GUILLAIN BARRE SYNDROME IN EARLY INFANCY: A CASE REPORT

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Abstract

Guillain Barre Syndrome (GBS) off late has been the most frequent cause of acute flaccid paralysis worldwide, replacing poliomyelitis which dominated in the past few decades. A 4 months old female infant was presented with sudden onset; nonprogressive weakness of both lower limbs with areflexia and urinary retention such clinical features consistent with Guillain Barre Syndrome. She was treated with immunoglobulin 0.4gm per kg per day for 5 days and responded well to treatment and started recovery after two weeks. Though Guillain Barre Syndrome is uncommon before six months of life, it should be suspect in infant those presented with bilateral and relatively symmetric weakness with hyporeflexia or areflexia with preceding antecedent events.

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INTRODUCTION

With the reduction of poliomyelitis, Guillain Barre Syndrome has become the most common cause of acute flaccid paralysis in both developed and developing countries with an incidence 0.6-2.4 cases per 100000 children below 15 years of age\textsuperscript{[1]}. Guillain, Barre and Strohl (1916) described a benign polyneuritis with albumin-cytological dissociation in the cerebrospinal fluid \textsuperscript{[2]}. Age is an important factor determining outcome and prognosis in children and is said to be favourable as compared to adults \textsuperscript{[3]} and there is a small predominance in male. In the past, Guillain Barre Syndrome was considered a single disease but now it has become clear that this clinical entity can be produced by different pathological subtypes like acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy, acute motor sensory neuropathy and mixed type. There is substantial evidence to support an autoimmuno cause of this syndrome and the autoantibody profile has been helpful in confirming the clinical and electrophysiological relationship. The main feature is progressive bilateral and relatively symmetric weakness of the limbs with generalized hyporeflexia or areflexia. A cranial nerve involvement and autonomic disturbances is not uncommon in most of the cases. In two third of cases, it is preceded by an acute infectious disease. Guillain Barre Syndrome is uncommon before six months of life. We describe a case of 4 months infant with clinical features consistent with Guillain Barre Syndrome.

CASE REPORT:

A 4 months old female infant of primipara mother with adequate antenatal care and immunization was born vaginally with uneventful perinatal and natal period and she was on exclusively breast fed. She was presented with sudden onset, nonprogressive weakness of both lower limbs with urinary retention. She had also history of loose
motion one week prior to illness and was not recently vaccinated.

On examination, she was afebrile, vital stable. She was conscious without cranial nerve involvement with grade 0 power and areflexia in both lower limb and normal power and reflexes in both upper limb with palpable distended bladder and exaggerated triple response. Head, neck, throat and spine examination was within normal limit.

Blood investigations showed normal hemoglobin, electrolytes, urea/creatinine except polymorphonuclear leucocytosis on peripheral smear. ECG was within normal limit. Nerve conduction study was suggestive of acute demyelinating polynueropathy with loss of F wave.

She was treated with immunoglobulin 0.4gm per kg per day for 5 days and responded well to treatment and she was in follow up and there was no recurrence.

**DISCUSSION:**

Guillain-Barre syndrome was defined more than a century back but the clinical characteristics in children from different studies are not consistent, which might be due to geographical and racial diversity. The clinical characteristics in pediatric Guillain Barre Syndrome differ from that in adult ones [4]. Guillain Barre Syndrome is very rare in early infancy and in childhood, it is usually occurs after the age of 3 years but van der Linden et al [5] reported youngest infant of 7 months old with Guillain Barre Syndrome and our case is 4 months old. There is a small predominance of male in occurrence of Guillain Barre Syndrome cases [6]. Seasonal or monthly variation in the occurrence of Guillain Barre Syndrome is noted by various studies [6, 7, 8]. The reason for this disparity remains unclear and whether the seasonal variation correlates to different pathogens, which might be asymptomatic or
lead to non specific symptoms, warrants further elucidation.

A previous infection should always be searched particularly when trying to define the presence of some microbiological agents more frequently related to Guillain Barre Syndrome. Among these events, the most frequent was unspecific upper respiratory tract infection followed by diarrhea. Our patient had diarrhea one week prior to illness like most of the studies in literature noted that antecedent’s factors associated with occurrence of Guillain Barre Syndrome and most commonly URTI and diarrhea [1, 7, 9, 10]. In relation with clinical presentation, the main feature is progressive bilateral and relatively symmetric weakness that progresses over a period of 12 hours to 28 days before a plateau is reached with hyporeflexia or areflexia. Our patient had classical clinical presentation of symmetrical bilateral lower limb weakness with areflexia. We did not found cranial nerve or respiratory involvement but dysautonomic signs and symptoms in form of urinary retention and exaggerated triple response were noted in our case. Dysautonomic signs and symptoms like urinary retention, urinary incontinence, constipation, fecal incontinence, systemic arterial hypertension, tachyarrhythmia and bradyarrhythmias were recorded by Chatterjee et al and van der Linden et al in their studies [11]. Abnormal rise of CSF protein in Guillain Barre Syndrome may be due to inflammatory reaction in the choroid plexus or disturbances in process of transport or breakdown of blood brain barrier [9]. We did not perform CSF examination as patient was presented in first week of presentation.

The classification of Guillain Barre Syndrome is based on nerve conduction study includes demyelinating and axonal subtypes and there is notable difference in geographic distribution. In Europe and North America, the demyelinating subtypes accounts for up to 90% cases whereas in China, Japan,
Bangladesh and Mexico, the frequency of axonal subtypes ranges from 30-65% and the frequency of demyelinating ranges from 22-46% \(^1\). There is a paucity of comparable data from India but most of Indian studies were reported demyelinating subtypes is the commonest one followed by axonal. Our patient had demyelinating subtypes of Guillain Barre Syndrome.

Immunotherapy like plasma exchange and immune globulin are appeared to be most effective when it was started within the first two weeks after disease onset. Plasma exchange nonspecifically removes antibodies & complement and appears to be associated with reduced nerve damage and faster clinical improvement while immune globulin may act by neutralizing pathogenic antibodies and inhibiting autoantibody-mediated complement activation, resulting in reduced nerve injury and faster clinical outcome. The combination of plasma exchange followed by a course of immune globulin is not significantly better than plasma exchange or immune globulin alone \(^1\). Our case did not have any progression nor any neurodeficit post the therapy with immunoglobulin.

**CONCLUSION:**

Excellent recovery can be expected in nearly all children with Guillain Barre Syndrome; provided respiratory, bulbar and autonomic impairments during the acute phase are managed meticulously. Guillain Barre Syndrome is very rare in early infancy and in childhood. It is usually occurs after the age of 3 years. Inspite that diagnosis of Guillain Barry Syndrome should be considered in infant those presented with bilateral and relatively symmetric weakness with hyporeflexia or areflexia with preceding antecedent events.

**REFERENCES**


(PubMed).