

# Remarkable Response of Autoimmune Basal Ganglia Encephalitis to Early Steroid and Immune Therapy: A Case Report

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**Abstract:** *Basal ganglia encephalitis is a rare inflammatory condition affecting the basal ganglia. The etiology of basal ganglia encephalitis can vary and includes infectious, autoimmune, and paraneoplastic causes. It can affect individuals of all ages, with a higher incidence in children and young adults. The clinical presentation is diverse, with movement disorders, cognitive impairment, behavioral changes, seizures, and autonomic dysfunction may also be present. The diagnosis of basal ganglia encephalitis requires a combination of clinical assessment, neuroimaging, cerebrospinal fluid analysis, and specific laboratory tests. Treatment involves addressing the underlying cause if identified, along with symptomatic management and immunotherapy for autoimmune cases. Prognosis can vary depending on the etiology, with infectious cases often responding well to appropriate antimicrobial therapy, while autoimmune and paraneoplastic cases may require long - term immunosuppression.*

**Keywords:** Basal ganglia encephalitis, Parkinsonism, Autoimmune encephalitis, Caudate, Putamen

## 1. Introduction

Basal ganglia encephalitis (BGE) is a rare neurological condition characterized by inflammation and dysfunction of the basal ganglia, a group of structures deep within the brain that play a crucial role in movement control, cognition, and behavior regulation. It causes motor and behavioral abnormalities symptoms and symptoms of parkinsonism such as akinesia, rigidity, and tremors. It can affect individuals of any age, although it is more commonly observed in children and young adults. It occurs due to autoimmune inflammation of the basal ganglia structures. There are various autoantibodies recognized to trigger this type of inflammation like dopamine D2 receptor (D2R), N - methyl - D - aspartate receptor (NMDAR), and anti - LGI<sup>1</sup>. It also occurs as a paraneoplastic manifestation due to some tumors, then anti - recoverin antibodies are classically associated. Herein, we report a case of autoimmune basal ganglia encephalitis in an 11 - year - old child presented with typical clinical feature of basal ganglia encephalitis, who benefited from steroid and immune therapy.

## 2. Case Presentation

A previously healthy 11 years old male child was presented to our hospital with 1 month history of intermittent fever, Vomiting and peri - umbilical pain before 15 days. He was treated by a private practitioner before admission despite that fever spikes continued. Fever was low grade maximum documented 100 F, inter febrile period for 2 - 3 days in which child was playful followed by a febrile period for 1 week then reappearance of fever. In view of persistent symptoms, he was admitted to our hospital. On admission his temperature was 98 f, pulse rate was 110/min, spO<sub>2</sub> was 100% and respiratory rate was 23/min. Fever and abdominal symptoms subsided after a week, but he started having difficulty in walking. He must hold up both hands while walking. However, he could get up from the bed on his own.

He was awake and sitting independently. There was Speech loss (Motor Aphasia), Mask like facies and Rigidity in all limbs. The gait was narrow based and reduced arm swing with Chorea presented in all limbs. There was brisk dip tendon reflex and Extensor plantar reflex present. This problem persisted for a week without worsening or improving then he developed difficulty in speaking. Although he could understand and follow commands, he had fluent speech with frequent pauses. Investigations showed Hb 10.4 g/dl (11.8 - 15), MCH 24.9 pg (26.5 - 33.5) RDW 15.7% (11.7 - 14.4), CPR (C reactive protein) 69.70 mg/l (<6); liver function test showed SGPT 48.9 IU/L (0 - 42), Protein 8.73 gm/dl (6.3 - 8.3). Serum Widal showed titers of S. typhi O - 1: 120 & S. typhi H - 1: 160. X ray chest was inconclusive. USG abdomen suggestive of Enteritis. Other tests including Neuromyelitis Optica antibody, Myelin oligodendrocyte glycoprotein antibody, Antinuclear antibodies and anti-dsDNA were done to rule out vasculitis. COVID- 19 antibody tests were found non - reactive. CSF Oligoclonal band - HO showed CSF IgG INDEX 0.91 INDEX (0.3 - 0.7) & serum IgG 28.7 g/dl (6.98 - 15.6) suggestive of inflammation. No organism found on CSF Culture. MRI Brain showed bilateral asymmetrical T2WL and FLAIR hyperintensity with subtle diffusion restriction involving bilateral caudate & lentiform nucleus, thalamus, bilateral cerebral peduncle, bilateral medial temporal lobes, hippocampus and anterior part of body of corpus callosum. Autoantibodies in serum, as well as CSF for NMDAR, dopamine D2 receptor (D2R), LGI1, anti - recoverin, anti - contactin associated protein 2 (CASPR2), were all found negative. He was treated with a combination of Methyl Prednisolone and IV Immunoglobulin as per advice of pediatric neurologist. In addition to this he was also treated with Ceftriaxone, Paracetamol, Clonazepam for infection control and symptomatic management. On treatment child's symptoms started to improve. He started speaking on the 2nd day of treatment. Gradually tone improved and involuntary movement decreased and ultimately subsided.

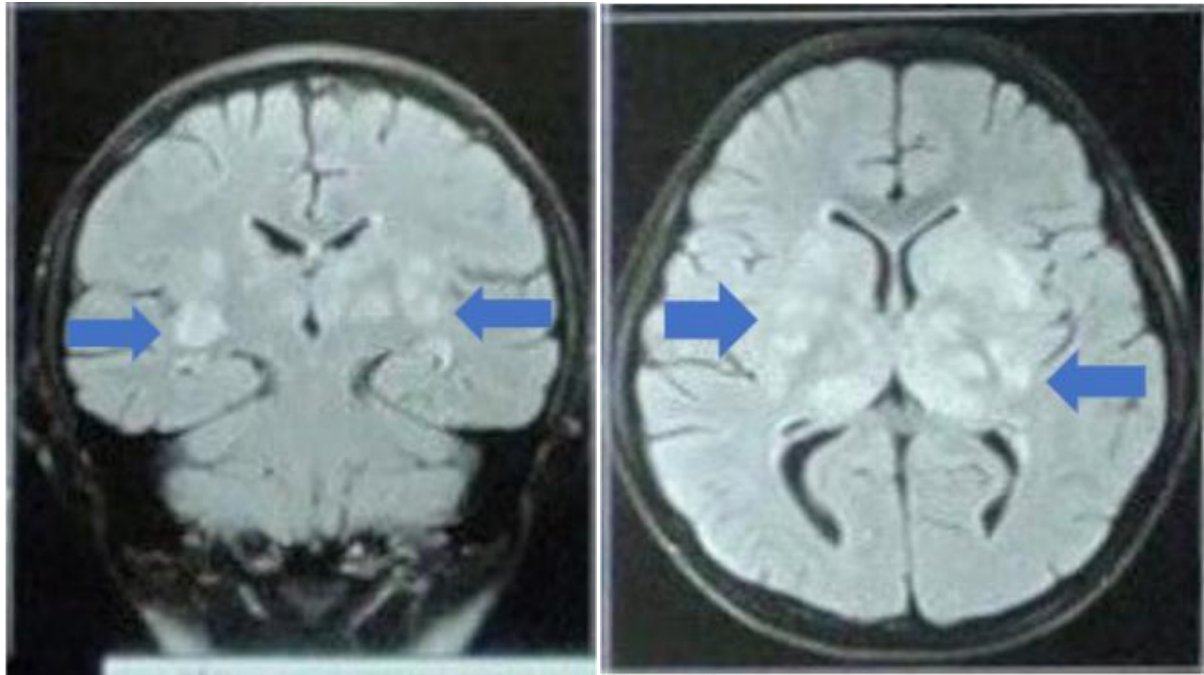
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He had transient complaints of urgency and increased frequency of micturition which subsided spontaneously. He

was later discharged when he was vitally stable with good oral intake on oral steroids.



**Figure 1:** MRI of coronal and horizontal section of brain showing bilateral asymmetrical enhancement involving caudate & lentiform nucleus and thalamus

### 3. Discussion

Basal ganglia encephalitis (BGE) is a rare neurological condition, where the body's immune system mistakenly targets and attacks the receptors in the basal ganglia of the brain. It can affect individuals of all ages, but it appears to be more common in children and young adults. It is associated with tumors in a subset of cases. However, it is important to note that the majority of cases do not have an identifiable tumor. Here we discuss the case where it most likely triggered by an infection. It is thought that the immune response to the infection may cross-react with the basal ganglia receptors, leading to the development of autoantibodies and subsequent autoimmune encephalitis. Genetic factors may contribute to an individual's susceptibility to developing encephalitis. So, it is thought that the interplay between genetic factors, environmental triggers (such as infections), and other underlying predisposing factors likely influences the development of this type of encephalitis<sup>3</sup>.

The clinical features can vary among individuals, but there are common patterns of symptoms that have been observed. It can present with various types of movement disorders including chorea (involuntary, jerky movements), dystonia (sustained muscle contractions causing abnormal postures), and parkinsonism (tremors, rigidity, and bradykinesia)<sup>2</sup>. These movement abnormalities often affect the limbs but can also involve the face and trunk. Psychiatric symptoms include behavioral changes, mood disorders (such as depression or mania), psychosis (hallucinations, delusions), agitation, anxiety, and cognitive impairments and speech and language difficulties, such as dysarthria (slurred speech), expressive or receptive aphasia (difficulty with speaking or

understanding language), or verbal perseveration (repetitive speech patterns).

The diagnosis of Basal ganglia encephalitis involves a combination of clinical evaluation, neuroimaging, cerebrospinal fluid (CSF) analysis, and the detection of specific autoantibodies targeting the receptors. Brain imaging, particularly magnetic resonance imaging (MRI), is an important diagnostic tool. It can help identify any structural abnormalities, inflammation, or atrophy in the basal ganglia or other affected brain regions. These findings can support the diagnosis and exclude other potential causes of symptoms. CSF analysis often shows increased protein levels and lymphocytic pleocytosis, indicating an inflammatory process within the central nervous system. Detection of specific autoantibodies targeting the basal ganglia receptors is essential for confirming the diagnosis. Laboratory testing methods, such as cell-based assays or enzyme-linked immunosorbent assays (ELISAs), can be used to detect these autoantibodies in the blood or CSF. Positive autoantibody results support the diagnosis, although it is important to interpret the results in the context of the clinical presentation and other diagnostic findings. Here we did a comprehensive evaluation for the accurate diagnosis and to differentiate it from other neurological and psychiatric conditions with similar manifestations.

The treatment of Basal ganglia encephalitis involves a multidisciplinary approach that aims to suppress the autoimmune response, manage symptoms, and address any underlying triggers. High-dose intravenous corticosteroids, such as methylprednisolone, are often used as the initial treatment to rapidly suppress the immune response. Oral prednisone may be continued as a maintenance therapy. Intravenous Immunoglobulin (IVIG) can be administered

intravenously and acts by modulating the immune response. It is often used as an adjunctive treatment in combination with corticosteroids or as an alternative in cases where corticosteroids are not well-tolerated. This patient showed good response to this combination therapy. Rituximab or cyclophosphamide are considered for those who show inadequate response to first-line immune therapies or develop relapse despite appropriate maintenance therapy.<sup>4</sup> If infectious etiology is suspected, antibiotics or antiviral can be used. Symptomatic treatment for stiffness and movement disorders can be used to alleviate symptoms. If an underlying tumor is identified, appropriate treatment modalities such as surgical resection, chemotherapy, or radiation therapy should be considered.

Early recognition and initiation of appropriate treatment, including immunotherapy, can significantly impact the prognosis. With timely and effective treatment, there is a potential for neurological and functional recovery in many cases. However, the extent and speed of recovery can vary among individuals. Some may experience complete resolution of symptoms, while others may have residual deficits or persistent symptoms. Relapses or exacerbations of symptoms can occur, particularly in cases associated with underlying tumors. Some individuals may experience long-term cognitive and behavioral sequelae including memory deficits, executive dysfunction, mood disorders, and behavioral changes. Our patient showed excellent recovery during his follow-up visit. His tone improved and involuntary movements subsided, and he also started walking on his own.

#### 4. Conclusion

Our case highlights that although the detection of auto-antibodies is a confirmatory diagnostic test, many autoimmune encephalitis series are antibody negative. Therefore, it is very important to treat this patient rapidly with steroids and immunoglobulins when clinical suspicion of basal ganglia encephalitis is so high, is crucial for better outcomes.

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