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Case Report

Guillain-Barre Syndrome Masquerading as Acute Lymphoblastic Leukemia Relapse

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Abstract

Gullain-Barre Syndrome (GBS) is an acute immune-mediated inflammatory demyelinating polyneuropathy [1]. it is characterised by motor, sensory symptoms, cranial nerves, particularly facial and bulbar with autonomic involvement [1]. In seventy percent of cases there is a preceding infection. Thus, it represents a post-infectious auto-immune disorder [2]. Rarely GBS has been reported in association with hematologic malignancies in adults [3]. In this literature, we present a unique case of ALL with GBS mimicking as relapse while on maintenance therapy. This case report demonstrates the necessity of appropriate diagnostic workup required to parse out the differentials.

Keywords: Guillain Barre syndrome; Acute lymphoblastic leukemia; AMAN variant GBS

Abbreviations: GBS: Gullain-Barre Syndrome; ALL: Acute Lymphoblastic Leukemia; ICiCLE: Indian Childhood Collaborative Leukemia Group; CMAP: Compound Muscle Action Potential; AMAN: Acute Motor Axonal Neuropathy; IVIG: Intra Venous Immunoglobulin

Introduction

Gullain-Barre Syndrome (GBS) is an acute immune-mediated inflammatory demyelinating polyneuropathy [1]. In 1916 it was first described by Guillain, Barré and Strohl [2]. Incidence ranges from 0.8-1.9 per 100,000 [2]. It is characterised by symmetric weakness in the limb muscles with sensory symptoms, cranial nerves, particularly facial and bulbar with autonomic involvement [1]. In seventy percent of cases there is a preceding infection. Thus, it represents a post-infectious auto-immune disorder [2]. In twenty-five percent of cases, there is respiratory muscle involvement requiring ventilatory support. Respiratory involvement is a life-threatening feature in GBS. Rarely GBS has been reported in association with hematologic malignancies in adults [3]. It is very rarely seen in children [3]. It must be differentiated from other forms of neuropathies. It is important from the therapeutic point of view. Most common malignancy of childhood is Acute Lymphoblastic Leukemia (ALL). In ALL, five to eight percent have neurological involvement at initial diagnosis. But up to thirty percent have neurological involvement at relapse [4]. We present the case of a pediatric ALL with GBS mimicking a relapse while on maintenance therapy. This case report demonstrates appropriate diagnostic workup required to parse out the differentials.

Case Report

A 4-year-old boy child, was on treatment for acute B cell Lymphoblastic Leukemia (Precursor B-cell ALL) intermediate risk, with Indian Childhood Collaborative Leukemia group (ICiCLE) protocol and on the maintenance phase of therapy, presented with pain in all four limbs followed by acute onset weakness of all four limbs of one week duration, more in lower extremities. He also had difficulty in swallowing both solid and liquids. There was also urinary retention and constipation. No prior history of fever or infectious foci. On admission, the child was afebrile, conscious, stable on cardiorespiratory evaluation. On neurological examination, the patient had quadriparesis with hypotonia (3/5 proximally and distally in upper extremities; 2/5 proximally and distally in lower extremities on the Medical Research Council muscle strength grading), absent deep tendon reflexes on all limbs. Gag reflex was also absent. Plantar response was flexor. Sensory system intact. No cerebellar or meningeal signs were elicited. Subsequently in period of three days, child developed respiratory failure, was intubated and ventilated. Provisional differential diagnosis considered were symmetrical polyradiculopathy of ALL relapse or GBS. CSF analysis done showed albumin cytological dissociation. Nerve conduction velocity study

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showed decreased CMAP (Compound Muscle Action Potential) amplitude with prolonged distal latency/decreased conduction velocity and F wave was poorly formed. It was suggestive of axonal motor neuropathy with secondary demyelination. Later child developed autonomic dysfunction in the form of excessive sweating and tachycardia, which was managed with propranolol. The pattern and evolution of the neurological symptoms and electrodiagnostic features were conclusive for Acute Motor Axonal Neuropathy (AMAN) variant of GBS. The child received Intravenous immunoglobulin (IVIG) at a dose of 2gm/Kg. An elective tracheostomy was done on day 10 of mechanical ventilation. Post IVIG, he improved clinically. Maintenance phase with oral 6-Mercaptopurine and methotrexate were restarted. He improved gradually over a period of two months. Tracheostomy was decannulated then. Currently child is six months post GBS, on maintenance therapy and on remission without any neurological deficit.

Discussion

Guillain-Barre syndrome is an acute immune-mediated polyneuropathy. Demyelinating and axonal subtypes are described [1]. It is extremely rare in paediatric haematological malignancies [5,6]. The pathogenesis proposed for GBS in lymphoproliferative disorder include the increased immunological vulnerability of the peripheral nervous system. Infective triggers could mount an immune neuropathy in this background. The other differential for GBS is symmetrical polyradiculopathy. It is a rare complication of ALL, occurs due to leukemic cells infiltration. It should be differentiated from acute inflammatory demyelinating polyneuropathy. Polyradiculopathy associated with ALL is usually clinically indistinguishable from GBS [7].

Vincristine is an important cause for neuropathy in ALL. "Dying back" sensory involvement is classically described for vincristine [8]. In patients with Charcot-Marie-Tooth disease, Vincristine may also cause a fulminant neuropathy with severe weakness. However, our child, was on maintenance phase with 6 mercaptopurine and methotrexate. The clinical and electrophysiological features were not suggestive of vincristine neuropathy.

Autoimmune disorders occur as a part of paraneoplastic syndrome in ALL [9]. There is depletion of the regulatory T cells in ALL. They usually suppress auto-reactive T cells. This leads to auto-immune trigger. This could be either due to disease or intensive chemotherapeutic drugs. This has been hypothesised for immune thrombocytopenia in ALL [9]. Same can be postulated for genesis of acute immune neuropathies in ALL. The association can be coincidental or causal. In our child as the disease was in remission, probability of leukemic infiltration or paraneoplastic neurologic manifestation is unlikely. The etiology may be GBS secondary to some infectious trigger. But could not be proven. Prior absence of fever may be attributed to immune suppression by chemotherapeutic drugs.

On nerve conduction velocity study showed decreased CMAP (Compound Muscle Action Potential) amplitude with prolonged distal latency/decreased conduction velocity and poorly formed F wave was noticed. This was suggestive of axonal motor neuropathy with secondary demyelination.

Conclusion

A high index of suspicion is needed for differentiating from other causes for neuropathies like leukemic infiltration, paraneoplastic process, GBS and vincristine related neuropathy. CSF analysis and nerve conduction studies aid in diagnosis. It has therapeutic significance, to initiate timely immunomodulatory therapies for GBS or diagnosis of neurological relapse.

Author Statements

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Conflict of Interest

There is no conflict of interest for all the authors.

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