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Bickerstaff Brainstem Encephalitis Masquerading As Snake Bite: A Case Report

Case Report

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Abstract

Introduction: Bickerstaff brainstem encephalitis (BBE) is a rare immune mediated disease that has good prognosis. It presents with several clinical and immunological similarities with Guillain Barre syndrome and miller fisher syndrome.

Aim: To sensitize the pediatrician to revise the diagnosis of snake bite in absence of improvement with antisnake venom.

Case: We report a case of 5-year-old male child presented with drooping of eyelids followed by difficulty breathing and altered sensorium, provisionally diagnosed and treated as a case of snake bite later diagnosed as Bickerstaff encephalitis clinically and supported by laboratory and radiological investigations.

Results: The child had fully recovered with supportive care and IVIG. Conclusion: The interest in this observation lies in its rarity, presenting symptoms and drastic clinical improvement with immunotherapy. Any case of neurogenic snake bite which did not respond to conventional treatment, we should look for alternative diagnosis.

Keywords: Bickerstaff Brainstem Encephalitis; Snake Bite; GuillianBarre Syndrome Variant

Introduction

Bickerstaff Brainstem Encephalitis first described in 1957 by Bickerstaff et al., is a rare autoimmune encephalitis characterized by an acute brainstem dysfunction occurring few days after an infection or vaccination, characterized by ophthalmoplegia, ataxia and altered sensorium.[1] It presents with several clinical and immunological similarities with Guillain Barre syndrome and miller fisher syndrome. [2] The aim of this work is to acknowledge atypical presentation of GBS and consider BBE as differential diagnosis in case of suspected neurotoxic snake bite with no improvement following ASV.

Case Report

We report the case of 5-year-old male child presented with complaints of drooping of eye lid followed by unable to stand and slurring of speech since last 2 days.There was no history of recent travelling, exposure to toxic materials, or head injury. He was fully vaccinated, and had previously been a healthy child. At local hospital, child received total of 20 vials of antisnake venom (ASV) in 2 divided doses, Atropine, Neostigmine suspecting as a case of neurogenic snake bite, also symptomatic treatment done with Mannitol for features of raised ICP. There in view of low GCS and difficulty breathing child was intubated and referred to our hospital on day 2 of illness. We received child in emergency room with bag and tube ventilation, child was afebrile with bradycardia,hypertension and abnormal breathing pattern. Child was immediately shifted to PICU and stabilized. There is history of flu like illness 15 days before onset of symptoms. Neurological examination showed ophthalmoplegia, brisk deep tendon reflexes with bilateral plantar extensor, rest clinical examination was insignificant. Routine blood investigation revealed

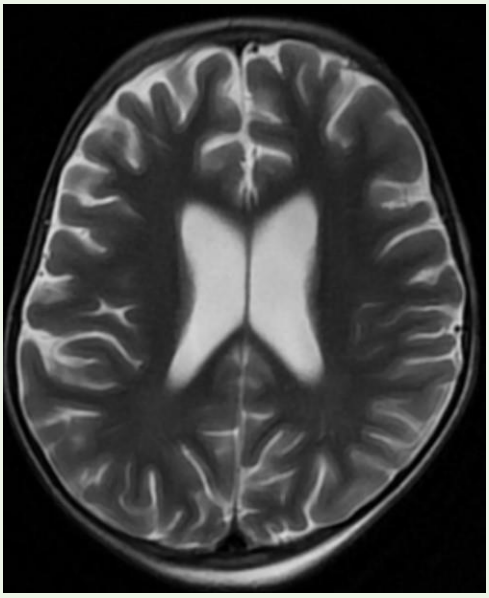

Figure 1: Diffuse cerebral atrophy.

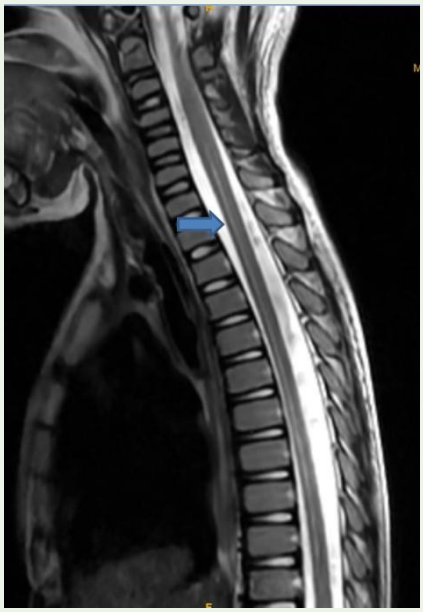

Figure 2: Minimal syrinx with T2 hyperintensity in a long segment of cervico-dorsal cord

hemoglobin of 11.7g/dl, total leukocyte counts 8150/ microliter (60% neutrophil)with negative sepsis screen. Liver function test, renal function test and coagulation parameter are within normal limits.CSF examination showed total 2 cells, glucose 88mg/dl,protein 76.5mg/dl (albumin-cytological dissociation). Presence of albumin cytological dissociation along with ophthalmoplegia, hyperreflexia and altered sensorium compel us to think of BBE. Serum Anti Gq1b antibody was sent and came out to be positive.Nerve conduction study showed axonal type of motor sensory neuropathy.MRI brain and spine showed diffuse cerebral atrophy with minimal syrinx with T2 hyper intensity in a long segment of cervico-dorsal cord.Electroencephalography (EEG) showed high amplitude slow wave activities without epileptiform discharges, suggesting encephalopathy. Child received IvIg on day 4 of illness at 2gm/kg over 5 days. Intubated for 13 days following which tracheostomy was done and on ventilator support for total of 20 days. Gradually regained power in bilateral upper and lower limbs by day 30 of hospitalization and was being discharged. On follow up child is healthy and completely recovered.

Discussion

Bickerstaff brainstem encephalitis is an autoimmune disorder that falls under same spectrum as Miller Fisher syndrome and Guillian Barre syndrome. This is classified as a central nervous system (CNS) disease; whereas, Guillain–Barre syndrome and Miller Fisher syndrome are peripheral nervous system (PNS) disorders [3]. It is similar to Miller Fisher syndrome, a variant of Guillain-Barre syndrome, in that they share features such as ophthalmoplegia and ataxia. The difference is that patients with Bickerstaff's brainstem encephalitis have impaired consciousness & hyperreflexia, whereas patients with Miller Fisher syndrome have alert consciousness and areflexia. Progressive, relatively symmetric external ophthalmoplegia and ataxia by 4 weeks' and 'disturbance of consciousness or hyperreflexia' are required as clinical features for the diagnosis of BBE [4]. The Etiopathogenesis of the disease is still unclear. Infectious etiology could be considered as an antecedent history of upper respiratory tract infection is usually present before the development of the neurological symptoms. [5] Production of gangliosides from some bacteria, similar to those of myelin constituent, may induce a molecular mimicry phenomenon in which the production of specific antibodies (anti-GQ1b). Infection-induced immunological mechanisms may play a pathogenic role in BBE as anti-G1Qb IgG antibody is positive in more than 60% of patients (3), which is positive in our case.Anti-GQ1b antibodies are commonly found in both, but more frequently in Miller Fisher syndrome.[6] Typical MRI finding (hyper intensity on T2-weighted images of the pons, medulla, thalamus, or cerebellum) [6]are absent butpresence of minimal syrinx with T2 hyper intensity in a long segment of cervico-dorsal cord supports ourdiagnosis.According to a previous report, 11% of 47 patients with Bickerstaff's brainstem encephalitis had abnormalities on brain MRI, whereas 57% of 30 patients with Bickerstaff's brainstem encephalitis had abnormalities on EEG[6]. Another study on 37 patients with Bickerstaff's brainstem encephalitis reported abnormalities at 23% and 50% for brain MRI and EEG, respectively[7]. Thus, brain MRI could not detect abnormalities in more than two-third of the patients with Bickerstaff's brainstem encephalitis. As most cases of Bickerstaff's brainstem encephalitis show no abnormal lesions on brain MRI, functional imaging tools such as PET could be useful to document CNS involvement[8].According to the epidemiological study in Japan, similar to our case, 56% of the patients with Bickerstaff's brainstem encephalitis showed less than 5/mm3 of CSF cell count and 20% showed more than 50/mm3 of CSF cell count [7]. In the study by Odaka et. al., most patients with BBE were given immunotherapy, such as steroids, plasmapheresis, and IVIg. [9]Fox et al. have suggested that plasmapheresis and IVIg have a beneficial effect in patients with BBE. [10]. Treatment with either intravenous immunoglobulin or plasma exchange as both have been established as efficacious in improving outcome based on randomised

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control trials in GBS. As seen in our case child showed improvement with IVIG and recovered completely.

Conclusion

Alternative diagnosis should be kept in mind when neurogenic snakebite cases did not showed improvement with conventional treatment (ASV). Complete neurological examinations should be performed in all case of suspected snake bite before giving ASV to ruled out GBS like illness.In any case of acute brainstem dysfunction especially after infectious etiology, BBE should be suspected. MRI brain,CSF analysis and presence of Anti GQ 1 antibody are aid to diagnosis of BBE. Early detection and timely intervention plays pivotal role in achieving remarkable recovery.

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