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CASE REPORT Open Access

Bickerstaff brainstem encephalitis, an uncommon presentation in a child: a case report

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Abstract

Background The typical clinical and radiological presentation of Bickerstaff brainstem encephalitis (BBE) has been highlighted in this case report.

Case presentation Bickerstaff encephalitis is a rare autoimmune inflammatory disorder and is considered a subtype of Guillain–Barré syndrome (GBS) along with Miller Fisher syndrome. The diagnosis of BBE is largely clinical, though laboratory tests and imaging can be of supportive value. We report a case of a 5-year-old child who presented with a classical clinical triad of BBE with characteristic magnetic resonance imaging (MRI) findings.

Conclusions BBE is a rare disease with very few cases being reported with typical clinical and radiological findings. Hence, we reported a typical case of BBE to make an addition to the available literature.

Keywords Radiological, Bickerstaff encephalitis, MRI

Background

Bickerstaff brainstem encephalitis (BBE) is a rare autoimmune inflammatory disorder that involves the central nervous system (CNS) (1). It is considered a subtype of Guillain–Barré syndrome (GBS) along with Miller Fisher syndrome. The patients with BBE clinically present with the triad of altered consciousness, ophthalmoplegia, and ataxia (2). The antecedent history of infection is usually present. As BBE is rare in the pediatric population with only a few cases reported in the past, we present a case of Bickerstaff encephalitis in a 5-year-old boy presenting with the classical clinical triad and typical MR imaging findings.

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Case presentation

A 5-year-old male firstborn child presented to the neurology outpatient department (OPD) in January 2023, with chief complaints of insidious onset, gradually progressive drooping of the eyelids (Right > Left) with a downward deviation of the left eye for 7 days. Parents also complained of the hypersomnolence of the child and abnormal gait. History was significant for viral fever 20 days back. Neurological examination showed brisk deep tendon reflexes and the right plantar reflex showed extension. The rest of the clinical examination was unremarkable. Blood investigations revealed a hemoglobin of 12.6 gm/dl and a total leucocyte count of 7900 cells/ul. CSF examination was significant for lymphocytosis (> 90%), sugar-59 mg/dl (normal range 45–80 mg/dl), and protein 23.7 md/dl (normal range 15–45 mg/dl). No viral or bacterial pathogen was isolated from blood or CSF samples. GQ1B antibodies in the CSF were negative.

Magnetic resonance imaging (MRI) brain was done on the next day of presentation (8^{th} day of illness) using a Philips Ingenia 3.0T MR scanner, which included T1WI, T2WI, DWI, SWI, FLAIR and post-contrast

T1FS images in axial, coronal and sagittal planes. Images were reviewed by two radiologists with 4 and 15 years of experience each. It showed confluent areas of T2/FLAIR hyperintensities in the midbrain (sparing red nucleus) and dorsal pons (Fig. 1) showing an isointense signal on T1WI. There was no diffusion restriction, blooming, or post-contrast enhancement. No other abnormality was identified on the MRI. Thus the diagnosis of the BBE was made based on the typical clinical and imaging findings (Figs. 2 and 3).

The patient was treated with intravenous IV steroids and showed significant clinical improvement. However, a follow-up MRI could not be performed to evaluate the interval change in radiological findings.

Discussion

Bickerstaff brainstem encephalitis is an autoimmune disorder that falls under the same spectrum as Miller Fisher syndrome and Guillain–Barre syndrome (2). These are postinfectious disorders that share common clinical traits such as ataxia and ophthalmoplegia. Additionally, patients frequently exhibit prodromal upper respiratory infection, cerebrospinal fluid (CSF) albuminocytological dissociation, and serum IgG antibody to ganglioside GQ1b3, all of which are indicators of a similar origin

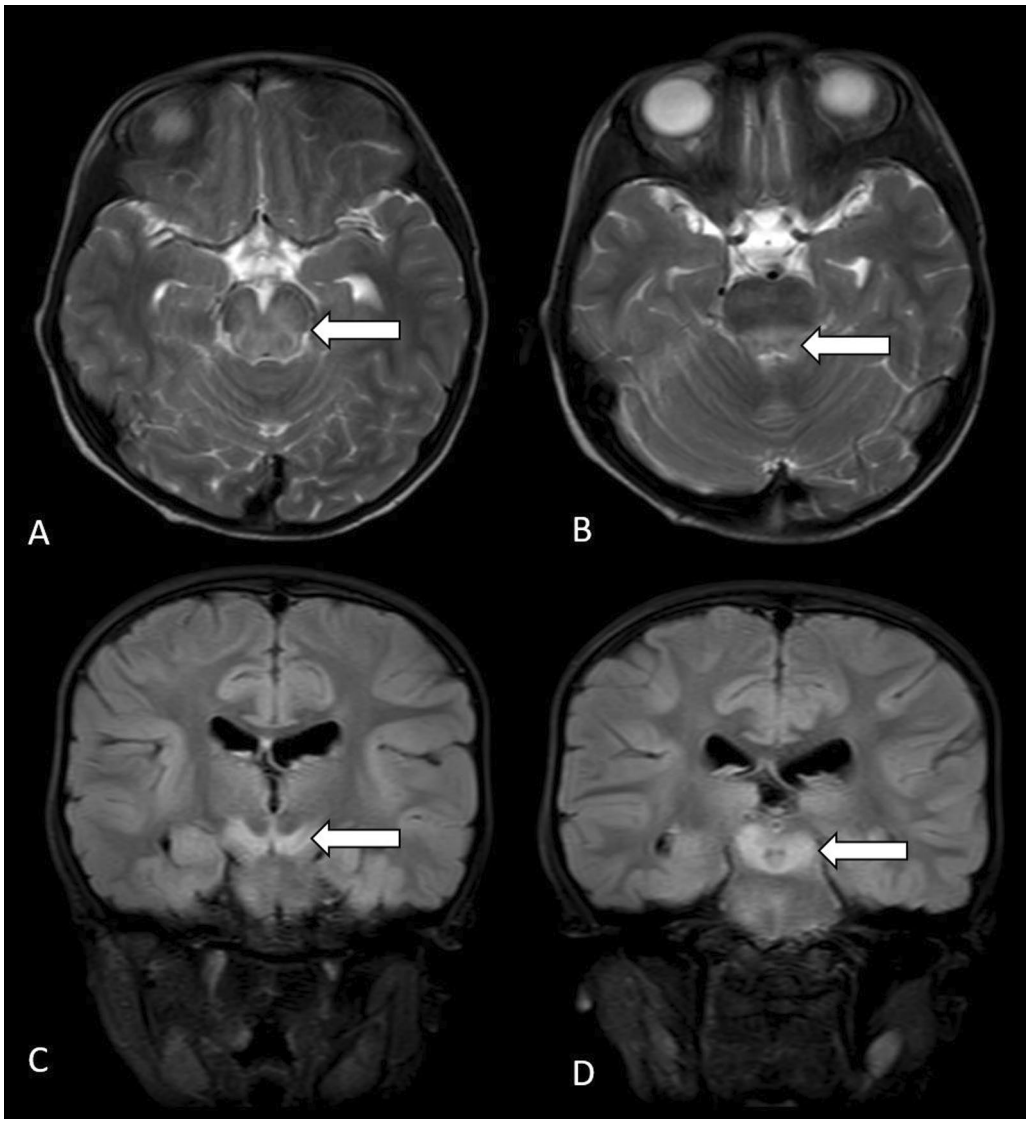

Fig. 1 T2 axial images (**A** and **B**) and FLAIR coronal images (**C** and **D**) of the MRI brain show T2/FLAIR hyperintensity involving the midbrain (sparing red nucleus) and dorsal pons

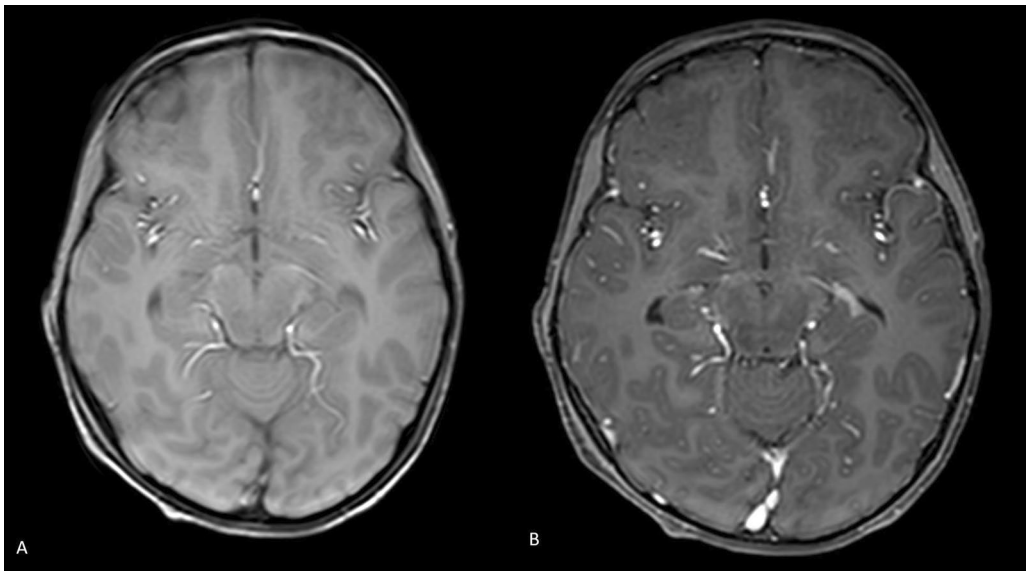

Fig. 2 T1 FFE (**A**) and post-contrast T1FS (**B**) axial images of the MRI brain show an isointense signal in the midbrain with no post-contrast enhancement

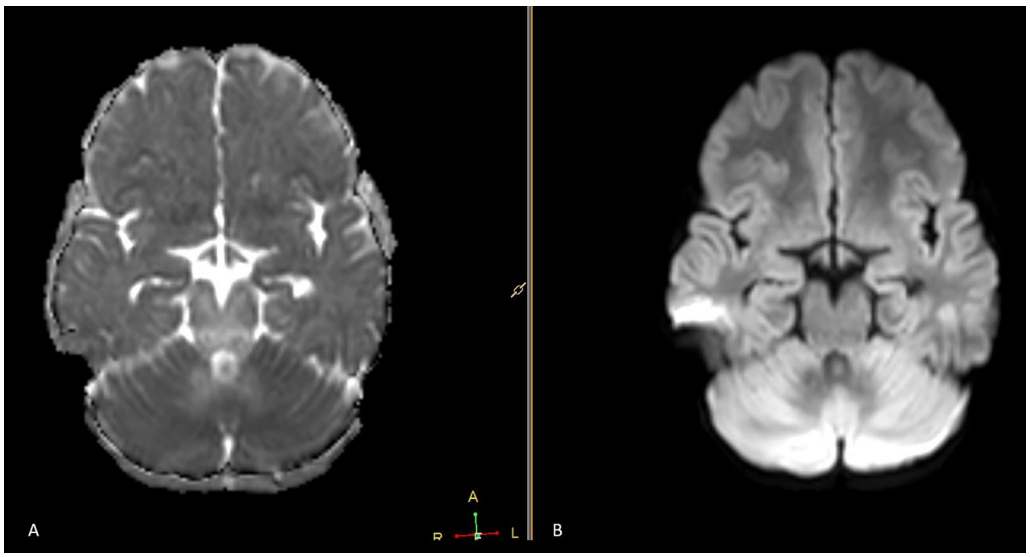

Fig. 3 ADC (**A**) and DWI (**B**) images of the MRI brain show no area of diffusion restriction in the midbrain

(3). This is classified as a central nervous system (CNS) disease; whereas, Guillain–Barre syndrome and Miller Fisher syndrome are peripheral nervous system (PNS) disorders (2).

'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). Additionally, Wernicke's encephalopathy, botulism, myasthenia gravis, brain stem tumors, pituitary apoplexy, acute disseminated encephalomyelitis, multiple sclerosis, neuro-disease, Behcet's vasculitis, lymphoma, and Creutzfeldt–Jakob disease must be ruled out before diagnosing the patient as BBE (5).

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. Another possibility is that infection-induced immunological mechanisms may play a pathogenic role

in BBE as anti-G1Qb IgG antibody is positive in more than 60% of patients (2). The CSF Anti-Gq1B antibodies were absent in our patient. However, BBE's clinical criteria were met.

Imaging of the brain supports the diagnosis of BBE as abnormal findings are present in only 30% of patients (2). Typical findings of BBE include patchy or confluent, moderately extensive T2/FLAIR hyperintensity in the midbrain and pons, which were evident in our case (5).

There are no specific treatment guidelines for BBE. In a resistant case of BBE, it was discovered that steroids and immunoglobulins brought about a resolution(6). Patients with BBE frequently recover spontaneously, regaining the baseline functional status within six months of diagnosis in the majority of cases, as was the case with our patient.

Conclusions

In conclusion, BBE is majorly a major clinical diagnosis characterized by the triad of ataxia, ophthalmoplegia, and altered sensorium. The MRI plays a supportive role in the diagnosis of BBE. We reported a case with typical clinical and radiological findings of BBE; however, further studies with multiple numbers of patients with follow-up imaging are needed. Moreover, newer studies have also mentioned the overlap syndromes involving both CNS and PNS; hence, more detailed studies with larger sample sizes are recommended to fully understand the disease etiopathogenesis, natural course of disease, and imaging findings.

Abbreviations

BBE	Bickerstaff brainstem encephalitis
CSF	Cerebrospinal fluid
OPD	Out-patient department
MRI	Magnetic resonance imaging
CNS	Central nervous system
PNS	Peripheral nervous system

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SS, AM contributed by literature search, writing the initial draft and figures. KB, PN contributed by conception and design of the work, critical revision of the article. All authors have read and approved the manuscript.

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