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A Case Report On Bicker Staff Brainstem encephalitis

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ABSTRACT
Bickerstaff brainstem encephalitis (BBE) is a rare inflammatory and demyelinating disease, with an estimated annual incidence of 0.078 per 100 000[2]. This case was presented with symptoms such as high grade fever spikes, altered sensorium, irritability, vomiting, abnormal movements, myoclonic jerks, Rhabdomyolysis, intentional tremor, ataxia and DTR weakness.MRI was reported to be brainstem encephalitis-Bickerstaff. Here we report a case of a male pediatric patient with clinical presentation of Bicker staff brainstem encephalitis.

KEYWORDS: Bickerstaff brainstem encephalitis, Autoimmune encephalitis, Encephalitis.

I. INTRODUCTION

Bickerstaff brainstem encephalitis (BBE) is characterized by progressive bilateral ophthalmoparesis and ataxia with loss of consciousness or pyramidal signs[1]. In addition to this feature, hyperreflexia, limb weakness, sensory changes, and bulbar and facial paralysis have been reported. It is part of the "anti-GQ1b syndrome", which forms a common serological profile supporting both Miller-Fisher syndrome (MFS) and BBE[2].Serum anti-GQ1b IgG antibodies (Abs) are observed with varying frequency in patients with BBE and are a common feature of Guillain-Barré syndrome (GBS) and Miller Fisher syndrome (MFS)[3]. It is generally believed that BBE is not a distinct neurological entity but lies at the other end of a spectrum of disorders known as anti-GQ1b syndrome. A previous infectious disease has been observed in the majority of cases of BBE [4,5] and is considered as a possible trigger for the autoimmune response. Bickerstaff encephalitis (BBE) is rare, with an estimated annual incidence of 0.078 per 100,000[6].

II. CASE PRESENTATION

A3yearold developmentally normal child admitted in pediatric ICU and developed with high grade fever spikes (106F) for 3 days, altered sensorium for 3 days, irritability, vomiting, abnormal movements and myoclonic jerks. Rhabdomyolysis, intentional tremor, ataxia and DTR weakness were developed in the child indicating an overlap with Bickerstaff brain stem encephalitis. The child had sudden jerky abnormal movements involving the limbs and eyes initially which gradually reduced infrequency.

Child had E1V1M4 response with preserved brainstem reflexes. Possibility of acute CNS infection/ demyelinating event was considered. History given by parents was not suggestive of rabies. As child had capillary leak features, fever. tachycardia, hyperglycemia at admission suggestive of hospital acquired sepsis/tropical infection antibiotic was upgraded to Meropenem and doxycycline. Acyclovir was continued. He was reintubated with 4.5cuffed tube to prevent aspiration. No papilledema was noted. Lumbar puncture was done which showed lymphocyticpleocytosis with mild protein elevation.

Many of the laboratory parameters of the child are abnormal which are Urea- 49 md/dl,creatinine-0.9 mg/dl sodium- 156 mmol/L, procalciton in- 115, CPK- 22370, high sensitivetroponin-373,SGOT-721,SGPT-577,phosphorus-5.3mg/dl

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MRI was reported to be brainstem encephalitis-Bickerstaff. MRI study showed ill definedhyper intense signals in the brain stem, cerebellar peduncles show hyperintense signal and diffuse hyperintensity in cerebellum in DWI with mild restriction in ADC map, suggesting Bicker staff brain stem encephalitis.

Hence, after discussing with relatives about possible benefit of IVIG, 2g/kg IVIG was given as 3 aliquots over the next 3 days. No seizures were noted during the course and IV Phenytoin was continued. Sensorium gradually improved and child was extubated after 4 days of ventilation (Total 6 days). As his muscle power was weak and he was supported with HFNC post extubation. He had sudden jerky abnormal movements involving the limbs and eyes initially which gradually reduced in frequency. Soft feeds were tried and full oral feeds were started. Repeat MRI was done after 7 days which showed resolving lesions Child is able to stand with support and communicates well with parents now.

Child had capillary leak, hyperthermia, multi organ dysfunction syndrome, hyperglycemia atadmissionsuggestingpossibilityofsecondarysepsis Procalcitoninwas>200.Meropenemwasstartedaftertaking cultures.Hehadthickseropurulentsecretionsfrom ETtubewithsignificantbronchospasm affecting ventilation on day 1 of PICU stay. Airway problems improved afterday 2ET aspirate was reported to grow Klebsiella Pneumoniae. Nobacteria was isolated from blood stream/ urine. Possibility of viral/ leptospiral sepsisis considered.

Possibility of refeeding syndrome was considered and IV thiamine 100 mg was started and feeds were reduced and hiked gradually over next 5 days. Potassium and phosphate were corrected withIV potassium phosphate, Oral sodium phosphate and oral potassium chloride for the next 3days.

Patient had rhabdomyolysis which indicated by CPK was in 15000 U/L range at admission andwentcloseto40000.Urinewasdarkcolouredonday 1howeverurinemyoglobinwasreportedto be negative. Child was managed with 2 times the maintenance fluid requirement. He had reduced urine output with good fluid status, that responded to2 furosemidebol uses.

During the hospital days the patient was treated with medications such as Inj. Thiamine 30mgIVOD, Multivitamin syrup5mlBD,Inj. Ondansetron2mgIVTDS,IV Immunoglobulin2Vial(5mg/100ml),Inj. Meropenem 600mg IV TID 31 Doses, Inj. Dexamethasone 2.2mg IV, Neb with Adrenaline5ml Q4H, Neb. Levosal but amol +2ml NS +$O_2$2ml Q4H, Inj. Acyclovir 225mg IV Q12H 5Doses, Tab. Doxycycline 65mg OD, Inj. Phenytoin 30mg IV Q12H, Inj. Pantoprazole 15mgI VOD, Inj.VitK5mg OD 3Doses, Syrup. Ibugesicplus 5.5ml SOS ,Lacrinagel BD, Tear Drops Q4H, Tab. Pyridoxine 200mg OD, Addphos sachet BD, Syp. Ambroxol 5ml TID, Dermadewcaloe lotion LIA, Inj. Levetiracetam 300mg BD 11 Doses, Inj. Ceftriaxone 750mg IV BD 9Doses, T Bact ointment, Fucidine Cream BD and other supportive measures. In subsequentdays, the child's condition was improved but patienthad Nystagmus.

III. DISCUSSION

Edwin Bickerstaff4 described BBE in the 1950s as "a serious syndrome with a benign prognosis". He reported a syndrome of ophthalmoplegia, ataxia and somnolence preceded by infection [10,11], There were parallels with MFS and Guillain-Barré syndrome (GBS), including areflexia and increased protein in the CSF. Bickerstaff encephalitis is an acute demyelinating pathology that affects the brainstem and develops a few days after an infectious episode. Although the exact pathogenesis remains unclear, it is thought to be related to an immune response triggered by previous infection with pathogens such as Campylobacter jejuni, Mycoplasma pneumonia, or Haemophilus influenza [7]. Guillain–Barré syndrome (GBS), Miller–Fisher syndrome, and BBE share some similarities, including the presence of antiganglioside antibodies. Together with GBS and Miller-Fisher syndrome, these three syndromes form a group of post-infectious demyelinating diseases[7]. There is no specific biological marker for BBE and diagnosis is based on a combination of anamnestic, clinical and radiological features [8,9]. Lumbar puncture is systematically performed to rule out infectious meningitis requiring specific antibiotic therapy. CSF analysis shows lymphocytic pleocytosis in 40% of cases, hyper proteinuria in 59%, and albumin cytological dissociation in 19% of cases [9].

IV. CONCLUSION

BBE is characterized by progressive bilateral ophthalmoparesis and ataxia with loss of consciousness and pyramidal signs[1]. Diagnostic criteria for BBE are ophthalmoplegia and ataxia with loss of consciousness and/or pyramidal

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signs.The patient was administered intravenous immunoglobulins and then subjected to plasmapheresis. Some cases of BBE may resolve on their own and require intravenous immunoglobulin or plasma exchange. Anti-CD20 monoclonal antibodies in the form of rituximab have been used in resistant cases[12].Early diagnosis and appropriate treatment reduce morbidity and mortality.

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