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Case Report

Guillain-Barré Syndrom Masquerading as Brain Death

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Abstract:

Brain death is irreversible loss of function of the brain including the brain stem. Many clinical conditions may mimic brain death. This is a case review of a 27 year old Bangladeshi male who complaints heaviness of tongue and slurring of speech and was diagnosed as a case of bulbar palsy and found to develop ascending flaccid paralysis of limbs, subsequently he developed respiratory failure was placed on mechanical ventilator (MV) with GCS -3 and brain stem and all spinal reflexes were absent. The MRI & CSF studies were unremarkble. He was on MV and had no spontaneous breathing. A provisional diagnosis of GBS was made with a dfferential diagnosis of Brain stem death. Patient recieved 5 cycles of plasma pheresis without any clinical change. EEG showed cerebral reactivity on tactile stimulation and external noise stimulation. 31^{ist} day of his illness patient developed involuntary tongue movement with pupils weakly reacting to light. 37th day of his illness he opened his eyes with vocal command. His neurological recovery continued till discharge

Introduction:

GBS is a disease of peripheral nervous system, which is caused by aberrant immune response, directed against some components of peripheral nerves[1]. Many variants of the typical syndrome are recognized. An unusual variant, the Fischer syndrome presents with ataxia, areflexia and external opthalmoplegia but weakness often presents later in its course. Other variants are notable for an axonal pattern of electrodiagnostic findings and axonal pathology with little inflammation. In two thirds of cases there are occurrences of gastroenteritis or flu like illness within six weeks of onset of GBS[2-3].The present report describes an unusual presentation of GBS, which initially suggested brain death.

Case Report:

A 27 years old unmarried young man developed heaviness of his tongue and slurring of speech on first day of his illness. He also felt difficulties during opening mouth and swallowing along with drooling of saliva. Then he sends a text massage stating his condition to his mother as he was unable to talk. On the same day he was taken for ENT consultation. He was conscious, alert and ambulant and was able to communicate by writing his problems on paper. On 2nd day of his illness he was admitted into a hospital & was treated as a case of bulbar palsy. But gradually he developed weakness which started from lower limbs, ascending to upper limbs followed by closure of both eyes. On 3rd day of his illness he developed respiratory failure and was intubated. He was grade IV unconsciousness (GCS 3/15) and having flaccid quadriplegia. His pupil was fixed, dilated (9mm) and non reacting to light. Brain stem reflexes were absent (Corneal reflexes were - absent, Occulo-cephalic reflexes test showed no eye movement. Cold caloric test showed- no vestibulo-ocular movement, Gag reflexes were absent), Muscle tone was flaccid in all four limbs, muscle power (MRC 0/60). All deep tendon reflexes were absent bilaterally. Planter response bilaterally were absent, Neck was supple, fundoscopy showed normal fundus. He was mechanically ventilated on Assist/control mode and had no spontaneous respiration and was not on sedation or neuromuscular blocking agent. Apnoea test was not done as because history does not support the case as brain death though patient met the clinical criteria for brain death. He had no recent history of diarrhea, immunization, fever, headache, convulsion or vomiting. There were dysautonomic signs with development of high blood pressure, tachycardia and profuse sweating and constipation and urinary retention and repeated gastroperesis. Provisional diagnosis was Guillain-Barré syndrome, differential diagnosis -Brain stem death, Bickerstaff's brainstem encephalitis. MRI of Brain on 5th day of his illness was normal. CSF study on same day

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revealed normal (colour – clear, RBC 2-4 / HPF, WBC - 4 cell/cmm mostly lymphocyte, Glucose 5 .7mmol/l, Protein 49.2 mg/dl). He was adviced I/V Immunoglobulin which his relatives could not afford. All biochemical investigations were normal. Viral markers (HBsAg , Anti HBc IgM, Anti HCV, CMV (IgM), HSV1 (IgM), HSV2 (IgM) were negative.

Repeat CSF study was done on 19th day of his illness, which showed albumin-cytological dissociation (protein 180 mg/dl, glucose 5 mmol/l, total cell count 05 /cmm). NCV Couldn't be done because the patient could not be transported.

He was treated with 5 cycle of plasma-pheresis with replacement of albumin and saline bedside with CRRT machine started on 13th day of his illness. There was no neurological improvement during therapy except reduction in pupil size after 5th cycle to 4-5 mm which was non reacting to light.

Bed side EEG was done on 20th day of his illness which showed cerebral reactivity on tactile stimulation and external noise stimulation (Fig.-1).

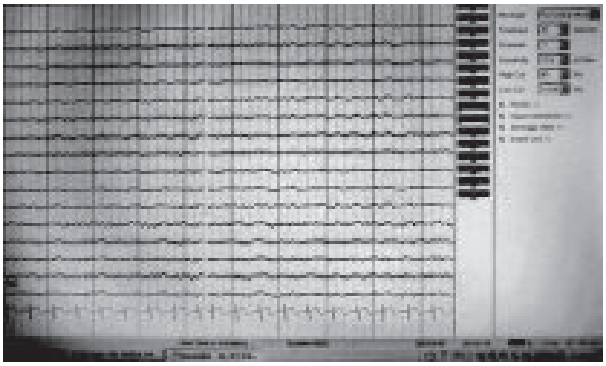

Fig.-1: *Bedside EEG*

Anticipating prolonged ventilator dependency tracheostomy was done on 21th day after admission (19th day of intubation).

Evolution of the neurological findings

Day 1 Bulbar palsy

Day 3 GCS 3/15 (E1V1M1), complete ophthalmoplegia, absence of brainstem reflexes, no spontaneous breathing

Day 19 decrease in size of pupil

Day 31 Involuntary tongue movement and left pupil sluggishly reacted to light

Day 37 Slowly responding to vocal command with movement of head, lower jaw and great toe but there was no eye opening to vocal command

Day 41 He condition was improving and started communicating by moving head and both eyes were open

Day 47 Muscle power improved in both lower and upper limbs (1/5)

Day 57 He was on spontaneous breathing trial requiring pressure support and PEEP

He was on mechanical ventilator for 150 days and tracheostomy was removed after 6 months and 19 days. Total hospital stay 7 months 19 days.

He is wheelchair bound but able to stand and walk with support and can help himself and is receiving regular physiotherapy at present.

Discussion:

The striking feature of this case was the initial clinical impression. In this case report we present a case with bulbar palsy, subsequently rapidly developing flaccid tetraperesis with absent brain reflexes. GBS is a disease of peripheral nervous system. Though features of brainstem dysfunction can be explained by severe peripheral neuropathy involving the cranial nerves. But patient is not unconscious & some sensory perception like vision or hearing maybe functioning which can be detected by auditory & visual evoked potentials. But in our case we only did bedside EEG to evaluate cerebral cortical activity & as there is no bedside evoked potential facilities was not available in our center. Patient may not express his consciousness because of his total motor paralysis.

He was treated like as he is watching and understanding & listening his surroundings. Treating physician instruct all the staff including the family to try to communicate with the patients in every chance. Probably that worked a lot and the patient claimed that he could see & hear the conversation of his surrounding when he seemed nonresponsive.

In our case the differential diagnosis was BBE (Bickerstaff's brainstem encephalitis). Several features argument against the diagnosis of BBE: initial absence of opthalmoplegia, early preservation of consciousness, signs of dysautonomic failure and finally absence of brainstem lesion upon brain MRI[4]. In the review of 13 similar cases, the suspicion of brain death diagnosis occurred on day 2 in most of the cases.[5] In our case sign of brain death notified on third day of illness.

Various EEG patterns can be recorded in these patients clearly allowing to rule out brain death. The most common pattern was the presence of an alpha rhythm, unresponsive to painful and auditory stimulation.[6,7] In other cases, the EEG showed preserved sleep patterns or reactivity to sound.

He had no spontaneous breathing and we did not perform an apnoea testing, one of the prerequisites for the diagnosis of brain death. Brain death is the current medical definition of death when the other body organs continue to function and is defined as an irreversible cessation of all functions of the entire brain, including the brainstem. Before the declaration of brain death, an etiology must be identified that could explain the clinical picture and all reversible causes must be excluded. GBS mimicking brain death is very rare [8, 9 10, 11,13,14]. To add to the difficulty in the initial diagnosis, our patient presented with non-reactive mydriasis. Pupillary abnormalities have rarely been described[12].

The outcome of patients with fulminant GBS who appeared as brain dead is reported in litareture. Three patients died, two from cardiac arrest related to dysautonomia and one from a massive anterior myocardial infarction. Two patients showed poor recovery, with permanent disabling weakness. The others could walk with persistent minor problems. GBS mimicking brain death has a poor recovery rate and a high mortality, particularly in relation to dysautonomia. It is a fact that nerve inexcitability on EMG, the need for ventilatory support for more than 1 month and severe rapidly progressive disease can all lead to residual weakness [15].

There are four common sub types based on clinical and neuro-physiological studies e.g. 1. Acute inflammatory demyelinating polyneuropathy (AIDP), 2. Acute motor axonal neuropathy (AMAN), 3. Acute motor sensory axonal neuropathy (AMSAN), 4. Millar fisher's syndrome[16]. In Bangladesh large proportion of children with acute flaccid paralysis have GBS & 56% of them are AMAN variety.[17]

Conclusion:

This case presented as an extreme polyneuropathy to the point of being able to masquerade as brain death. This was a rare variant of GBS with distinct case in the sense that it is a reversible condition and misdiagnosis without a good antecedent history is potentially easy & carries disastrous consequence[18] & EEG may helps in evaluating brain function.

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