all ischemic strokes. Of these, 11% involve multiple arterial territories. Survivors of insult to multiple regions are left with extensive neurological deficits and present a unique challenge to the rehabilitation team. We present a case of recurrent posterior circulation stroke involving extensive areas of the brainstem, cerebellum, bilateral thalami and occipital cortex. Fluctuating sensorium and complete bilateral ophthalmoplegia along with anarthria and quadriaparesis made initial assessment of level of consciousness difficult. The establishment of consistent communication and challenges posed to cognitive and physical rehabilitation are discussed.

Keyword: posterior circulation stroke, midbrain, locked-in syndrome, rehabilitation

Case Study: The foot that spoke - Rehabilitation in Midbrain locked-in syndrome

Abstract: Posterior circulation strokes account for approximately 20% of all ischemic strokes(1). Of these, 11% involve multiple arterial territories(2). Survivors of insult to multiple regions are left with extensive neurological deficits and present a unique challenge to the rehabilitation team. We present a case of recurrent posterior circulation stroke involving extensive areas of the brainstem, cerebellum, bilateral thalami and occipital cortex. Fluctuating sensorium and complete bilateral ophthalmoplegia along with anarthria and quadriaparesis made initial assessment of level of consciousness difficult. The establishment of consistent communication and challenges posed to cognitive and physical rehabilitation are discussed.

Background -

The vertebrobasilar system of arteries consists of the paired vertebral arteries, the basilar artery, and the paired posterior cerebral arteries, which supply blood to the upper spinal cord, brainstem, cerebellum, and parts of the subthalamus, thalamus and hippocampus (3). The intracranial vertebrobasilar system can be divided into proximal, middle, and distal territories (4). Infarcts involving multiple brain stem levels may have overlapping features and are very difficult to diagnose and localise (5). The commonest cause for these infarcts is intraarterial embolism from vertebral artery occlusive disease. Patients with multiple intracranial territory infarcts have a higher risk of poor outcomes (4).

Case Presentation –

A 49 year old businessman was admitted for inpatient rehabilitation with complaints of decreased responsiveness to external stimuli, weakness of bilateral upper and lower limbs and dependence for all activities of daily living since the last 7 months. He was known to be under treatment for hypertension since 15 years and diabetes mellitus since 1 year and was apparently well till seven months earlier when he had an episode of giddiness and unsteadiness while driving his car. He was managed on an outpatient basis for hypoglycaemia and recovered within a few hours. 2 days later, he developed sudden abnormal tonic posturing of bilateral upper and lower limbs, slurring of speech and altered consciousness. He was taken to a hospital where an MRI revealed infarcts in bilateral cerebellum, superior vermis, right half of pons, midbrain and red nucleus with complete occlusion of right vertebral artery. He was intubated and ventilated and a nasogastric tube was inserted for feeding. He was managed conservatively with dual antiplatelets and antihypertensives and a tracheostomy was done in view of need for long term management of ventilation and secretions. He was discharged to a nursing home within 5 days. Five months post stroke, the tracheostomy was decannulated. He was discharged and taken home. He developed one episode of aspiration pneumonia which was managed at a local hospital. He was then referred to our centre for further evaluation and neuro-rehabilitation. At the time of admission he was on a nasogastric tube for feeding and indwelling urethral catheter. His GCS was 6/15 i.e. E1(No eye opening), V2(Incomprehensible groaning sounds), M3(Abnormal flexion to pain – dorsiflexion of the right foot was seen in response to sternal rub). On manually lifting the eyelids both eyes were noted to be deviated downwards and upwards, both pupils were 4mm and not reactive to light, corneal reflex was present bilaterally, no spontaneous eye movements were noted and oculocephalic reflex was abnormal. The Coma Recovery Scale-R score was 5/23 corresponding to a vegetative state (6). Tone was increased in left upper and lower limb and normal in right upper and lower limb. Deep tendon reflexes were brisk bilaterally and plantars were upgoing.

Fig 1: MRI and MRA showing bilateral thalamic and midbrain infarcts and attenuated right vertebral artery.
complete bilateral oculomotor nerve paralysis(11). Following this, characterized by quadripariesis, anarthria, total bilateral ptosis and Berg in 1979 described a “Mesencephalic Locked-in Syndrome” This pattern of involvement is known to cause fluctuating sensorimotor and complete bilateral ophthalmoplegia along with anaphesia and quadriaparesis. These symptoms and signs are well explained by his imaging findings of bilateral thalamic, midbrain and pontine infarcts as described above. The JFK Coma Recovery Scale (CRS) was initially described by Giacino et al in 1991. It was developed to characterize and monitor patients with disorders of consciousness, with the aim of prognostic assessment and treatment planning. It was revised in 2004 and is now known as the JFK Coma Recovery Scale-Revised (JFK CRS-R)(6). It has since then been validated and has become the old standard for assessment of level of consciousness(13). There is still however a high percentage of patients with disorders of consciousness who are erroneously assigned a diagnosis of vegetative state (14). The high rate of misdiagnosis has been attributed to physician, patient and environmental factors(15). In our patient, fluctuation in sensorium, ophthalmoplegia, motor impairment and aphasia led to initial misdiagnosis of vegetative state and goals were set accordingly. With the evolving voluntary control of his right foot, goals were reset and he showed remarkable progress in communication. He was able achieve consistent functional communication and indicate his needs. This improved his quality of life and his interaction with his family. In the largest survey of chronic locked-in syndrome patients till date, more than 70% reported good subjective well being implying that patients with severe disabilities may have a good quality of life despite being socially isolated and dependant for activities of daily living(16). Every effort should be made to ensure that these patients and their families have access to comprehensive multidisciplinary rehabilitation programmes to maximize functional recovery.

Conclusion:
This case highlights the importance of repeated and accurate assessment in patients with disorders of consciousness. Failure to detect signs of consciousness may lead to early termination of treatment and lower goals of rehabilitation especially in the domain of communication.

References:
10. Saunders.