



Locked-in Syndrome in Snakebite

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Abstract

Two young patients are described who made complete recovery from locked-in syndrome (LIS) after snakebites. LIS was a presenting feature in a patient of presumed snake bite who showed complete response to polyvalent Anti-snake venom (ASV). This case suggests that elapid snake bite should be suspected in unresponsive patient found in early morning in endemic areas of snake bite in monsoon season. The second case was admitted in complete LIS state with history of rapidly progressive ptosis, diplopia, ophthalmoplegia, bulbar symptoms and quadriparesis, 6 hours after snake bite. Complete improvement by ASV in second patient highlights the need of differentiation of complete LIS from coma and brain death in patient of snake bite, as former have good prognosis. ©

INTRODUCTION

Locked-in syndrome (LIS) is characterized by quadriparesis and anarthria in conscious patient. Patient can be communicated by eye movement and blinking. Communication is not possible in total LIS.¹ LIS may be either of central, usually ventral pons¹, or of peripheral origin.² Diagnosis of central cause is important for rehabilitation purpose, as even limited physical activity can improve quality of life.¹ Knowing the peripheral causes is very important, as one may make erroneous diagnosis of brain death in a patient with complete LIS due to fulminant polyneuropathy and severe neuromuscular junction Blockade.^{2,3} In two case reports, we discuss the importance of LIS in patients of snakebite and review the literature.

CASE REPORTS

Case-1

A 40 years male was brought to the emergency room in unresponsive state with no significant past history. In early morning patient did not show any response when his wife tried to make him awake. At presentation, patient had no motor response to painful stimuli. There was ptosis, no spontaneous eye movement and absent oculocephalic and oculovestibular reflexes. Deep tendon reflexes and planter reflexes were absent. Pupil was of 3 mm size with sluggish reaction to light. Physical examination revealed no abnormality. His respiratory effort was sluggish. Endotracheal intubation was performed and patient was mechanically ventilated. A provisional diagnosis of stroke was made, but MRI Brain

was normal. Hematological examination and blood\ serum biochemistry (sugar, creatine, liver function tests, electrolytes, and thyroid profiles) were within normal limits. Chest X-ray and cerebro spinal fluid (CSF) examinations did not reveal any abnormality. The electroencephalograph showed alpha and theta activity with no epileptiform discharges. Further careful examinations of skin revealed reddish spot over back of chest. It was not typical fang mark of snakebite, but patient was given injection polyvalent ASV. We noticed spontaneous eye movement within an hour of treatment. He communicated with eye movement. There was complete improvement of ptosis and ophthalmoplegia in about 30 hours. Patient was extubated after 3 days. Proximal muscle weakness took 7 days to improve completely. The patient could remember well the period when he was unresponsive. He did not give history of any possible snakebite or toxin and drug exposure.

Case 2

A 25 years old male was bitten on toe by an unidentified snake 8 hour prior to his admission. He was brought to emergency room in unconscious state with history of rapidly progressive ptosis, diplopia, dysphagia, dysarthria, dyspnoea, and weakness of all four limbs. Patient was unresponsive to painful stimuli. Light reflex, oculocephalic reflex, oculovestibular reflex, deep tendon reflexes and superficial reflexes were absent. Swelling at bite site was noted. Patient was mechanically ventilated because of poor respiratory efforts. Hematological examinations and blood/serum biochemistry were within normal limits. A provisional diagnosis of snakebite with hypoxic encephalopathy was made. Injection polyvalent ASV was started. There was spontaneous eye movement after 6 hour of treatment and patient communicated with eye movement. Patient improved completely in 5 days. Patient was able to recall

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the period of his unresponsiveness.

DISCUSSION

The locked-in syndrome (LIS) has been classified into three categories: Classical, Incomplete, and Total.¹ Classical LIS is characterized by quadriplegia and anarthria in conscious patient. Vertical eye movement and blinking are the only means of communication. Incomplete LIS is same as classic but with remnants of voluntary movement other than vertical movement. There is total immobility and inability to communicate in fully conscious patient in case of Total LIS.¹ Each of the three categories have been subdivided into transient and chronic forms.¹ The LIS is usually described in relation to damage of ventral pons by stroke. Non vascular etiologies of LIS affecting ventral pons are trauma, tumor, inflammatory conditions, etc.¹ In addition to ventral pons, LIS are reported due to involvement of bilateral cerebral peduncle, medulla, etc. Transient LIS due to involvement of brainstem are reported in case of TIA, post-infectious encephalitis, hypoglycemia, hyperglycemia, etc.¹ There are many peripheral causes of LIS, such as severe acute polyneuropathies (GBS), neuromuscular junction blockade (myasthenia gravis, toxins) etc.² The correct diagnosis is essential as peripheral causes of LIS are treatable conditions and complete recovery can be possible with timely interventions.²

Few cases of LIS after snakebite are reported in literature.³ But in case-1, LIS was the presenting complaint of presumed snakebite. Since krait bites are generally painless with little or no skin changes, victims may not feel or recognize the bite.⁴ Several cases of morning paralysis after krait bites have been reported⁵, but patient recognized as LIS in early morning have not been reported in literature. The common krait is nocturnally active snake and its bite is painless.⁴ In various studies 60%-70% of the cases of snakebite occurred when the patient were asleep. The site of bites were undetectable in 17% patients of snake bite in a prospective study.⁴ It is very challenging task to make diagnosis in a patient found in unresponsive states due to undetectable, painless bite during asleep state in night. High degree of suspicion is required. Unexplained unresponsiveness in a previously healthy person in early morning of rainy season, in areas where snakebites are common, should create the suspicion of possibility of snakebite even in the absence of history of snakebite or definite fang mark on clinical examination. Incidence of complication is directly proportional to the duration of venom in blood. Polyvalent ASV is relatively safe, and allergic reactions after ASV injection can be prevented by premedication with adrenaline, I.V. hydrocortisone and histamines.³ Therefore, suspected snake envenomation should be treated empirically with I.V. polyvalent ASV, and simultaneously should be investigated for differential

diagnosis and etiology of unresponsiveness,³ including the use of venom detection kit.

There was typical history of neuroparalytic symptoms after snakebite in our second patient. However, he was completely unresponsive to any stimuli at presentation. There are very few case reports of LIS after snakebite in literature.³ However, there are many reports of patient with coma who made a complete recovery. In one prospective study of snakebite, 22 patients out of 35 made complete recovery from coma.⁴ Author did not give exact reason for deep coma. However, complete improvement suggests that patients could be in Total LIS states. The differentiation of coma with LIS is essential in patient of snakebite. LIS is severe form of neuromuscular junction blockade, while coma may be due to complication of snakebite. Respiratory failure is the most common cause of mortality and morbidity in victims bitten by venomous snakes of elapidae family. Other causes of death are complication of mechanical ventilation, shock, intracerebral hemorrhage, complication of wound, tetanus, cortical venous thrombosis, renal failure, hypoxic brain damage, etc.⁶ Direct toxic effects of snake venom on brain parenchyma have not been reported in literature. Therefore, differentiation of LIS with coma and brain death is important in patient with definite history of snakebite for prognostic and management purpose. In fact, confirmatory tests like electroencephalography, cerebral blood flow, etc. are recommended in situation like LIS, where a misdiagnosis of brain death is possibility.³ Neuroimaging of brainstem is required to rule out central cause of LIS.

In conclusion, snakebite should be a differential diagnosis in a person who is found in unresponsive state in early morning. In addition, differentiation of LIS with coma is important in a patient with definite history of snakebite for prognostic and therapeutic purpose.

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