Hyper IgE Syndrome with Guillain-Barré Syndrome: Pure Coincidence or a Real Connection?

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ABSTRACT

Hyper 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 patient diagnosed with both HIES and GBS, exploring the clinical presentations, diagnostic challenges, and treatment approach.

INTRODUCTION

Hyper IgE Syndrome manifests with symptoms such as recurrent skin abscesses, pneumonia, eczema and a high serum IgE level, often exceeding 1000 IU/mL. Guillain-Barré Syndrome typically follows an infection and is characterized by rapid onset muscle weakness which can progress to paralysis. The coexistence of these two conditions in a single patient is exceptionally rare, posing significant diagnostic and therapeutic challenges.

CASE PRESENTATION

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, rapidly progressive such that since 4 days before presentation he also developed hoarseness of voice, with difficulty in swallowing liquids as well as solids. 2 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. His gag reflex as well as cough reflex were weak. His single breath count (SBC) was 20.

Diagnostic Workup

Laboratory Tests

Routine blood tests showed elevated white blood cell counts with eosinophilia (22%) and significantly high serum IgE levels (1500 IU/mL). Inflammatory markers in the form of ESR/CRP were elevated. CSF analysis revealed cyto-albuminological dissociation. ANA/ANCA profile were negative.

Electrophysiological Studies

Nerve conduction studies were suggestive of demyelinating polyneuropathy consistent with Guillain-Barré Syndrome.

Imaging

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

Treatment

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.

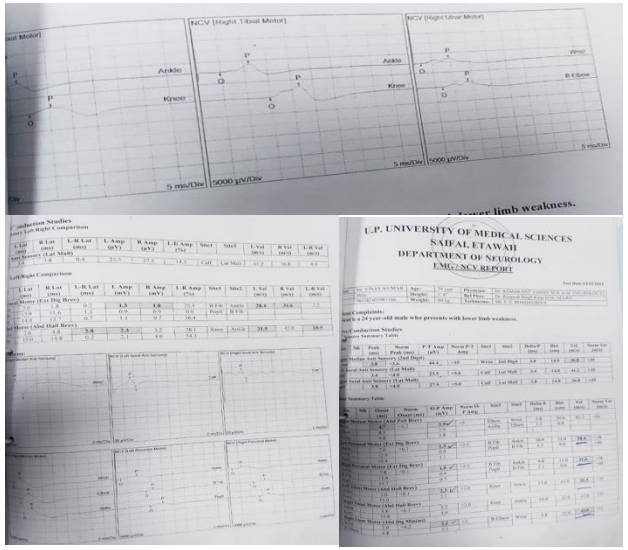

Figure 1:- Nerve Conduction Studies

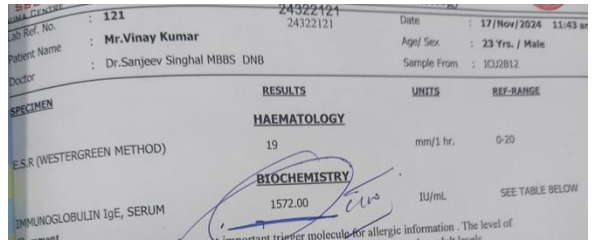

Figure 2:- Hematology report showing elevated IgE levels

DISCUSSION

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.[2] Mast cells activated by IgE trigger disruption of the BNB as an initial GBS insult, and this is followed by macrophage activation.[3] Therefore, hyper-IgE-emia may increase the magnitude and rate of neural damage in early GBS.

CONCLUSION

This case highlights the complexity of managing a patient with concurrent Hyper IgE Syndrome and Guillain-Barré Syndrome. Further research is needed to understand the potential immunological links between these two conditions and to develop targeted therapeutic strategies.

1. Amin K. The role of mast cells in allergic inflammation. Respir Med 2012;106:9-14
2. Kimura A, Yoshino H, Yuasa T. Chronic inflammatory demyelinating polyneuropathy in a patient with hyperIgEaemia. J Neurol Sci 2005;231:89-93.
3. Dines KC, Powell HC. Mast cell interactions with the nervous system: relationship to mechanisms of disease. J Neuropathol Exp Neurol 1997;56:627-640.

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