A CASE OF BICKERSTAFF'S BRAINSTEM ENCEPHALITIS IN CHILDHOOD

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ABSTRACT

Bickerstaff's brainstem encephalitis is characterized by acute progressive ophthalmoplegia, ataxia, and disturbance of consciousness. It is similar to Miller Fisher syndrome, a variant of Guillain-Barre syndrome, because they have some features in common like; ophthalmoplegia and ataxia. The difference is that patients with Bickerstaff's brainstem encephalitis have impaired consciousness and hyperreflexia while patients with Miller Fisher syndrome have alert consciousness and areflexia.

Here, we present a case 4 year and 3 month old girl with abrupt onset ophthalmoplegia, ataxia and disturbed consciousness, her brain Magnetic Resonance Imaging (MRI) was normal, cerebrospinal fluid analysis showed albuminocytological dissociation, and Nerve conduction study is suggestive of Acute Inflammatory Demyelinating-axonal motor polyneuropathy (AIDP). She has been treated successfully with steroid with complete recovery within two months.

Keywords: *Encephalitis, Bickerstaff's, brain stem, complete recovery*

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INTRODUCTION

A variant of Guillain-Barre Syndrome, Bickerstaff Encephalopathy was first described in 1951 by Edwin Bickerstaff who reported three patients with the triad of ataxia, ophthalmoplegia, and brisk tendon reflexes. Unlike the previous reported Miller-Fisher variant, these cases described disturbance of consciousness and hyper-reflexia as defining characteristics of the clinical course [1, 2].

Bickerstaff's brain stem encephalitis (BBE) is characterized by the acute onset of external ophthalmoplegia, ataxia, altered consciousness, and hyperreflexia after the exclusion for other diseases involving the brain stem In addition, the neurological signs that suggest this abnormality of the central nervous system presents with: abnormalities of the pupils, facial weakness, bulbar palsy, dysesthesia, limb weakness, positive Babinski sign, nystagmus, and blepharotosis [1-4].

BBE shows clinical overlap with Miller-Fisher syndrome a cranial nerve variant of Guillain-Barré syndrome as well as with the axonal forms of GBS (acute motor axonal neuropathy and acute motor-sensory axonal neuropathy) in patients with limb weakness, leading several authors to suggest that BBE, MFS and GBS represent variable manifestations of the same clinical spectrum. The differential diagnosis in patients with BBE should also include acute disseminated encephalomyelitis as well as rare post-infectious neurological disorders [5].

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Since Bickerstaff's initial paper, case reports confirm overlapping clinical symptoms of the two variants. Debates continue on whether these two entities may represent a single variant of GBS. Like Guillain-Barre Syndrome, Miller-Fisher and Bickerstaff Encephalopathy are frequently precipitated by an infectious etiology, often viral, that produces cross-reacting antibodies to nerve antigens [6].

There have been several reports of BBE associated with antiganglioside antibodies. Anti-GQ1b antibodies are frequently detected in as much as among 66% of patients with BBE [2-7]. Other antibodies that may be positive in patients with BBE are: anti-GM1, anti GD1a, and anti-Ga1NAc-Gd1a [8].

CASE REPORT

Four year and three month old girl (figure 1) presented to ER of Sulaimani Pediatrics Teaching Hospital with disturbed level of consciousness for 10 hours before admission . The condition started 3 days before admission with intermittent fever. On the fourth day of the illness the child was drowsy, ataxic and developed seizure followed by loss of consciousness. There were no associated symptoms such as diarrhea, vomiting, cough, coryza or sore throat.

On initial examination the child had disturbed level of consciousness, pupils had normal reaction to light, Glascow coma scale was 8. The muscle strength was reduced in both upper and lower limbs and was grade 2. There was spasticity on initial passive movement of all four limbs. Sensation was intact. Neck rigidity was positive in all directions, positive Kernig's sign and up going planter response. Deep tendon reflexes were grade 3 without clonus. She also had bilateral abducens nerve palsy and left side facial nerve palsy.

On admission, the vital signs were body temperature of 37 C, pulse rate of 110 beats/min, respiratory rate 30 cpm, blood pressure of 110/80 mmHg, oxygen saturation 96% in room air.

Routine tests on admission were done for her including: complete blood count, ESR, blood chemistry (including plasma glucose, blood urea, electrolytes), C-reactive protein, and urine analysis. CT scan of brain; all were normal, after supportive measure we did lumbar puncture on second day of admission. Cerebrospinal fluid (CSF) study revealed a WBC count of zero cells/μL, a protein level of 97 mg/dL, and a glucose level of 54 mg/dL. Cerebrospinal fluid (CSF) study repeated on day 7 revealed a WBC count of zero cells/μL, a protein level of 192 mg/dL, and a glucose level of 72 mg/dL.

The brain Magnetic resonance imaging (figure 2) and Electroencephalogram done on 7th day showed no abnormalities (figure 3). Nerve conduction study done on 10th days of admission, was suggestive of acute inflammatory demyelinating-axonal motor polyneuropathy (predominantly a motor type of AIDP-GBS) (figure 4). The patient treated empirically with steroid IV (for the first 4 days), acyclovir and mannitol.

On 4th day of hospitalization she regained consciousness. The muscle weakness, spasticity, extraocular nerve palsies and ataxic gate had gradually improved within the next 2 weeks of admission. The patient discharged and was followed in outpatient clinic, all symptoms completely resolved by two month after discharge.

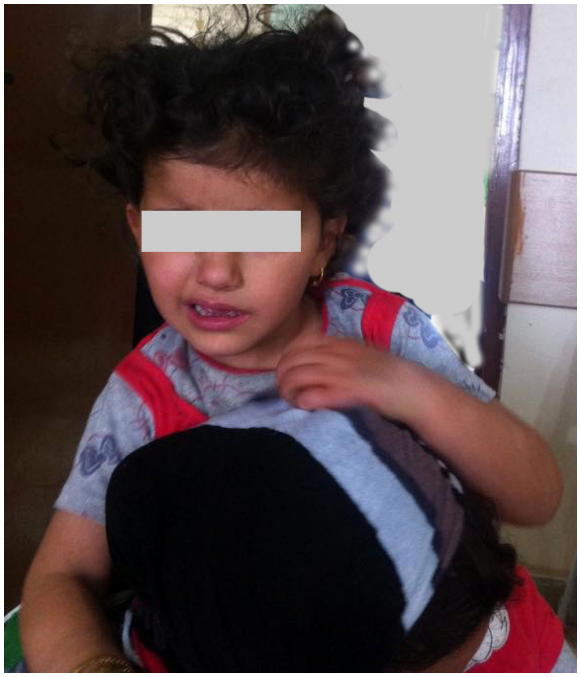

Figure 1 . Picture of case BBE.

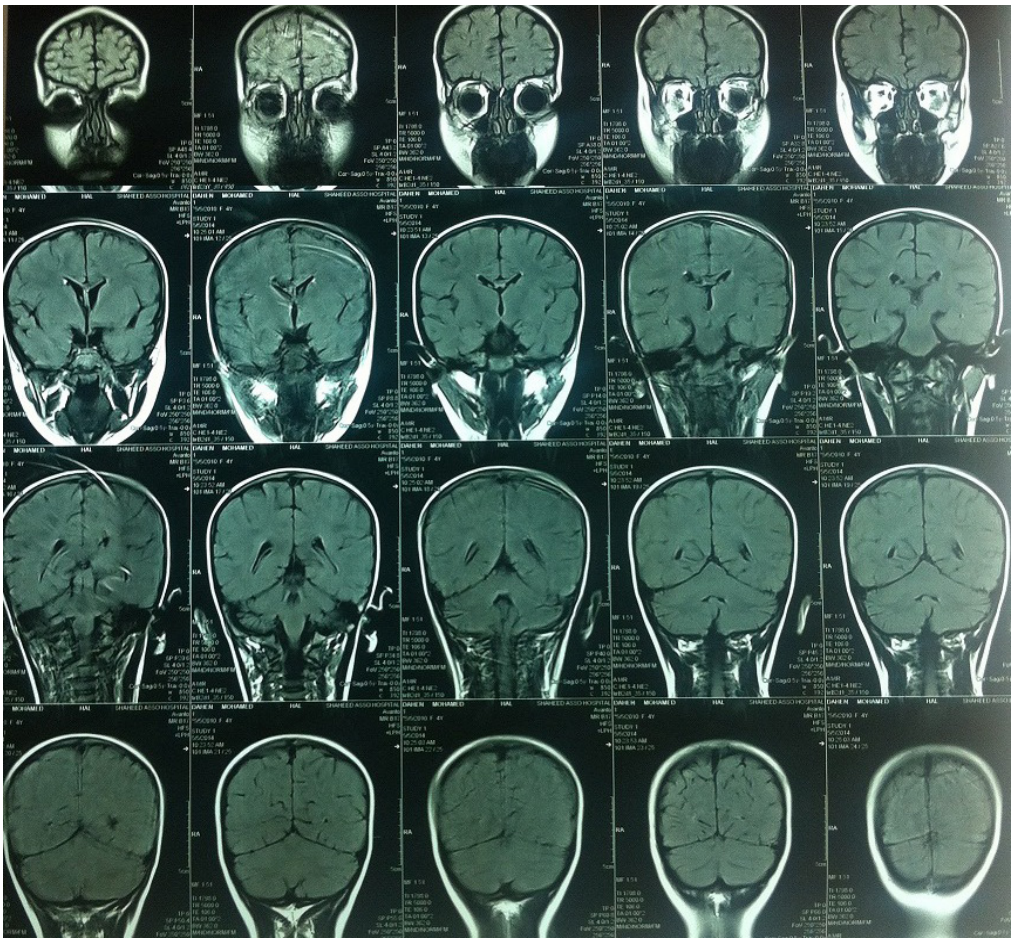

Figure 2. Brain Magnetic resonance imaging of the case.

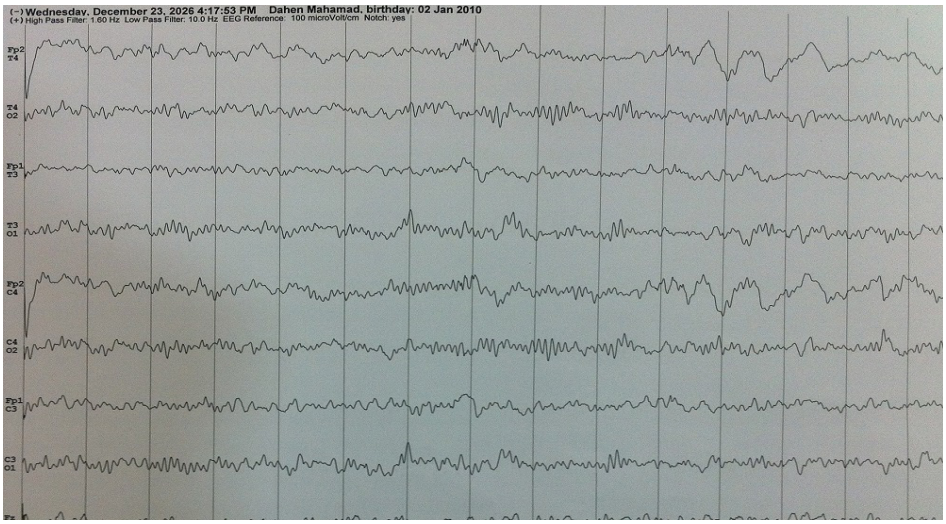

Figure 3. Electroencephalogram of the case.

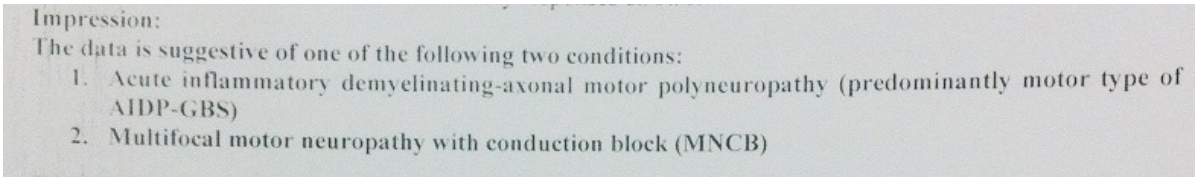
Impression:
The data is suggestive of one of the following two conditions:
1. Acute inflammatory demyelinating-axonal motor polyneuropathy (predominantly motor type of AIDP-GBS)
2. Multifocal motor neuropathy with conduction block (MNCB)

Figure 4. The final report of NCT by neurophysiologist.

DISCUSSION

Bickerstaff and Cloake reported three cases of drowsiness, ophthalmoplegia and ataxia in 1951 [3]. The diagnostic criteria for BBE include: 1- progressive, relatively symmetric ophthalmoplegia and ataxia by four weeks; 2-either altered consciousness (coma, semicoma or stupor) or pyramidal signs (hyperreflexia or pathological reflexes), and 3-limb strength of 5 or 4 on the Medical Research Council scale [9]. In all cases the course of illness and anatomical structure involved were similar with gradual improvement to eventual complete recovery.

In our case, initial presentation with ataxia and ophthalmoplegia was suggestive of Miller Fisher syndrome but disturbance of consciousness, seizure and hyperreflexia were in favour of Bickerstaff's brainstem encephalitis rather than Miller Fisher syndrome. It is difficult to differentiate between the two as they share common features. Anti-GQ1b antibodies are commonly found in both, but more frequently in Miller Fisher syndrome [2, 7]. According to the Japanese epidemiological study, Bickerstaff's brainstem encephalitis accounted for 43% of brainstem encephalitis and anti-GQ1b antibodies were present in 75% of the patients with Bickerstaff'sbrainstem encephalitis [2]. Unfortunately in our case we didn't have facilities to measure antiganglioside antibodies.

CSF albuminocytological dissociation is present in GBS, FS and Bickerstaff brainstem encephalitis. Two of the eight patients described in the original paper of Bickerstaff had CSF albuminocytological dissociation and the other six had CSF pleocytosis (median, 26 cells/ml; range, 10±153 cells/ml). Odaka M reported that CSF protein concentration was normal in about two-thirds of BBE patients during the first week, but its frequency increased during subsequent weeks. CSF albuminocytological dissociation was found 35% during the first 4 weeks, and the frequency increased from 19% in the first week to 57% in the third and fourth weeks. In our case CSF protein in first week was 97 mg/dl and increased to 192 mg/dl in the second week and there were no pleocytosis.

Normal MRIs have been reported for some BBE patients, whereas in others high-signal lesions, documented on T2-weighted images of the upper mesencephalon, cerebellum, thalamus or brainstem, may move and regress with the clinical course of the illness [10, 11]. In our case MRI finding was normal, Brain imaging is rarely useful for the diagnosis of BBE, but its essential for exclusion of intracranial pathology.

During the first week of illness, CSF albumin-cytological dissociation was reported in 25% of patients and increase with time in BBE like our case and other [11].

The EEG is often abnormal but shows only slow wave activity, which also occurs in many other conditions, and is therefore of limited value in diagnosis [2]. Nerve conduction studies may show an axonal polyneuropathy. In Japanese study (38%) patients showed abnormal conduction in peripheral nerves, including decrease compound muscle action potential amplitude or reduced F wave frequencies or both [5]. In our case NCT was suggestive of acute inflammatory demyelinating-axonal motor polyneuropathy.

In conclusion: we report for first time in our country a rare childhood case of BBE with a good clinical outcome after successful treatment with I.V. steroid. The diagnosis of BBE in our case was supported by the specific clinical symptoms (e.g. cranial polyneuropathy presented as ophthalmoplegia, dysarthria, dysphagia, and facial weakness; progressive motor weakness; disturbed mental status, seizure; and hyperreflexia) and the presence of cytoalbuminodissosciation in CSF.

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