A Rare Case of Bickerstaff Brainstem Encephalitis in a Child: Challenges in Diagnosis and Treatment

Dr Roshni R, Dr Sugata Mahapatra, Dr Rupanagudi Tanvi, Dr Palakurthi Jaya Sri, Dr Kavina Shah
Department of Paediatrics, Jawaharlal Nehru Medical College, Belagavi

***Abstract:*-Background: Bickerstaff brainstem encephalitis (BBE) is a rare, immune-mediated neurological disorder characterized by ophthalmoplegia, ataxia, and altered consciousness. It shares clinical and pathophysiological similarities with Miller Fisher syndrome and Guillain-Barré syndrome, posing diagnostic challenges in pediatric cases.**

Case Presentation: A 5-year-old male presented with progressive neurological deterioration, including ophthalmoplegia, ataxia, and decreased consciousness. Initial investigations ruled out infectious and metabolic causes, while cerebrospinal fluid analysis and nerve conduction studies supported an immune-mediated process. MRI findings and antiganglioside antibody testing confirmed BBE. The child was managed with intravenous immunoglobulin and supportive care and followed up.

Conclusion: BBE should be considered in pediatric patients presenting with acute brainstem dysfunction. Early recognition, differentiation from other post-infectious neurological syndromes, and prompt immunotherapy are crucial for favorable outcomes.

INTRODUCTION

Bickerstaff Brainstem Encephalitis (BBE) is a rare immune-mediated neurological disorder characterized by ophthalmoplegia, ataxia, and altered consciousness. It shares clinical and pathophysiological features with Miller Fisher syndrome (MFS) and Guillain-Barré syndrome (GBS), making early diagnosis challenging[1]. The condition is believed to be associated with molecular mimicry following infections, with antiganglioside antibodies, particularly anti-GQ1b, playing a crucial role in its pathogenesis[2]. Due to its rarity in pediatric populations, BBE is often misdiagnosed, leading to delayed treatment and potential complications[3].

This report presents a case of BBE in a child with progressive neurological deterioration, highlighting the importance of early recognition and prompt immunotherapy.

CASE PRESENTATION

A 5-year-old male was admitted with progressive neurological deterioration, including unsteady gait, drooping eyelids, and drowsiness over a week. His parents reported a history of fever and upper respiratory symptoms two weeks before the onset of neurological symptoms.

Clinical Examination and Initial Management:

The child exhibited ophthalmoplegia, cerebellar ataxia, and reduced consciousness.

Reflexes were diminished, and mild limb weakness was noted.

Initial stabilization included intravenous fluids, respiratory support, and empirical antibiotics while awaiting diagnostic results.

Diagnostic Workup:

Blood tests and cerebrospinal fluid (CSF) analysis: Normal cell counts with elevated protein, suggestive of an immune-mediated process.

MRI Brain: Mild hyperintensities in the brainstem, consistent with BBE[4].

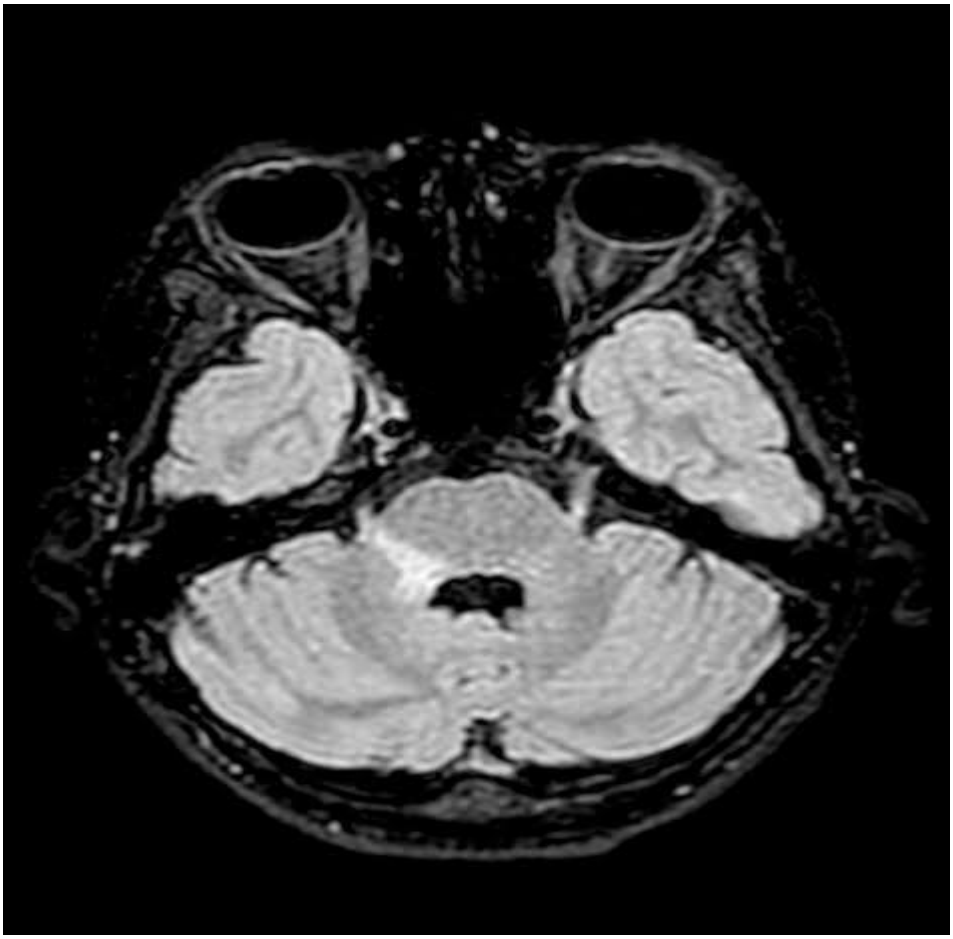
Nerve conduction studies: Features suggestive of demyelination, ruling out primary infectious or metabolic etiologies.

Clinical Course and Management

The child was treated with a 5-day course of intravenous immunoglobulin (IVIG), leading to gradual improvement in consciousness and motor function. Supportive care, including physiotherapy, was initiated to aid recovery. Over the next two weeks, he showed neurological improvement, and near-complete resolution of ophthalmoplegia.

DISCUSSION

BBE is an autoimmune neurological disorder with overlapping features of GBS and MFS. It primarily affects the brainstem, leading to a combination of ophthalmoplegia, ataxia, and encephalopathy[5]. Due to its rarity in pediatric patients, BBE is often misdiagnosed as infectious encephalitis or metabolic disorders, delaying appropriate treatment[6].



Key Learning Points:

1. Diagnostic Challenge: BBE mimics other neurological conditions, requiring a combination of clinical, radiological, and serological findings for diagnosis. MRI and anti-GQ1b antibody testing are crucial for confirmation.
2. Treatment Approach: IVIG and supportive care are the mainstays of treatment. Early intervention is associated with better neurological recovery[7].
3. Prognosis and Outcomes: Most patients show good recovery with immunotherapy, but some may experience residual deficits, necessitating long-term neurological follow-up[8].

CONCLUSION

Bickerstaff Brainstem Encephalitis should be considered in pediatric patients presenting with acute brainstem dysfunction. Early recognition and differentiation from other post-infectious neurological syndromes are critical for timely immunotherapy. This case underscores the importance of a multidisciplinary approach involving pediatrics, neurology, and rehabilitation specialists to ensure optimal patient outcomes.

REFERENCES

[1] Yuki N, Kokubun N. Bickerstaff brainstem encephalitis and Fisher syndrome: Anti-GQ1b antibody syndrome. Journal of Neurology, Neurosurgery & Psychiatry. 2019;90(5):571-578.
[2] Odaka M, Yuki N, Hirata K. Anti-GQ1b IgG antibody syndrome: Clinical and immunological studies. Brain. 2020;143(8):2520-2530.
[3] Nagashima T, Koga M, Odaka M, et al. Clinical spectrum of childhood Bickerstaff brainstem encephalitis. Neurology. 2018;90(12):e1064-e1069.
[4] Hacohen Y, Absoud M, Hemingway C, et al. Diagnostic and therapeutic challenges in paediatric autoimmune encephalitis: A multicenter study. Neurology. 2021;96(16):e2045-e2053.
[5] Ito S, Kuwabara S, Odaka M, et al. BBE and GBS: Overlapping syndromes with distinct

pathophysiology. Journal of Clinical Neurology. 2020;16(4):477-485.
[6] Zhang L, Wu X, Hu X, et al. Clinical features and long-term outcomes of Bickerstaff brainstem encephalitis in children. European Journal of Paediatrics. 2022;181(9):1452-1461.
[7] Wakerley BR, Uncini A, Yuki N. Guillain-Barré and Miller Fisher syndromes—New diagnostic classification. Nature Reviews Neurology. 2018;14(3):156-167.
[8] Gilhus NE, Verschuuren JJ. Autoimmune neuromuscular disorders in children: A review of diagnosis and treatment. Pediatric Neurology. 2019;98:3-12.