Paradoxical Facial Palsy with Intravenous Immunoglobulin (IVIG) In Acute Inflammatory Demyelinating Polyradiculoneuropathy

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ABSTRACT

Guillain – Barre syndrome is an acute ascending flaccid paralysis that is often preceded by a mild bacterial or viral infection. Anti –ganglioside antibodies detected in serum have been proposed to contribute to the immunopathogenesis of GBS. Management options including supportive care, physiotherapy, intravenous immunoglobulin and plasmapheresis. IVIG have been shown to effectively remove serum anti –GM1 IgG antibo dies which is considered as one of the major contributing factor for the pathogenesis of GBS. We document an interesting case of a four year old girl who was diagnosed with GBS, and later on developed a facial palsy during the IVIG treatment, while weaknes, areflexia and other symptoms were improving. The IVIG treatment has a beneficial effect on GBS, but some times it may not arrest the development of facial palsy. The mechanism of such a variable effect of IVIG on GBS is hypothesized and discussed here.

Key words: Guillian Barre syndrome, IVIg, Facial palsy.

Introduction:

Guillain- Barre syndrome is a peripheral poly neuropathy characterized by acute onset of symmetrical muscle weakness with or without cranial nerve involvement. Some patients develop cranial nerve palsies early on and generalized limb weakness appears as the illness progresses, whereas some experience only regional weakness throughout its course. The facial nerve is the most frequent cranial nerve affected in GBS and is usually affected when limb weakness is severe. Elevated anti ganglioside antibody levels mainly of anti GM1 and anti GD1 have been reported in the serum of patients with GBS and strongly associated with immunopathogenesis and severity of GBS. IVIG use in GBS has been reported to be beneficial. We describe a case of a four year old female child with GBS in whom facial palsy developed during the IVIG treatment when other neurological signs and symptoms were improving.
Case report:

A four year old female child was brought to the department of neurological sciences with a seven days history of gradually progressive weakness of all four limbs. The weakness began initially in the lower limbs with frequent falls and difficulty in standing from sitting and squatting position, followed by difficulty in getting up from the bed and turning sidewards over the bed. After four days of onset of illness child noticed difficulty in raising upper limbs and holding objects. There were no complaints of ocular or bulbar dysfunction. She had no preceding history of diarrheal episodes, respiratory tract infections, exanthematous illness, recent travel, exposure to toxins recent immunization or shellfish ingestion.

On examination the child was alert but irritable on handling. There was hypotonia noted in all four limbs. The best observed power of the lower limbs was reduced to a greater extent and in the upper limbs; there was some reduction in strength bilaterally. Neck and truncal muscle weakness was noted. No facial weakness was noted. A detailed sensory and cerebellar examination could not be done.

Results of laboratory studies at the time of admission included a normal white blood cell count with a normal differential count and normal serum electrolytes. Anti – nuclear antibodies were negative. Thyroid functional tests were normal. Creatinine phosphokinase levels were within normal limits. Cerebro spinal fluid from the spinal tap revealed an increased CSF protein with normal cell count and glucose levels. IgG anti –GM antibody and anti IgM GD1 a are detected in the serum by enzyme linked immunosorbent assay, but IgG and IgM antibodies against other gangliosides were not detected. Nerve conduction studies showed gross reduction in the compound muscle action potentials in both upper and lower limb nerves with absent ‘F’ wave response, suggestive of acute motor axonal neuropathy.

Child was diagnosed as Guillain Barre Syndrome and was started on intravenous immunoglobulin on the first day of admission. Subsequently over the next two days child had shown significant improvement in the motor power with evidence of being able to sit on the bed without support and actively lifting the lower limbs against gravity. During the recovery period child had developed facial asymmetry, with drooling of saliva from right corner of the mouth, deviation of mouth to left side and with a wide palpebral fissure of the right eye. To account for LMN facial weakness MRI brain was done which was unremarkable and blink reflex confirmed right facial nerve dysfunction. Child was given facial nerve stimulation and physiotherapy and completed 5 day treatment of IVIG. Child was discharged on the 9th post admission day with significant improvement in the power of all the limbs and truncal musculature with mild recovery in facial functions. On subsequent review visit after a month, we noticed good improvement in the facial functions along with the other muscle groups.

Discussion:

IVIG and plasmapheresis are the two most commonly used immunotherapies in GBS. It has been shown that IVIG and plasmapheresis are equally effective in reducing morbidity and
mortality in GBS. The practicalities and expense of administering plasmapheresis makes IVIG the immunotherapy of choice in practice, though expensive IVIG is simple to administer with fewer side effects. IVIG is not in routine use as a first-line therapy in the management of GBS in India today and would generally be administered only in a tertiary care center.

Anti ganglioside antibodies other than anti- GM1 IgG and anti- GD1a IgM have not convincingly been linked to myelin and/or axonal damage. Press et al showed there has been a significant decrease in anti GM1 IgG in the blood of GBS patients with no significant effect on anti GD1a IgM after IVIG treatment. Shanbag et al reported statistical significant benefits with IVIG regarding time to recovery and stopping the progression of the disease. In this document we reported a case of GBS in whom IVIG given in the early stage has successfully stopped the progression of disease, by improving the weakness of all limb and truncal musculature. However, facial palsy was developed on the third day of the IVIG treatment. However, there is higher rate of relapse in adults treated with plasmapheresis than IVIG in GBS. We consider this patient did not had a relapse as child developed facial weakness during recovery of the motor power of limb and truncal musculature in spite of IVIG treatment. The most common cranial nerve involved in the GBS is facial nerve and usually affected when limb weakness is more severe which shows similar immunopathological process involved in the cranial nerves along with the peripheral nerves.

The reason for the new onset of facial palsy in a GBS patient who was improving from her signs and symptoms was not clear. Different possibilities and hypothesis were suggested. First possibility is different immune histochemical studies showed prominent staining with anti GT 1a IgG and anti GQ1b IgG antibodies which might be responsible for several cranial nerve palsy. But these antibodies were detected in the serum of this patients as mentioned above. This may suggest some unknown factors might be responsible for the facial nerve palsy in this patient. Second possibility is IVIG has been shown to effectively remove serum anti GM 1 IgG antibodies which is considered as one of the contributing factor of the pathogenesis of GBS but not IgM anti ganglioside antibodies. This suggests IVIG may not clear all types of antibodies which may contribute to the pathogenesis of the facial nerve palsy in this patient. However we are not able to prove this hypothesis because of the lack of serial serum antibody measurements.

Our observation supports the effectiveness of IVIG use in the treatment of GBS of acute motor axonal type in which symptoms improved drastically but may not have removed the unknown contributing factors for pathogenesis of facial palsy.

References:


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