

## Case Report

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# Guillain Barre syndrome and hyper-immunoglobulin E syndromes: an unusual correlation

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## ABSTRACT

Hyper immunoglobulin E (IgE) syndrome (HIES), also known as Job's syndrome, is a rare primary immunodeficiency disorder characterized by elevated levels of serum IgE, recurrent skin and lung infections, and eczema. Guillain-Barré syndrome (GBS) is an acute autoimmune condition that affects the peripheral nervous system, leading to muscle weakness and paralysis. This case report presents a unique instance of a 24-year-old male diagnosed with both HIES and GBS, exploring the clinical presentations, diagnostic challenges, and treatment approach.

**Keywords:** Hyper IgE syndrome, Guillain Barre syndrome, Autoimmune, Peripheral nervous system

## INTRODUCTION

Hyper IgE syndrome (HIES) manifests with symptoms such as recurrent skin abscesses, pneumonia, eczema and a high serum IgE level, often exceeding 1000 IU/ml. Elevated IgE can also be detected in a spectrum of conditions like asthma, atopic eczema, vasculitis, anaphylaxis, parasitic infection, IgE myeloma, as well as HIES.<sup>1</sup> The binding of IgE to specific antigens causes mast-cell degranulation, resulting in an inflammatory reaction.<sup>2</sup> GBS typically follows an infection and is characterized by rapid onset muscle weakness which can progress to paralysis.<sup>3</sup> Dysimmune neuropathy is an etiologically heterogeneous entity with the diverse clinical presentations.<sup>4</sup> The coexistence of these two conditions in a single patient is exceptionally rare, posing significant diagnostic and therapeutic challenges.

## CASE REPORT

### Patient history

A 24-year-old male presented to our OPD with complaints of weakness of both upper and lower limbs since 12 days

before admission, which was sudden in onset, gradual in progression such that since 4 days before presentation he also developed hoarseness of voice, with difficulty in swallowing liquids as well as solids.

Two months ago, he had a history suggestive of eczema in the form of periorbital puffiness and redness which improved gradually with medical treatment.

### Clinical presentation

On examination, patient was having flaccidity in both upper and lower limbs with absent reflexes, and absent plantar reflex. Patient gag reflex as well as cough reflex were weak. Patient single breath count (SBC) was 20.

### Diagnostic workup

#### Laboratory tests

Routine blood tests showed elevated white blood cell counts and significantly high serum IgE levels (1500 IU/ml). Inflammatory markers in the form of ESR/CRP were elevated. However, ANA/ANCA profile were negative.

### Electrophysiological studies

Nerve conduction studies were performed, revealing demyelinating polyneuropathy consistent with GBS.

### Imaging

Magnetic resonance imaging (MRI) of the brain and spine showed no abnormalities, ruling out central nervous system involvement.

### Treatment approach

#### Initial management

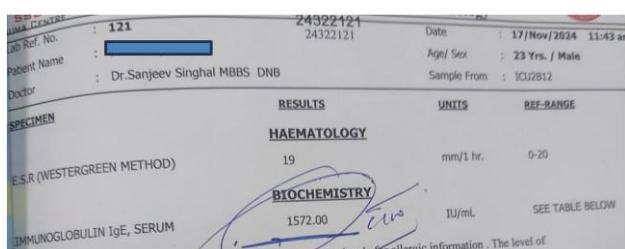
The patient was treated with intravenous immunoglobulin (IVIG) for GBS, which led to a gradual improvement in muscle strength. Mast cell inhibitors and anti-histaminic drugs were given for management of conditions associated with hyper-IgE-emia. Rest management involved providing supportive treatment and physiotherapy.

#### Long-term management

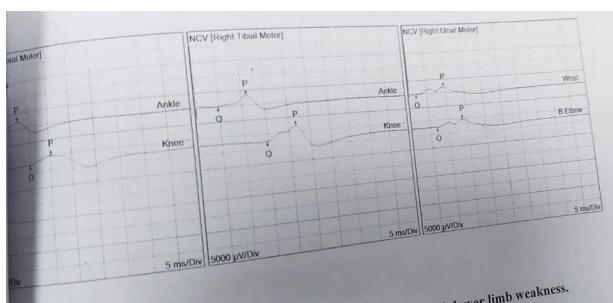
Physiotherapy was recommended to aid in the recovery of muscle strength and function.

### Prognosis

The patient showed significant improvement in neurological function over the subsequent months. He also subsequently did not develop episodes of eczema or skin infections.



**Figure 1: Haematology report showing elevated IgE levels.**



**Figure 2: Nerve conduction studies showing prolonged latency, decreased amplitude and remarkably decreased conduction velocity.**

**Table 1: Laboratory parameters.**

Laboratory parameters	Report
ESR	49 mm/hr
Hemoglobin	13.5 gm/dl
Total cell counts	8500/mm <sup>3</sup>
Platelet count	2.54 lac/mm <sup>3</sup>
Absolute eosinophil count	240
Serum IgE levels	1572 IU/ml
Nerve conduction studies	Prolonged distal latencies with conduction block



**Figure 3: MRI cervical spine showing no significant abnormality.**

### DISCUSSION

Although peripheral nerve involvement is not a well-known complication, a few cases of hyper-IgE-emia associated with subacute or chronic polyneuropathy have been reported.<sup>4,5</sup>

As per our research, only one previous case of GBS with raised IgE was reported, although the occurrence could be coincidental. Elevated IgE might have played a role in the pathogenesis of this case of GBS.<sup>6,7</sup>

In the pathogenesis of GBS, multiple factors including breakdown of blood-nerve barrier (BNB) and extent of inflammation are all relevant to producing auto-antibody-mediated nerve fiber injury.<sup>8</sup> A healthy peripheral nervous system is tightly sealed by the BNB, which has a very low permeability to serum immunoglobulins.<sup>9</sup>

Therefore, in the absence of a local inflammatory response, diffusion of an antibody into the nerve is unable to initiate a functional deficit.<sup>8</sup> Mast cells activated by IgE trigger disruption of the BNB as an initial GBS insult, and this is followed by macrophage activation.<sup>10</sup> Therefore, hyper-IgE-emia may increase the magnitude and rate of neural damage in early GBS. Further research regarding the role of IgE in GBS might help to identify the mechanisms underlying the induction of GBS.

## CONCLUSION

This case highlights the complexity of managing a patient with concurrent HIES and GBS. It underscores the importance of a multidisciplinary approach in diagnosing and treating such rare clinical intersections. Further research is needed to understand the potential immunological links between these two conditions and to develop targeted therapeutic strategies.

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