

CASE REPORTS

BICKERSTAFF BRAIN-STEM ENCEPHALITIS

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Abstract

Bickerstaff's Brainstem encephalitis is a very rare localized central nervous system inflammatory demyelinating disorder. This has not been reported in children in India before. We here describe a 12 year old girl with Bickerstaff's brainstem encephalitis who had multiple cranial nerve palsies. The etiologies, clinical course and therapeutic interventions of this rare condition are briefly reviewed.

Key words: Bickerstaff's Brainstem encephalitis, demyelinating disorder, cranial nerve palsies.

Introduction

Acute demyelinating encephalomyelitis is an immune-mediated disorder localized to the central nervous system (CNS). The demyelinating lesion can be localized (referred to as clinically isolated syndrome or CIS) or disseminated (then called as acute disseminated encephalomyelitis, or ADEM).(1) When localized to the brainstem, it is referred to as Bickerstaff's brainstem encephalitis (BBE). The disease is characterized by the acute onset of signs of brainstem involvement, days to weeks after a variety of viral and bacterial infections or vaccinations. Because of its rarity, there has been no report on BBE patients in pediatric age group in India. We here report the first case of BBE in a girl, who presented with multiple cranial nerve palsies.

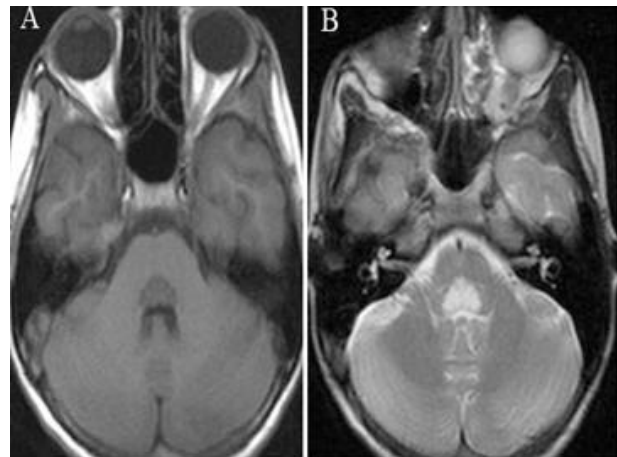
Case Report

A 12-year-old previously healthy girl developed headache and vomiting, 2 days before admission to our hospital. There was no associated fever, cough or nasal discharge. Headache was not associated with diplopia, altered sensorium, or neck stiffness. On examination, she was fully conscious and her general examination was unremarkable. The neurologic examination showed bilateral facial nerve paralysis (right more than left). She had an expression-less face and horizontal gaze paralysis. She could not converge her eyes also. Vertical eye movements were normal. Over a period of one-week, she developed difficulty closing her jaw, chewing and nasal regurgitation while feeding. She had moderate dysmetria on finger-to-nose testing. Her gait became unsteady, and wide-based but Romberg sign was negative. Deep tendon jerks were normal initially and later were not elicitable. Plantar response was flexor. A possibility of Fisher syndrome (FS) or BBE was considered.

Her investigations revealed that the blood-counts and blood-chemistry results were normal. Cerebrospinal fluid (CSF) analysis showed 3 white blood cells/mm³ (all lymphocytes), with a protein of 25 mg/dL and glucose of 83 mg/dL. CSF culture was negative. Her nerve conduction studies done on the 10th day of illness were normal and blood was negative for anti-nuclear antibodies. Magnetic resonance imaging (MRI) brain was also normal. In view of progression of symptoms,

MRI was repeated after one week. This revealed a small midline pontine lesion, 11 mm in diameter, adjacent to the floor of fourth ventricle that showed low signal on T1 and high signal on T2-weighted images (Figure-1). This MRI finding was consistent with a demyelinating process and suggested a possibility of brainstem encephalitis. In view of low sensitivity and specificity of antibodies like anti GQ1b in differentiating MFS from BBE, serological tests were not done. The patient received a 5-day course of IVIG (total of 2.0gm/kg). As there was not much improvement and the bulbar palsy worsened, 5 pulses of i.v. methylprednisolone of 25mg/kg/day were given starting on the 4th day of IVIG. On the 5th day, there was marked improvement in facial tone and paresis gradually resolved by the end of therapy. Even the cerebellar signs, bulbar signs and ocular movements steadily improved. At discharge, she just had mild difficulty with tandem gait. Clinical and radiological follow-up after 6 months was normal.

Figure 1: MRI of brain. A: Axial T1W image shows a lesion involving pontine tegmentum with hypointense signals. B: The same lesion is hyperintense in T2W sequence.



Discussion

BBE which is one of CNS inflammatory demyelinating disorders is described as a type of Clinically Isolated Syndrome (CIS) in the consensus definitions proposed by the International Pediatric Multiple Sclerosis Study Group. It is the only CIS that can be associated with encephalopathy.(1)

Bickerstaff reviewed the syndrome for "The Handbook of Clinical Neurology" under the title "brainstem encephalitis" in 1978.(2) The diagnostic criteria for BBE were: (1) progressive, relatively symmetric ophthalmoplegia and ataxia by 4 weeks; (2) either consciousness disturbance (coma, semicoma, or stupor) or pyramidal signs (hyperreflexia or pathological reflexes); (3) limb strength of 5 or 4 on the Medical Research Council scale. Patients showing limb weakness (3 or less on the Medical Research

Council scale) which is not uncommon are diagnosed as having overlapping BBE and Guillain-Barre syndrome (GBS).(3)

The clinical symptoms of BBE are similar to FS, but BBE must have consciousness disturbance or pyramidal signs/long tract signs, which reflect serious brainstem lesion.(4) Upward gaze and horizontal gaze disability, pupillary abnormalities and nystagmus are frequent ocular manifestations. Other clinical features of BBE are facial weakness, blepharoptosis, bulbar palsy and long-tract sensory disturbance.(5) The etiology of BBE is similar to that of FS and GBS. Abnormal lesions (high-intensity areas on T2-weighted images of the brainstem, thalamus, cerebellum and cerebrum) on MRI findings are present in about one-third of BBE patients (more common when compared to FS). The relationship of BBE to FS and GBS remains controversial. It is suspected that BBE, FS and GBS form a continuous spectrum; whether BBE is a distinct disease entity or a variant of FS and GBS is still to be clarified.(6) Many authors even consider FS and BBE as the same disease. Our case had ophthalmoplegia and ataxia but with intact sensorium, flexor plantar and normal deep tendon reflexes. The initial MRI was normal but repeat MRI showed a demyelinating lesion. Like in ADEM, there could be a gap of 5-14 days before the MRI changes appear.(7) Thus our case also highlights the overlap between BBE and FS. While the brainstem lesion was favoring the diagnosis of BBE the absence of altered sensorium and pyramidal signs favored the diagnosis of FS. BBE has a monophasic remitting course, with 2/3rd of patients becoming symptom-free at six months and around 16% patients requiring ventilation. Deaths have been reported in BBE.(8) Thus patients with BBE need careful observation and in cases that worsen or have respiratory involvement, immunomodulatory therapy to prevent residual deficit and death needs to be started.

There are no randomized controlled trials of immunomodulatory therapy in FS or related disorders (BBE) on which to base practice.(8) Until recently, corticosteroids were the only widely used treatment for acute demyelinating encephalitis.(9) However, there is disagreement regarding their efficacy and there have not been any prospective studies or treatment guidelines. Several reports suggested that plasmapheresis and IVIG have a beneficial effect on patients with BBE. Moreover, combined therapy of IVIG and high-dose methylprednisolone should be more efficacious therapy. No advantage of any particular treatment can be inferred from the data

available.(8) Controlled clinical trials are needed to test this proposal.

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E-published: 1st October 2011. **Art#64**
