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Case Report

Speech language characteristics and intervention approaches in lockedin syndrome post snake envenomation

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ABSTRACT

The present case study illustrates hypoxic-ischemic encephalopathy as a result of neuroparalytic snake envenomation in an 11 year old male patient. Detailed speech assessment was done which revealed diverged oral structures and language loss. The study aims to document speech and language characteristics as well as rehabilitation in terms of speech therapy. The study enlightens the role of speech language pathologist in assessment and intervention of locked in syndrome. Study shows the swapped roles of traditional therapy approaches and the importance of augmentative and alternative communication as compassion and a beneficial technique in an intricate incident like hypoxic-ischemic encephalopathy as a result of neuroparalytic snake envenomation.

Keywords: Neurotoxins, Locked in syndrome, Augmentative and alternative communication, Kinetic mutism

INTRODUCTION

Snake poison is a complex combination of numerous affluence such as neurotoxins, enzymes, growth factors, activators, and inhibitors with a wide band of biological activities which causes different metabolic disorders by altering the cellular processes and biocatalyst activities of different organs. Irrespective of its modes of administration (orally or via snake bite) poison possess the same conclusions. Compounds transported by oral feeding effect are distributed to all parts of the body in their metabolized form. The pharmacological properties of toxins are sorted into subtypes like hemotoxic, neurotoxic, and cytotoxic. The neurotoxic nature of venom intrudes the neurons and their functioning while hemotoxic intrudes blood and cytotoxic the cells. The venom can lead to paralysis and an inability to control one's muscles. Toxicants can attack the aerobic system that supplies the human body with energy. The peril from venom depends on what class of snake it is and how much venom is found in the person's system. Locked-in syndrome (LIS) can best be described as a disease process where the brain is fully functional while confined within a non-functional body.[1] The locked-in syndrome is a rare neurologic disorder defined by the presence of sustained eye-opening, preserved awareness, aphonia or hypophonia, quadriplegia, or quadriparesis and complete loss of speech and language.

CASE REPORT

An 11 years old male reported to the ENT department of Maharishi Markandeshwar institute of medical sciences and research Mullana, Haryana, India in April 2019 with a chief complaint of inability to communicate with associated difficulties in walking and standing. He had a history of neuroparalytic snake envenomation with hypoxic-ischemic encephalopathy with a neurologic sequel in late 2017. The child was having complete speech and language development and was a healthy person before the episode of neuroparalytic snake poisoning. But after the episode, the child became

completely paralyzed resulting in regression of speech and language following a complete loss of it.

Speech and language assessment

Detailed speech and language assessment was administered on this child. Oral peripheral examination revealed all structures normal in appearances and inadequate in functions. Involuntarily tongue movements and inadequate lip seal could be observed which resulted in intermittent drooling. The child was not able to carry any direction of verbal and gestural commands. On language assessment, irregular and inappropriate social smile was present. Receptive vocabulary consists of the concept of eatables vs. non-eatables. Moreover, the child initiates vocalization directly by keen sighting the parents and sometimes for hunger. Speech imitation was poor. No utterance of single meaningful word or jargon speech could be discerned. Besides no self-help skill could be observed. The child was hypersensitive towards few eatables when placed in the mouth. No cognitive pre-requisites for language learning were evident after the episode of the neuroparalytic condition. Nevertheless, the child could perceive some forms of facial expression and voices expressing different emotions like sadness, anger and joy.

According to Reynell's attention scale, the child's attention was at level 1 which indicates that the child cannot attend to what you say. The child attends to a thing fleetingly however, gets distracted by any new activity. On formal testing like receptive expressive emergent language scale (REELS), receptive and expressive language scale corresponds to 1-2 months.

Audiological assessment

Immittance audiometry was administrated which revealed bilateral 'A' type tympanogram with both ipsilateral and contralateral acoustic reflexes present. Conventional test like pure tone audiometry couldn't be assessed since the child was cognitively not stable. OAE was done to find out the auditory integrity up to outer hair cells and it displayed the presence of both distortion product otoacoustic emission (DPOAE) and transient otoacoustic emission (TOAE) suggesting normal auditory integrity up to outer hair cells. Further ABR was performed which revealed hearing sensitivity within normal limits and healthy brainstem conduction of sound.

DISCUSSION

Language is a compute of intellectual capacity whereas speech is the physical fabrication of sounds. The acquisition of language is the process of building the ability to understand a language using it to communicate. Communication is every child's necessity for quality of life. The acquisition of language and speech is the affirmation of a developed intellectual child. Loss of language closes every access to forthcoming opportunities.

Locked in syndrome is the sleeping beauty of the medical world. It is a very rare condition that results in an extreme and sudden loss in the self-expression of linguistic, intellectual, emotional, and motor abilities. Locked-in syndrome has been recognized in medicine for ages but the diagnosis is delayed or entirely missed. Because of the rarity of the condition in children, the diagnosis of locked-in syndrome may be missed and patients may wrongly be considered as being in a coma, vegetative state, or akinetic mutism.[2] The mentioned case is an 11 years old boy with hypoxic-ischemic encephalopathy in a known case of the locked-in syndrome as a result of neuroparalytic snake envenomation with regression of speech and language development.

Locked in Syndrome in Ischemic Encephalopathy due to snake envenomation is a rare entity which causes damage to different structures of the brain involving cerebrum, cerebellum and brainstem (predominantly). Broca's and Wernicke's area are two primary known centre for speech and language production located in the frontal and temporal part of the brain respectively. The coordination of voluntary muscles of different oral structures, language processing centre and balance maintenance are the foremost task of the cerebellum. Besides, the different cranial nerves which conduct distinct nerve impulses in and out of the brain are located in the brainstem. Any anomaly that intervenes their blood circulation and nerve conduction cause impediment in speech and language production as in the prior mentioned case.

The presence of sustained vertical eye movement and opening, preserved awareness, aphonic or hypophonic voice, pathological bite reflexes, quadriplegia, unresponsive to pain stimuli, difficulty with conscious swallowing and breathing and complete loss of speech and language are the chief characteristics exhibited by an individual with locked-in syndrome.

Another important term to be contemplated is kinetic mutism which is a very rare neurological condition in which an affected individual does not move (akinetic) or talk (mute) despite being awake and are in a conscious state with normal sleeping patterns. The rehabilitation of locked-in patients is a long process and requires close cooperation of a specialized group of experts.[3]

The management of this case is done in different domains consisting of speech language therapy, physiotherapy and medical interventions. Speech language therapy focuses on communication needs and moulding of inverted J shaped graph of deprivation of language into a linear function of the development of speech language skills.

Initially, the traditional speech-language approach was given the chief focus. However significant progress could not be observed that might help the child in a longer

duration. Hence, usage of augmentative alternative communication (AAC) was emphasized. For the restoration of the disabling condition, AAC is important. AAC can help in managing the frustration, anger, isolation, depression, and can focus to enhance meaningful interactions. The main goal is identifying the needs and the assistive technology application for cognition and language impairment. AAC enlightens the residual challenges and capabilities. AAC can be used in a creative way to improve their participation in meaningful communication activity.

CONCLUSION

Although the particular incident is unknown, locked in syndrome appears to be a sporadic condition in which language disorder is alleged. The findings should trigger a detailed assessment to establish whether it is isolated or if there are any associated problems. This case enlightens the importance of augmentative and alternative communication as a therapy tool. This case study shows the importance of a speech language pathologist in diagnosing and management of this rare language disorder. Augmentative and alternative communication is the best implementation in this case as after months of consistent therapy there is very little progress. AAC is a modified practice area to treat the communication abilities of individuals with little or no functional speech.

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