LETTER TO EDITOR (VIEWERS CHOICE)

PERIPHERAL LOCKED - IN SYNDROME FOLLOWING SNAKE ENVENOMATION

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A 4 years old girl bitten by a snake at about 1.30am presented after 3 hours with drooping of eyes, loss of voice and paralysis of all 4 limbs. Endotrachial intubation was done in view of apnea at another hospital and she was shifted to our hospital in 30 minutes. Physical examination at the emergency room revealed an unresponsive flaccid child with a Glasgow Coma Scale of 3, dilated unreactive pupils, bilateral ptosis, absent dolls eye movement due to complete ophthalmoplegia, absent superficial and deep tendon reflexes and complete paralysis of limbs and facial muscles. Her heart rate was 160/min, blood pressure 140/80mmHg and she was on manual ventilation. Her distal pulses were weak. Two fang marks were seen in the right arm without evidence of local envenomation. A provisional diagnosis of neuroparalytic snake envenomation (Krait) was made. Shock was corrected with 20ml/kg of normal saline (NS) and child was shifted to the Pediatric Intensive Care Unit (PICU) where she received 20 vials of polyvalent anti snake venom. Neostigmine did not show any improvement in the neuroparalysis. A mechanical ventilator in the intensive care unit could not be arranged due to full
occupancy and manual ambu-bag ventilation was continued. Child remained unresponsive without any improvement for up to 48 hours of admission showed signs of recovery initially in the big toe movements in response to mother's request to move the toe indicating that the child was conscious, followed by thumb movements at 49 hours. Dolls eye movement was elicitable on day 4 of hospitalization and ptosis started improving from day 5. Deep tendon reflexes were elicitable by day 6. Pupils were reactive from day 7 and spontaneous limb movements on day 8. Child was manually ventilated for a period of 5 days and 16 hours, mechanically ventilated from day 6 and was extubated on day 13. Minimal head lag and limb weakness persisted at the time of transfer to the ward. Child was discharged on day 19. Follow up after 2 weeks of discharge revealed complete neurological recovery. However the child was too young to recall the events during LIS but for remembering the painful pricks and nasogastric tube insertions, which she said she could perceive but unable to react at that time.

Locked-in syndrome (LIS) is characterized by total paralysis and anarthria in a conscious patient. (1) Complete LIS is a state where the patient cannot communicate in any form. Central LIS, the more commonly encountered form is due to lesions in ventral pons.(1) Recently described are the peripheral causes of LIS, severe acute polyneuropathy, neuromuscular junction blockade (myasthenia gravis, toxins)(1) and snake bites.(2) Knowledge about the peripheral cause of LIS is very vital, as one may make an erroneous diagnosis of brain death in LIS. (1-3) Confirmatory tests like electroencephalography, cerebral blood flow, nerve conduction velocities are recommended to avoid a misdiagnosis of coma or brain death.(3) However the rise in heart rate following atropine injection prior to neostigmine in this child did give a clue that the child is not brain dead. The child we have described is a complete peripheral LIS due to snake bite. There are no pediatric case reports of a total peripheral LIS following snake bite. She is also the first reported critically ill child to have survived after prolonged manual ambu-bag ventilation (5 days and 16 hours). However there are a few previous case reports of brief manual ventilation in neuroparalytic snake bites in adults.(4-6) Recently, reports from Lucknow have documented that prolonged ambu bag ventilation can be life saving in adults with snake bites.(7) Thus, in any child presenting with sudden onset of unexplained flaccid paralysis, snake envenomation has to be suspected, as treatment is lifesaving. It is thus important that emergency room physicians recognize locked-in syndrome, to prevent the dangerous errors of diagnosing brain-death/coma in snakebite victims presenting unresponsive with dilated unreactive pupil but conscious and non communicative. Manual ventilation with self inflating manual resuscitation bag (ambu bag) can save the life in a resource constrained situation.

REFERENCES

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